SPNR
Sociedade Portuguesa de Neurorradiologia Diagnóstica e Terapêutica

XVII CONGRESSO

SPNR Sociedade Portuguesa de Neurorradiologia Diagnóstica e Terapêutica

Resumos

NEUROPEDIATRIA

25 e 26 de novembro 2022

Ordem dos Médicos, Lisboa

Organizado pelo
Serviço de Neurorradiologia
do Hospital de Santa Maria.

Índice


SPNR
Sociedade Portuguesa de
Neurorradiologia
Diagnóstica e Terapêutica

Boas-Vindas

Caros sócios e Colegas Neurorradiologistas

É com enorme prazer que convido todos a juntarem-se em 25 e 26 de novembro no nosso XVII Congresso de Neurorradiologia em Lisboa na sede da Ordem dos Médicos.

Será, a exemplo dos últimos anos, uma nova demonstração da vitalidade da Neurorradiologia Portuguesa.

A organização ficou a cargo dos Colegas do Hospital de Santa Maria. A Graça Sá, a Luisa Biscoito e restantes médicos do Serviço de Neurorradiologia do HSM constituiram-se em Comissão Organizadora, pensaram numa série de temas da actualidade e juntaram um conjunto de prelectores nacionais e internacionais, conseguindo fazer um excelente programa sobre Neurorradiologia Pediátrica.

Espero que da Vossa parte também haja uma grande participação com Comunicações Orais e Posters sobre os mais variados temas da Neurorradiologia, tanto Diagnóstica como Terapêutica.
O jantar do Congresso será mais uma vez na Ordem dos Médicos.

Mãos à obra!

Esperando ver-Vos a todos e todas em Lisboa em fins de novembro, aceitem as cordiais saudações.

Rui Manaças
Presidente da SPNR

Comissões

Comissão Organizadora HSM-CHLN

Graça Sá
Luísa Biscoito
Carlos Morgado
Sofia Reimão
Lia Neto
Gonçalo Basilio
Carla Guerreiro
Manuel Correia

Comissão Organizadora Internos HSM-CHLN

Filipa Proença
João Madureira
Inês Carneiro
David Berhanu
Inês Sá
Guilherme Martins
Joana Freitas
Hugo Cadilha
Angelo Dias

Comissão Científica

Ana Mafalda Reis (H Pedro Hispano)
Carla Conceição (CHLC-HDE)
Cristina Rios (HGO)
Graça Sá (CHLN-HSM)
Inês Carreiro (CHUC)
Inês Caldeira (H Evora)
José Fonseca (CHS João)
Luisa Biscoito (CHLN-HSM)
Pedro Vilela (HBA)
Rui Manaças (HFF)
Teresa Garcia (CHUC)

Comissão de Discussão e Avaliação de Posters

Joana Graça
Tiago Baptista
Carlos casimiro

Secretariado:

Isabel Gregório - Sociedade Portuguesa de Neurorradiologia Diagnóstica e Terapêutica – SPNR
R. Coronel Júlio Veiga Simão, 3025-307 Coimbra – Portugal
Telefone: +351 239497160 | Telemóvel: +351 927811456
Telemóvel SPNR MB WAY: 925 246 670
secretariado.spnr@gmail.com

Logística:

Organideia, Lda.
https://organideia.pt
geral@organideia.pt

Informações Úteis

Apresentação de Comunicações Orais (Slide Desk)

Se vai apresentar uma Comunicação Oral, por favor, entregue o seu trabalho em USB disk ou outro suporte digital o quanto antes no "Slide Desk", não poderão ser usados os próprios computadores para fazer a apresentação.

Horário do Slide Desk: 8:30h-18h - 25 de novembro | 8:30h-15:30h - 26 de novembro.
Terá à sua disposição um computador onde poderá rever a sua apresentação, caso o pretenda.

Apresentação de Póster

Se vai apresentar um póster, por favor afixe-o no placard que tem o número do seu póster. Pode consultar na lista de pósteres qual é o número do seu. Terá fita adesiva adequada no placard para a afixação.

Acesso à Internet

Auditório: Wi-Fi network: **WIFI_SPNR** | Password: **omsul2021**
Área dos Almoços/C-B: Wi-Fi network: **MEO-FD0D03** | Password: **883JYGJEHB4**

Coffee Breaks e Almoços de trabalho

Espaço anexo no jardim

Exposição Técnica

Espaço anexo no jardim

Jantar do Congresso

O jantar terá lugar na Ordem dos Médicos pelas 20:30h, 25 de novembro de 2022.

Programa

Sexta-feira, 25 novembro 2022

08:30h - Abertura do Secretariado

09:00h - Abertura do Congresso

SESSÃO 1 - Segurança e proteção radiológica em imagiologia pediátrica + RM Fetal

Moderadores: Graça Sá e Sofia Reimão

9:15h-9:35h - Neuroimagem em pediatria: questões de segurança e contrastes - Filipa Proença

9:35h-9:55h - RM Fetal Crânio e Coluna - principais indicações e 'check-list' - Carla Conceição

10:05h-10:25h - Comunicações orais

10:05h-10:15h - ID8 | "FEED AND WRAP" - A NOVEL NON-SEDATION MRI PROTOCOL FOR INFANTS - Maria Inês Prisco, *Unidade Local de Saúde de Matosinhos*

10:15h-10:25h - ID23 | THE PREDICTIVE VALUE OF THE WEEKE SCORE IN THE PROGNOSIS OF HYPOXIC-ISCHEMIC ENCEPHALOPATHY TREATED WITH THERAPEUTIC HYPOTHERMIA - Liliana Igreja, *Centro Hospitalar Universitário do Porto (CHPorto)*

10:30h-11:00h - **Café/Discussão de Posters** (Joana Graça, Tiago Baptista, Carlos Casimiro)

SESSÃO 2

Moderadores: Rui Manaças e Gonçalo Basílio
11:00h-11:30h - Aspetos Neurorradiológicos das Leucodistrofias - Leandro Lucato (Brasil)

11:40h-12:10h - Comunicações orais

11:40h-11:50h - ID29 | NEUROIMAGING IN THE DIAGNOSIS OF L-2-HYDROXYGLUTARIC ACIDURIA - Sílvia Pino Martins, *Centro Hospitalar e Universitário de Coimbra*

11:50h-12:00h - ID52 | CLCN-2, GLIALCAM AND MLC1 MUTATIONS RELATED CNS DISEASES - DISTINCT CLINICAL AND MRI PHENOTYPES IN A COMMON MOLECULAR PATHWAY - Vasco Sousa Abreu, *Centro Hospitalar Universitário do Porto*

12:00h-12:10h - ID13 | CENTRAL NERVOUS SYSTEM INVOLVEMENT IN PRIMARY IMMUNODEFICIENCY DISORDERS - Pedro Sousa Brandão, *Department of Neuroradiology, Centro Hospitalar Universitário de Lisboa Central, E.P.E., Lisbon*

12:10h-12:30h - Simpósio Indústria MC Medical "First generation carotid stents vs. second generation - Clinical comparative results", José Luís Gomez, MD

12:30h-14:00h - Almoço

14:00h-14:30h - Homenagem ao Professor Doutor Jaime Cruz Maurício

Moderador: Rui Manaças

Fernando Costa Reis, Eduardo Medina e Carlos Morgado

SESSÃO 3 - Malformações Congénitas

Moderadores: Pedro de Melo Freitas e Sofia Reimão

14:30h-14:45h - Craniossinostoses - abordagem diagnóstica - Inês Caldeira

14:45h-15:00h - Craniossinostoses - abordagem terapêutica - Cláudia Faria (Neurocirurgia)

15:00h-15:15h - Disrafismos - Teresa Morais

15:30h-16:20h - Comunicações orais

15:30h-15:40h - ID26 | CONGENITAL NON-TUMORAL DISORDERS OF THE HYPOTHALAMIC-PITUITARY AXIS - João Saraiva, *Hospital de Braga EPE*

15:40h-15:50h - ID25 | PITUITARY GLAND HEIGHT AS A PREDICTOR OF RESPONSE TO GROWTH HORMONE THERAPY IN CHILDREN - Rui Duarte Armindo, *Hospital Beatriz Ângelo*

15:50h-16:00h - ID42 | MULTIPLE SCLEROSIS: IS IT THE SAME IN CHILDREN? - A PEDIATRIC CENTER EXPERIENCE - Gonçalo Gama Lobo, *Hospital de São José, Centro Hospitalar Lisboa Central*

16:00h-16:10h - ID10 | CAN YOU SPOT THE DIFFERENCE? DISTINGUISHING MULTIPLE SCLEROSIS AND ACUTE DISSEMINATED ENCEPHALOMYELITIS ON FIRST EPISODE MAGNETIC RESONANCE IMAGING - Ricardo João Gaspar Pires, *Functional Unit of Neuroradiology, Department of Medical Imaging, Coimbra Hospital and University Centre*

16:10h-16:20h - ID19 | PEDIATRIC MOGAD - SINGLE-CENTER IMAGIOLOGIC PATTERNS REVIEW - João Pedro Freitas Gonçalves, *Neuroradiology Department, Hospital de São José, Centro Hospitalar Lisboa Central*

16:25h-16:45h - Simpósio Indústria Johnson & Johnson ""Tough" Thrombectomies - How to make it easier?" - Dr[a]. Sarah Power

16:45h-17:15h - Café/Discussão de Posters (Joana Graça, Tiago Baptista, Carlos Casimiro)

SESSÃO 4 - Tumores e Epilepsia

Moderadores: Rui Pedro Pais e Tiago Baptista

17:15h-17:30h - Classificação de tumores pediátricos da OMS: o que há de novo - Carla Guerreiro

17:30h-17:45h - A estratégia diagnóstica dos tumores encefálicos (fluxograma) - Graça Sá

17:45h-18:00h - Epilepsia na idade pediátrica - Carlos Morgado

18:15h-19:00h Comunicações orais

18:15h-18:25h - ID28 | VERTEBRAL ANEURYSMAL BONE CYST (ABC) - COMPLETE NEURORADIOLOGIC DIAGNOSTIC AND TREATMENT FROM A TO Z - João Tarrio - *Centro Hospitalar Universitário do Porto, Neuroradiology Department*

18:25h-18:35h - ID55 | DISTINCT VOLUMETRIC SIGNATURE ALONG THE LONGITUDINAL AXIS OF THE HIPPOCAMPUS IN THE ALZHEIMER'S DISEASE CONTINUUM DEFINED BY CEREBROSPINAL FLUID BIOMARKERS PROFILE - Torcato Meira - *Neuroradiology Department of Hospital de Braga*

18:35h-18:45h - ID31 | ANTE-MORTEM MAGNETIC RESONANCE IMAGING GREY-WHITE MATTER CONTRAST REGIONAL SIGNATURES OF ALZHEIMER'S DISEASE NEUROPATHOLOGY - Francisco Almeida - *Life and Health Sciences Research Institute (ICVS), School of Medicine, University of Minho, Braga*

18:45h-18:55h - ID50 | MR FINDINGS IN THE FOLLOW-UP OF TRANSORAL LASER MICROSURGERY FOR LARYNGEAL CANCER - Bruno Cunha - *Neuroradiology Department, Centro Hospitalar Universitário de Lisboa Central*

Sábado, 26 novembro 2022

08:30h - Abertura do Secretariado

SESSÃO 5 - Vascular

Moderadores: Isabel Fragata e Teresa Garcia

09:00h-09:15h - Padrões imagiológicos do AVC na criança e abordagem terapêutica - Luísa Biscoito

09:15h-09:30h - Malformações vasculares na criança - Pedro Vilela

09:30h-09:45h - Tratamento endovascular dos Retinoblastomas - Egídio Machado

09:55h-10:30h - Comunicações orais

09:55h-10:05h - ID35 | ENDOVASCULAR TREATMENT OF BRAIN ARTERIOVENOUS MALFORMATIONS IN PEDIATRIC PATIENTS: A SINGLE CENTER EXPERIENCE - Gonçalo Almeida, *Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central*

10:05h-10:15h - ID7 | ACUTE CAROTID ARTERY STENTING VERSUS BALLOON ANGIOPLASTY FOR TREATMENT OF TANDEM OCCLUSIONS IN ACUTE ISCHEMIC STROKE - ANALYSIS OF A TERTIARY CENTRE - Inês Carneiro, *Serviço de Imagiologia Neurológica, Centro Hospitalar Universitário Lisboa Norte*

10:15h-10:25h - ID47 | THE CHALLENGE OF DIFFERENT CLINICAL AND IMAGIOLOGICAL PRESENTATIONS IN MOYAMOYA VASCULOPATHY - Henrique Coimbra Queirós, *Centro Hospitalar Universitário de Coimbra*

10:30h-11:00h - Café/Discussão de Posters (Joana Graça, Tiago Baptista, Carlos Casimiro)

SESSÃO 6

Moderadores: Ana Mafalda Reis e Cristina Rios

11:00h-11:30h - Mimickers of HIE - Andrea Rossi (Itália)

11:40h-12:00h - Comunicações orais

11:40h-11:50h - ID18 | PEDIATRIC HYDROCEPHALUS: A RETROSPECTIVE ANALYSIS OF THE CLINICAL AND RADIOLOGICAL FEATURES IN CHILDREN - Carolina Chaves - *Unidade Funcional de Neurorradiologia, Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra*

11:50h-12:00h - ID51 | PEDIATRIC ORBITAL VASCULAR ANOMALIES - Tiago Lorga, *Serviço de Neurorradiologia do Centro Hospitalar Lisboa Ocidental*

12:40h-13:00h - Simpósio Indústria Bayer "Why do we use gadobutrol in neuroimaging? Considerations based on literature and on a large neuroradiology center experience" - Dra. Claudia Godi, *San Raffaele Hospital, Milan, Italy*

13:00h-14:00h - Almoço do Congresso

SESSÃO 7

Moderadores: Inês Carreiro e Pedro Vilela
14:00h-14:30h - ADEM, MOG e NMO - é possível o diagnóstico diferencial por imagem? - Carolina Tramontini (Colômbia)

14:40h-15:30h - O seu caso mais difícil. Casos desafiantes PEARL and PITFALLS

Moderadores: Graça Sá e Luísa Biscoito

15:30h-16:10h - Comunicações Orais

Moderadores: Lia Neto e Carla Guerreiro

15:30h-15:40h - ID48 | NEUROIMAGING FINDINGS IN SICKLE CELL DISEASE - Francisca Sena Batista, *Hospital Garcia de Orta*

15:40h-15:50h - ID44 | UNILATERAL MICROBLEEDS IN PEDIATRIC POPULATION: A CLUE TO THINK IN SCLERODERMA - Mariana Ribeiro dos Santos, *Serviço de Neurorradiologia do Hospital de Braga*

15:50h-16:00h - ID27 | UNDER PRESSURE: ABORDAGEM CLINICO-RADIOLÓGICA DA HIPERTENSÃO INTRACRANIANA IDIOPÁTICA EM IDADE PEDIÁTRICA - Liliana Igreja, *Centro Hospitalar Universitário do Porto*

16:00h-16:10h - ID41 | THIS SHOULDN'T BE HERE! - A PICTORIAL REVIEW OF FOREIGN BODIES IN NEURORADIOLOGY - Gonçalo Gama Lobo, *Neuroradiology Department, Hospital de São José, Centro Hospitalar Lisboa Central*

16:30h-16:45h - Entrega dos Prémios

17:00h - Encerramento do Congresso

Comunicações Orais

ID8 | "FEED AND WRAP" – A NOVEL NON-SEDATION MRI PROTOCOL FOR INFANTS

Maria Inês Prisco - inesprisco98@hotmail.com
Unidade Local de Saúde de Matosinhos
André Miranda, Joana Nunes, Ana Filipa Geraldo
Centro Hospitalar Vila Nova de Gaia, 4434-502, Portugal

Introduction: Magnetic resonance (MR) imaging remains an unparalleled diagnostic tool for the assessment of central nervous system disorders, particularly in the pediatric population, following the principle of use of radiation as low as reasonably achievable (ALARA). However, its widespread applicability is limited by both long study duration and motion artifacts susceptibility. While sedation may present as an alternative, it not only requires human and technical resources, but also concerns remain on the anesthetic agents impact in long-term brain development. To mitigate these limitations, non-sedation protocols based on timed feed, induction of natural sleep and immobilization have been suggested. We sought to establish the usefulness of this protocol in MR imaging central nervous system pathologies in infants.
Materials and Methods: We implemented a non-sedation "Feed and wrap" protocol at the Diagnostic Neuroradiology Unit of Centro Hospitalar de Vila Nova de Gaia from January to September 2022. Clinically stable inpatient and outpatient infants, younger than 6 months without feeding difficulties, were included. A shorter and quieter imaging protocol was developed and included multiplanar T1 and T2 and axial T1 SE, DWI and T2* sequences.
Results: We developed a protocol based on prolonged fasting and restricted sleep before examination. Proper position and immobilization was performed, using heating devices and customized ear tampons, under cardiac and respiratory monitoring. A total of 10 infants were evaluated, with median age at examination of 1,8 months-old (min. 6 days-old, max. 3 months-old) and average examination time of 28 minutes (min. 14 minutes, max. 47 minutes). We report a 90% success rate in obtaining quality images and proper answering to clinical queries, with no need of repeated MR imaging under sedation.
Conclusions: Feed and wrap is an efficient alternative in MR screening in clinically stable newborns when performed by experienced personnel, minoring exposure to anesthetic agents in infancy.

ID23 | THE PREDICTIVE VALUE OF THE WEEKE SCORE IN THE PROGNOSIS OF HYPOXIC-ISCHEMIC ENCEPHALOPATHY TREATED WITH THERAPEUTIC HYPOTHERMIA

Liliana Igreja - lilianaigreja@gmail.com

Centro Hospitalar Universitário do Porto (CHPorto), Largo do Prof. Abel Salazar, 4099-001, Porto, Portugal

Adriana Ferreira[3], Rita Gomes[2,3], Bebiana Sousa[2,3], Ana Novo[3], Carmen Carvalho[3], Elisa Proença[3], José Eduardo Alves[1]

1 Neuroradiology Department, Centro Hospitalar Universitário do Porto (CHPorto), Porto, Portugal;
2 Department of Pediatrics, Centro Materno-Infantil do Norte (CMIN), Centro Hospitalar Universitário do Porto (CHUPorto), Porto, Portugal;
3 Intensive Neonatal Care Unit, Department of Neonatology and Pediatric Intensive Care, Centro Materno-Infantil do Norte (CMIN), Centro Hospitalar Universitário do Porto (CHUPorto), Porto, Portugal

Introduction: Neuroimaging plays an essential role in hypoxic-ischemic encephalopathy (HIE), helping to determine the timing and extent of lesions, to guide future therapeutic decisions, and to predict neurological prognosis. Different magnetic resonance imaging (MRI) scores have been validated in moderate/severe HIE, yet little applied in clinical practice. Our aim was to retrospectively apply the Weeke score in a group of neonates clinically diagnosed with HIE treated with therapeutic hypothermia (TH) and evaluate its prognostic value.

Material and Methods: An analysis of patients diagnosed with HIE and who underwent TH at a tertiary neonatal intensive care unit between January/2012 and July/2020 was conducted, and the Weeke score calculated based on the MRI findings. Demographic data, pre- and perinatal information, neurological sequelae at 12 and 24 months and mortality were collected. Mann-Whitney, Kruskal-Wallis, and Pearson correlation coefficient tests were used in the statistical analysis.

Results: 29 patients with a median gestational age of 39 weeks and 55% male were included and underwent MRI on average at day 6 (3-13) of life. 21 patients were clinically assessed at 12 and 24 months.

Clinical seizures on admission correlated significantly with gray matter (GM) subscore ($p=0.050$) and total score ($p=0.046$). The aEEG pattern at 48h correlated significantly with GM subscore and total score ($p=0.003$ and $p=0.004$), as did the aEEG after TH with GM, white matter (WM) and total scores ($p=0.006$, $p=0.011$, $p=0.005$). Thompson score after TH showed strong correlation with GM subscore ($p=0.791$) and WM to moderate degree ($p=0.613$).

After therapeutic hypothermia, Thompson score, GM subscore and total score correlated with mortality ($p=<0.001$) and motor sequelae ($p=0.048$, $p=0.002$ and $p=0.004$).

Conclusions: Our findings confirm the predictive value of the Weeke Score for neurological prognosis and mortality in HIE treated with therapeutic hypothermia, allowing detailed characterization of the imaging findings and grading of its severity.

ID29 | NEUROIMAGING IN THE DIAGNOSIS OF L-2 HYDROXYGLUTARIC ACIDURIA

Sílvia Alexandra Reigada Pino Martins - silvialexr@hotmail.com
Centro Hospitalar e Universitário de Coimbra, Praceta Professor de Mota Pinto 3400-561 Coimbra, Portugal
Ana Carolina Chaves, Bruno Gomes, Sílvia Carvalho, Luísa Diogo
1, 2, 3 Centro Hospitalar e Universitário de Coimbra, Praceta Professor de Mota Pinto 3400-561 Coimbra, Portugal
4 Hospital Pediátrico de Coimbra, Av. Afonso Romão, 3000-602 Coimbra, Portugal

Introduction: L-2-hydroxyglutaric aciduria (L-2-HGA) is a rare recessive autosomal genetic disease that results from mitochondrial L-2-hydroxyglutarate dehydrogenase deficiency caused by mutations in L2HGDH gene. The symptoms usually start in the early infancy, and include psychomotor delay, progressive cerebellar ataxia, dysarthria, dystonia and cognitive deterioration. Macrocephaly is found in most patients.
Brain magnetic resonance imaging (MRI) findings are virtually pathognomonic for the disease and typically show subcortical leukodystrophy and lesions in globus pallidus and dentate nucleus.
Materials and Methods: Retrospective analysis of the clinical records of patients diagnosed with L-2-HGA and followed at Centro Hospitalar e Universitário de Coimbra.
Results: Six patients with with L-2-HGA were included in our study, with the median age of 23.8 years and 3 were females. All patients had clinical manifestations that started in the first years of life, however the diagnose was made before 2 years-old in only one (15 months). In the others, the diagnose was made at 3 (4), 11 (1) and 12 (1) years-old. The main symptoms were psychomotor retardation (6/6), extrapyramidal symptoms (6/6), cerebellar ataxia (5/6), seizures (3/6) and pyramidal symptoms (2/6). Three patients had macrocephaly.
Five patients underwent brain MRI and in one patient only brain computed tomography scan was performed (subcortical leukodystrophy was present).
Characteristic MRI findings included subcortical cerebral white matter abnormalities (5/5), T2 hyperintensities of the dentate nucleus (5/5), globus pallidus (4/5), putamen and caudate nucleus (1/5).
Conclusion: Diagnosis of L-2-HGA is usually belated due to slow progression of the disease.
Clinical manifestations like macrocephaly, global psychomotor delay, extrapyramidal symptoms and cerebellar ataxia with the identification of typical lesions on brain MRI should point to the diagnosis of L-2-HGA.

ID52 | CLCN-2, GLIALCAM AND MLC1 MUTATIONS RELATED CNS DISEASES – DISTINCT CLINICAL AND MRI PHENOTYPES IN A COMMON MOLECULAR PATHWAY

Vasco Sousa Abreu - vrlsa1993@gmail.com

Centro Hospitalar Universitário do Porto, Largo do Prof. Abel Salazar, 4099-001 Porto, Portugal

José Silva[1], João Tarrio[1], Carla Conceição [2], José Eduardo Alves[1]

1 Department of Neuroradiology, Centro Hospitalar Universitário do Porto, Portugal

2 Department of Neuroradiology, Hospital Dona Estefânia, Centro Hospitalar Universitário Lisboa Central, Portugal

Introduction: Defects affecting ion and water homoeostasis may lead to different forms of CNS disease. The chlorine channel 2 (ClCN2) spans the cell membrane, being fundamental in transepithelial transport, homeostasis, and cell excitability. GlialCAM, a subunit of ClNC2, and membrane protein MLC1, both contribute for stabilizing this channel, forming a complex for astrocytic regulatory volume decrease after cell swelling.

However, ClCN2 mutations fundamentally differ from GlialCAM and MLC2, and our aim is to review their distinct clinical-imaging findings through three genetically proven pediatric patients.

Results: An 18-month-old premature boy had abnormal routine cranial-US. MRI showed diffuse white matter (WM) involvement, predominantly in the internal capsules, cerebral peduncles, and middle cerebellar peduncles, with associated diffusion restriction. Clinically he only presentes slight ataxia. Genetics confirmed ClCN2 mutation. A 12-month-old boy with increased head circumference undergoes MRI showing extensive diffuse supratentorial WM involvement, with increased diffusibility. He remained asymptomatic and imaging findings regressed in follow-up MRIs. GlialCAM mutation was genetically proven.

An 11-month-old boy with difficult-to-control epilepsy. MRI showed extensive WM involvement, with increased diffusibility and temporal subcortical cysts. Genetics confirmed MLC1 mutation. Clinically, there is significant developmental delay, epilepsy, and skeletal deformations.

Discussion: Our findings are in line with the literature: the first two mutations have a benign clinical course, unlike MLC1 mutation, typically leading to worse clinical outcomes.

MRI findings are also distinctive: ClCN2-related disease preferentially affects WM structures that are (relatively) spared by MLC1/GlialCAM-related diseases, namely the corpus callosum, internal capsules, brainstem, and cerebellum. Another intriguing question concerns the size of the myelin vacuoles: while in MLC1/GlialCAM-related diseases, ADC-values of the affected WM are highly increased reflecting large vacuoles/ increased extracellular spaces, ClCN2 mutation leads to low ADC-values due to small intramyelinic vacuoles/oedema.

Although the exact mechanisms around ClCN2-GlialCAM-MLC1-pathway are unclear, each genetic mutation leads to distinct clinical-imaging phenotypes.

ID13 | CENTRAL NERVOUS SYSTEM INVOLVEMENT IN PRIMARY IMMUNODEFICIENCY DISORDERS

Pedro Sousa Brandão - pedrodsbrandao@gmail.com

Department of Neuroradiology, Centro Hospitalar Universitário de Lisboa Central, E.P.E., Lisbon, Portugal

Sofia Bettencourt[1], João Farela Neves[2], Carla Conceição[1]

1 Department of Neuroradiology, Centro Hospitalar Universitário de Lisboa Central, E.P.E., Lisbon, Portugal

2 Department of Pediatrics, Centro Hospitalar Universitário de Lisboa Central, E.P.E., Lisbon, Portugal

Introduction: Primary immunodeficiency disorders (PIDs), also referred to as inborn errors of immunity, are a heterogeneous group of more than 480 sporadic or inherited conditions caused by damaging germline variants in genes related to immune system development and function. Although individually rare, their prevalence as a group is estimated to range between 1/1,000 and 1/5,000.

Based on shared pathogenesis and/or clinical phenotypes, PIDs are classified into ten categories according to the 2022 Update on the Classification from the International Union of Immunological Societies Expert Committee. Central nervous system involvement may be primary, as a core feature in certain syndromes, or secondary due to infections or immune dysregulation (neoplastic, inflammatory or autoimmune).

The broad clinical spectrum of PIDs often poses a diagnostic challenge at presentation, and imaging plays an important role in the differential diagnosis and in evaluating the full extent of its primary and secondary manifestations.

Methods & Results: A retrospective analysis of pediatric patients diagnosed or followed-up with PIDs and central nervous system involvement in Hospital Dona Estefânia, Portugal, was conducted. Clinical and imaging findings of the selected cases were discussed.

We provide a pictorial review of the main neurological manifestations in PIDs and accompanying imaging features according to their etiopathogenesis.

Conclusion: Increased susceptibility to infections is the most well-recognized feature of PIDs, and it is the history of atypical/opportunistic, persistent, or recurrent infections that typically suggests the diagnosis.

However, physicians and radiologists should be aware of additional potential neurological manifestations of PIDs, which may be grouped as autoimmune, vascular, malignant or neurodegenerative, or part of the disease itself. Recognizing the diverse imaging patterns of these disorders may assist in early diagnosis, and it is of great importance in guiding management approaches and treatment to prevent and reduce complications.

ID26 | CONGENITAL NON-TUMORAL DISORDERS OF THE HYPOTHALAMIC-PITUITARY AXIS

João Saraiva - joaosaraiva.md@gmail.com
Hospital de Braga EPE, 4710-367, Portugal
João Paulo Soares-Fernandes
Hospital de Braga EPE, 4710-367, Portugal

Introduction: in children, the hypothalamic-pituitary (HH) axis regulates growth, puberty, and maintains body homeostasis by integrating internal/external stimuli and controlling the autonomic nervous, endocrine and somatic systems. HH axis dysfunction includes both acquired (tumoral, inflammatory/granulomatous, or vascular) and congenital causes. Congenital disorders of the HH axis may be associated with endocrinopathies, vision impairment, hyposmia/anosmia, seizures and midline cranio-facial anomalies; hypopituitarism is a dynamic condition with potential long term effects (eg. short stature).
Methods: we collected congenital anomalies of the HH axis either diagnosed by our neuroradiology department or listed in the individual databases of our attending neuroradiologists.
Results: we selected six structural abnormalities depicting the broad spectrum of congenital non-tumoral disorders of the HH axis; in particular - incomplete closure of the neural tube and persistence of the craniophayngeal canal with meningocele, pituitary gland duplication, ectopic posterior lobe, pituitary stalk interruption syndrome, septo-optic dysplasia and hypothalamic hamartoma.
Conclusions: although many congenital anomalies of the HH axis reflect malformative or disruptive - hence irreversible - processes, their clinical repercussion (at presentation and evolution) is variable and, in some cases, as if anticipated based on certain cerebral and HH structural abnormalities identified on magnetic resonance imaging. Consequently, neuroradiology plays a critical role by increasing chances of a timely diagnosis, early definition of prognosis and inititation of proper treatment.

ID: 25 | PITUITARY GLAND HEIGHT AS A PREDICTOR OF RESPONSE TO GROWTH HORMONE THERAPY IN CHILDREN

Rui Duarte Armindo - ruimigueldarmindo@gmail.com

Hospital Beatriz Ângelo, Avenida Carlos Teixeira, 3 2674-514 Loures, Portugal

Ana Rute Manuel[1], Carolina Folques[2], Alice Mirante[3], Joana Pinto[3]

1 Hospital Prof. Doutor Fernando Fonseca E.P.E. IC 19, 2720-276 Amadora, Portugal

2 Centro Hospitalar de Leiria, Rua das Olhalvas, 2410-197 Leiria, Portugal

3 Centro Hospitalar e Universitário de Coimbra - Hospital Pediátrico, Rua Dr. Afonso Romão, 3000-602 Coimbra, Portugal

Introduction: There are no validated radiological predictors of response to growth hormone (GH) therapy in children with growth disorders. Most patients suggested for GH therapy are submitted to a brain MRI to exclude secondary causes of GH deficiency, so it would be useful to identify imaging markers of prognosis.

Methods: We retrospectively reviewed all cases of GH therapy initiated and completed between 2010-2019 at a tertiary pediatric hospital. Patients with missing clinical or MR data were excluded. The anterior pituitary was measured in the coronal plane and classified in: average size for sex and age, more than one standard deviation (SD) below average, and more than two SD below average. The height-for-age z-score variation was considered to represent the response to GH therapy and was analyzed according to the anterior pituitary size.

Results: Of a sample of 83 patients, 44 met the inclusion criteria (27 boys and 17 girls). The median age at the time of MRI was 12 years-old. Anterior hypophyseal heights were normal for age and gender in 63.6%, between one and two SD below average (1SDBA) in 27.3%, and more than two SD below average (2SDBA) in 9.1%. The median duration of GH treatment was of 55 months. The median height-for-age z-score variation was 0.79, 0.83, and 1.05, respectively for the normal, 1SDBA, and 2SDBA groups. These differences were not statistically significant (p=0.83). Response to therapy was not related to its duration, age at the beginning of therapy, or presence of other radiological findings.

Conclusion: There was a tendency for better response to GH therapy in patients with hypoplastic anterior pituitary glands, without statistical significance. Larger studies are needed to assess an eventual relationship between hypophyseal size and response to GH therapy, which could be an important factor in patient selection for this treatment.

ID42 | MULTIPLE SCLEROSIS: IS IT THE SAME IN CHILDREN? – A PEDIATRIC CENTER EXPERIENCE

Gonçalo Gama Lobo - goncalogamalobo@gmail.com
Hospital de São José, Centro Hospitalar Lisboa Central, Lisbon, Portugal
João Gonçalves, Sofia Ribeiro, Rita Silva, Carla Conceição
1 Hospital de São José, Centro Hospitalar Lisboa Central, Lisbon, Portugal
2 Hospital de São José, Centro Hospitalar Lisboa Central, Lisbon, Portugal
3 Hospital Dona Estefânia, Centro Hospitalar Lisboa Central, Lisbon, Portugal
4 Hospital Dona Estefânia, Centro Hospitalar Lisboa Central, Lisbon, Portugal

Introduction: Multiple sclerosis (MS) typically arises between the ages 20-40, but its diagnosis has increased at earlier ages. Between 3-10% of MS cases present in pediatric-age and <1% before 10 years-of-age. Diagnosis is challenging due to transient demyelinating events and other conditions with similar symptoms and imaging findings. Differences from the "classical" adult MS are described regarding epidemiology, symptoms, course, incapacity, and cognitive impairment. The imaging characteristics are also distinctive and the diagnosis criteria as well as the available therapies have some particularities.
Objectives and Methods: Clinical and imaging characterization of pediatric-onset MS (POMS) cases from Hospital Dona Estefânia. We retrospective analyzed the clinical-data and brain/spine MRI findings of our POMS data base from 2015 to 2020. **Results:** We found 13 cases of POMS, 10 female. Onset-age ranges from 11-17 (mean 13,77), 2 patients 10 lesions in 10 patients (77%) and >20 lesions in 7 patients (54%). 77% had infratentorial lesions and 10 patients had at least one enhancing lesion. 5 patients (38%) had confluent lesions, 3 patients had thalamic involvement and all patients had spinal cord lesions.
Conclusions: POMS is infrequent, however growingly reported, which may suggest that was underdiagnosed in the past. This diagnosis is challenging due to the higher prevalence of transitory demyelinating events and other pathologies with similar symptoms and imaging findings. POMS has some particular clinical and imaging features that have been described in the literature and are present in the majority of our cases. Early identification is essential for a prompt therapy to modify the disease course.

ID10 | CAN YOU SPOT THE DIFFERENCE? DISTINGUISHING MULTIPLE SCLEROSIS AND ACUTE DISSEMINATED ENCEPHALOMYELITIS ON FIRST EPISODE MAGNETIC RESONANCE IMAGING

Ricardo João Gaspar Pires - ricardogasparpires@gmail.com

Functional Unit of Neuroradiology, Department of Medical Imaging, Coimbra Hospital and University Centre, Praceta Professor Mota Pinto, 3004-561 Coimbra, Portugal

Filipe Palavra[1], Daniela Jardim Pereira[2]

1 Centre for Child Development - Neuropediatrics Unit, Pediatric Hospital, Coimbra Hospital and University Centre, Praceta Professor Mota Pinto, 3004-561 Coimbra, Portugal

2 Functional Unit of Neuroradiology, Department of Medical Imaging, Coimbra Hospital and University Centre, Praceta Professor Mota Pinto, 3004-561 Coimbra, Portugal

Introduction/Objectives: Acute disseminated encephalomyelitis (ADEM) is a demyelinating disorder, more frequently occurring in the paediatric population. While typically monophasic, the clinical presentation can overlap with a first manifestation of multiple sclerosis (MS). Differentiating these conditions is crucial to therapeutic decision and to establish long term risk of recurrent demyelination.

Materials and methods: We revised MRI scans acquired at first clinical episode of 27 children diagnosed with MS and 10 with ADEM. T2/FLAIR hyperintense lesions were quantified and categorized regarding location and characteristics, through visual assessment. Additionally, we performed automatic segmentation with lesion segmentation toolbox (LST) for SPM and volBrain (https://www.volbrain.upv.es). Statistical analysis was performed using IBM-SPSS (v26).

Results: Periventricular lesions were consistently more numerous in MS, on both visual and automatic assessment. Other statistically relevant characteristics included deep grey matter lesions (ADEM); well-defined lesions, oval lesions and black-holes (MS). Infratentorial lesions were more common in MS on visual assessment, while with the automatic methods there was no statistical difference, but these often failed to detect infratentorial lesions. Total lesion number and volume did not differ significantly between both diseases. Presence of oval lesions and number of periventricular lesions were the most significant features to differentiate MS from ADEM, losing statistical power when considering only cases with ≤ 2 lesions. Regarding the automatic tools, LST missed several infratentorial and juxtacortical lesions; volBrain was also unreliable for infratentorial lesions, but more accurate on the supratentorial compartment, although overestimating juxta-cortical lesions.

Conclusion: Although definitive distinction between ADEM and MS on first MRI remains challenging, careful analysis of lesion characteristics provides useful clues for the diagnosis. Automatic tools still have considerable limitations and must be used with caution in clinical practice.

ID19 | PEDIATRIC MOGAD - SINGLE-CENTER IMAGIOLOGIC PATTERNS REVIEW

João Pedro Freitas Gonçalves - joao.gonc94@gmail.com

1 Neuroradiology Department, Hospital de São José, Centro Hospitalar Lisboa Central, R. José António Serrano, 1150-199 Lisboa, Portugal

2 Unidade de Neurorradiologia, Hospital Central do Funchal, Av. Luís de Camões 6180, 9000-177 Funchal, Portugal

Gonçalo Gama Lobo[1], Alexandra Rodrigues[1, 2], Teresa Painho[3], Teresa Morais[1]

1 Neuroradiology Department, Hospital de São José, Centro Hospitalar Lisboa Central, Lisbon, Portugal

2 Unidade de Neurorradiologia, Hospital Central do Funchal, Funchal, Portugal

3 Pediatric Neurology Department, Hospital Dona Estefânia, Centro Hospitalar Lisboa Central, Lisbon, Portugal

Introduction: Myelin oligodendrocyte glycoprotein antibody–associated disease (MOGAD) is a CNS inflammatory disease that was recently recognized as an independent entity. It differs from Neuromielitis optica spectrum disorder (NMOSD), Multiple Sclerosis (MS) and seronegative Acute Disseminated Encephalomyelitis (ADEM), on the account of having its own biomarker (IgG anti-MOG), as well as some some characteristic clinical and imagiologic findings.

We aim to present the typical imagiologic patterns of MOGAD, as well as tips and tricks on how to differentiate them from NMOSD, MS and ADEM.

Methods: We retrospectively reviewed the clinical and MRI findings of all the cases of IgG anti-MOG positive pediatric patients (n=5) that occurred between 2017 and 2022 in our tertiary center.

Results: We present 5 cases that illustrate the typical imagiologic patterns found in MOGAD pediatric population, from younger to older patients. The younger patients usually presented with an ADEM-like pattern with or without transverse myelitis, while the older ones had optic neuritis as a clinical and MRI finding.

Conclusions: MOGAD has a different clinical course and prognosis when compared to other CNS inflammatory diseases. As Neuroradiologists it is important to learn how to recognize the characteristic MRI findings of this recent entity, so as to alert the clinicians for that possibility and allow a better diagnostic workup and prompt treatment.

ID28 | VERTEBRAL ANEURYSMAL BONE CYST (ABC) – COMPLETE NEURORADIOLOGIC DIAGNOSTIC AND TREATMENT FROM A TO Z

João Tarrio - joaotarrio@gmail.com

Centro Hospitalar Universitário do Porto, Neuroradiology Department, Porto, Portugal / Hospital Central do Funchal Dr. Nélio Mendonça, Neuroradiology Department, Funchal, Portugal

José Sá Silva, Vasco Abreu, José R. Pereira, Viriato Alves, Bruno Moreira, João Xavier

All: Centro Hospitalar Universitário do Porto, Neuroradiology Department, Porto, Portugal

Introduction: Aneurysmal bone cyst (ABC) are benign expansile osteolytic bony neoplasms, composed of numerous blood-filled channels and cystic spaces. They are rare and mostly seen in the first two decades of life, of which about 20-30% affect the vertebrae. Diagnosis is based on typical histopathological findings, but imaging is sometimes pathognomonic when fluid-fluid levels are found. Selective arterial embolization (SAE) has the best cost-to-benefit ratio in the treatment of patients with vertebral ABC.
Methods: Clinical and radiological case description and discussion.
Results: A 16-year-old boy presented with progressive motor weakness in the lower limbs for 10 days, with gait abnormality. Neurological examination also revealed pyramidal signs, sensory ataxia and lumbar sensory level. CT scan demonstrated an expansile osteolytic bony lesion involving T1 and T2 right posterior elements and vertebral bodies, with a soft tissue component generating spinal and homolateral foraminal stenosis. MR imaging confirmed compression of the spinal cord and right T1 and T2 spinal nerves. The tumor had a heterogeneous appearance on both T1 and T2-weighted MR, with focal areas of high T1 signal, presumably representing blood. There were multiple internal septations with enhancement and fluid-fluid levels, compatible with ABC. CT-guided percutaneous biopsy (CT-PB) was executed by a neuroradiologist, with the anatomopathological study establishing the definitive diagnosis of ABC. Three SAE were performed, with an interval of at least one month between them, with tumor devascularization by injection of particles, combined with corticosteroid therapy. Soon after the first SAE there was an improvement in neurological deficits, with progressive recovery of motor strength in the lower limbs. Control MRIs demonstrated a lesion volume decrease.
Conclusion: The important role of the neuroradiologist in this pathology is highlighted, both in the diagnosis, interpreting CT and MR images and executing the CT-PB, as well as in the treatment, performing the SAE.

ID55 | DISTINCT VOLUMETRIC SIGNATURE ALONG THE LONGITUDINAL AXIS OF THE HIPPOCAMPUS IN THE ALZHEIMER'S DISEASE CONTINUUM DEFINED BY CEREBROSPINAL FLUID BIOMARKERS PROFILE

Torcato Meira - torcatomeira14@gmail.com

1 Neuroradiology Department of Hospital de Braga, Rua das Comunidades Lusíadas 133, Braga, Portugal
2 School of Medicine of University of Minho, Campus de Gualtar, Braga, Portugal

Rafaela Morais-Ribeiro[2], Tiago Jesus[2], Marcelo Dias[2], Ana Coelho[2], Tiago Gil Oliveira[1,2]

1 Neuroradiology Department of Hospital de Braga, Rua das Comunidades Lusíadas 133, Braga, Portugal
2 School of Medicine of University of Minho, Campus de Gualtar, Braga, Portugal

Introduction: Cerebrospinal fluid (CSF) biomarkers have been increasingly used to support diagnosis of Alzheimer's disease (AD). Suspected non-AD pathophysiology (SNAP) refers to normal CSF levels of amyloid-β (Aβ) with increased tau, whereas Alzheimer's Disease continuum (ADc) is defined by Aβ pathology evidence. Since hippocampus studies have highlighted differential properties along its longitudinal axis, we aim to evaluate how its various subregional volumetric markers differ between ADc, SNAP and controls, as well as their association with clinical presentation.

Methods: We included 1242 participants from the Alzheimer's Disease Neuroimaging Initiative. Controls (n=234) were defined as having normal CSF Aβ (≥192 pg/ml), total tau (<93 pg/ml) and phosphorylated tau (<23 pg/ml). ADc individuals (n=784) were abnormal for Aβ, whereas SNAP subjects (n=224) had normal Aβ with either abnormal total or phosphorylated tau. Structural MRI acquisitions were analyzed with a method developed in our laboratory for segmenting the hippocampus in anterior, intermediate and posterior parts. Controlling for age and sex, groups were compared with one-way ANOVA (Tukey's post-hoc) for various hippocampal volumetric measures. Pearson coefficients were calculated to assess correlations between those volume variables and age, CSF biomarkers or neuropsychological scores – multiple comparisons adjustment with Benjamini-Hochberg (FDR<5%).

Results: ADc showed lower total and subregional hippocampal volumes than SNAP and controls (P<0.001). When normalizing the volume of each subregion for its total ipsilateral hippocampus volume, ADc showed increased posterior atrophy (P<0.001) and higher relative anterior volume (P<0.01) when compared with the other groups. Concisely, SNAP and controls (versus ADc) showed greater correlations between volumetric measures and age, with attenuated differences over age. Several subregional hippocampal volumes correlated significantly with altered levels of CSF biomarkers, cognition, neuropsychiatric features and with functional impairment (more strongly in ADc).

Conclusions: Our work contributes to better understanding of ADc and SNAP pathophysiology and to predicting their respective clinical features.

ID31 | ANTE-MORTEM MAGNETIC RESONANCE IMAGING GREY-WHITE MATTER CONTRAST REGIONAL SIGNATURES OF ALZHEIMER'S DISEASE NEUROPATHOLOGY

Francisco Almeida - franciscojcga@gmail.com

Life and Health Sciences Research Institute (ICVS), School of Medicine, University of Minho, Braga, Portugal
ICVS/3B's—PT Government Associate Laboratory, Braga/Guimarães, Portugal; Department of Neuroradiology, Centro Hospitalar Universitário do Porto, Porto, Portugal

Ana Coelho[1,2], Miguel Quintas-Neves[1,2,4], Merilee A. Teylan[5], Charles N. Mock[5], Walter A. Kukull[5], John F. Crary[6], Tiago Gil Oliveira[1,2,4]

1 Life and Health Sciences Research Institute (ICVS), School of Medicine, University of Minho, Braga, Portugal

2 ICVS/3B's—PT Government Associate Laboratory, Braga/Guimarães, Portugal

3 Department of Neuroradiology, Centro Hospitalar Universitário do Porto, Porto, Portugal

4 Department of Neuroradiology, Hospital de Braga, Braga, Portugal

5 Department of Epidemiology, National Alzheimer's Coordinating Center, University of Washington, Seattle, Washington

6 Neuropathology Brain Bank & Research Core, Department of Pathology, Nash Family Department of Neuroscience, Department of Artificial Intelligence & Human Health, Friedman Brain Institute, Ronald M. Loeb Center for Alzheimer's Disease, Icahn School of Medicine at Mount Sinai, New York

Introduction: Grey matter (GM), white matter (WM) contrast (GWC) in T1-weighted MRI has been shown to decrease with age in healthy populations and cognitive impairment in clinical Alzheimer's Disease (AD). However, the neuropathological determinants of this signal remain unknown. Here, we aimed to study whether GWC is associated with AD neuropathology and longitudinal hippocampal atrophy.

Methods: 157 patients with neuropathological, clinical data and ante-mortem MRI were selected from the National Alzheimer's Coordinating Center database. Cortical volume and GWC (WM/GM intensity) in T1/MRI were obtained with Freesurfer 6.0. Hippocampal volume was divided into anterior, intermediate and posterior regions using a lab-based algorithm. Correlational analysis was conducted for GWC with age, and GWC residuals after linear regression with age for volume, BRAAK stage (neurofibrillary tangles), CERAD score (neuritic plaque density) and Cognitive Dementia Rating Sum-of-Boxes (CDR-SB). APOE genotypes were compared using one-way-ANOVA.

Results: GWC was negatively correlated with age in widespread brain areas ($p < 0.05$, FDR), but only the entorhinal cortex showed a positive correlation between volume and GWC. There were no significant correlations between GWC and BRAAK stage, CERAD score, CDR-SB or differences according to APOE genotype. Entorhinal GWC correlated positively only with the posterior hippocampal volume in BRAAK III-IV ($p = 0.031$; $R = 0.34$).

Conclusion: GWC is highly modulated by age, but not by classical AD neuropathology, APOE genotype or dementia severity in this cohort. Entorhinal GWC correlates with entorhinal and posterior hippocampal volume, suggesting it might reflect an early pathophysiological process distinct from classical AD neuropathology.

ID50 | MR FINDINGS IN THE FOLLOW-UP OF TRANSORAL LASER MICROSURGERY FOR LARYNGEAL CANCER

Bruno Cunha - brunojorgecunha@gmail.com
Neuroradiology Department, Centro Hospitalar Universitário de Lisboa Central, Rua José António Serrano 1150-199 Lisboa, Portugal
Marco Ravanelli, Roberto Maroldi, Davide Farina
Radiology Department, University of Brescia, 25123 Brescia, Italy

Introduction: Transoral laser microsurgery (TLM) has demonstrated to be an effective therapeutic option for selected laryngeal tumors, offering high rates of tumor control and organ preservation. Imaging, in particular MR, plays a crucial part in the follow-up the post-TLM larynx. This study explores the various post-TLM findings that the radiologist may encounter with state-of-the-art MR (with surface coils) of the larynx.
Methods: We provide a pictorial review of laryngeal post-TLM findings, focusing on its characteristic MR findings and differential diagnosis.
Results: TLM for laryngeal cancer does not grossly change the anatomy of the larynx. Expected findings consist of visualization of volume loss and scar tissue within the surgical area, retracing the different types of cordectomies and partial laryngectomies. Compensatory adduction and edema of the contralateral vocal cord may be present. TLM complications include synechiae and granuloma formation and, more rarely, chondritis. If the patient was submitted to radiotherapy, chondroradionecrosis may ensue. Tumor recurrences can be focal submucosal or show extensive spread with invasion of adjacent structures. Careful evaluation of signal characteristics on unenhanced T1-WI, T2-WI and diffusion weighed imaging and of the contrast enhancing pattern frequently allows to discriminate tumor recurrence from scar tissue and inflammation.
Conclusion: MR with surface coils is a valuable tool for the evaluation of the post-TLM larynx, frequently allowing to differentiate expected findings and post-TLM complications from tumor recurrence.

ID35 | ENDOVASCULAR TREATMENT OF BRAIN ARTERIOVENOUS MALFORMATIONS IN PEDIATRIC PATIENTS: A SINGLE CENTER EXPERIENCE

Gonçalo Almeida - goncalo.almeida@campus.ul.pt

Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

Jaime Pamplona, Mariana Baptista, Rui Carvalho, Carla Conceição, Rita Lopes da Silva, Amets Sagarribay, Isabel Fragata, João Reis

1 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

2 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

3 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

4 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

5 Department of Pediatric Neurology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

6 Department of Neurosurgery, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

7 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

8 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

Introduction: Brain arteriovenous malformations (bAVMs) are abnormal vascular connections with direct arteriovenous shunts, generally symptomatic in the adult life. However, a small number of bAVMs may manifest in pediatric patients, with higher bleeding risk and mortality rates when compared to adults. The purpose of this work is to review our experience with endovascular treatment of bAVMs in pediatric patients, considering both efficacy and safety outcomes.

Material and Methods: Retrospective analysis of all bAVMs in pediatric patients (0-18 years) who underwent digital subtraction angiography (DSA) at our institution from January 2010 to June 2022.

Results: Twenty-six patients met the inclusion criteria, of which 12 underwent endovascular treatment. Treated patients had a mean age of 10,25 years and 58% were female: seven (58%) were treated in a single session and the remaining in multiple sessions (range 1-5 per patient). Complete angiographic exclusion was achieved in 5 (42%) patients with endovascular treatment. Five patients with residual bAVM after embolization needed adjuvant therapy with surgery (n=3) or stereotactic radiosurgery (SRS) (n=2). Two patients are still undergoing embolization sessions. Procedure-related complications occurred in two patients (17%) and included small vessel perforation and an occipital ischemic lesion. Two patients showed bAVM recurrence on follow-up (17%) and subsequently underwent SRS (n=1) or surgery (n=1), both resulting in complete bAVM exclusion. All patients were independent at follow-up (modified Rankin scale score 0-2).

Conclusions: Our experience supports the effectiveness and safety of endovascular treatment of bAVM in selected pediatric patients. A multidisciplinary approach combining surgery and SRS is warranted to achieve higher complete bAVM obliteration rates. Long term follow-up is important as these lesions undergo continuous vascular remodeling and may show recurrence over time, especially in the pediatric population.

ID7 | ACUTE CAROTID ARTERY STENTING VERSUS BALLOON ANGIOPLASTY FOR TREATMENT OF TANDEM OCCLUSIONS IN ACUTE ISCHEMIC STROKE –ANALYSIS OF A TERTIARY CENTRE

Inês Carneiro - inescarneiro93@gmail.com

Serviço de Imagiologia Neurológica, Centro Hospitalar Universitário Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

João Madureira, Manuel Alberto Correia, Gonçalo Basílio, Lia Lucas Neto, Paulo Sequeira, Luísa Biscoito

1 Serviço de Imagiologia Neurológica, Centro Hospitalar Universitário Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

2 Serviço de Imagiologia Neurológica, Centro Hospitalar Universitário Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

3 Serviço de Imagiologia Neurológica, Centro Hospitalar Universitário Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

4 Serviço de Imagiologia Neurológica, Centro Hospitalar Universitário Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

5 Serviço de Imagiologia Neurológica, Centro Hospitalar Universitário Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

6 Serviço de Imagiologia Neurológica, Centro Hospitalar Universitário Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

Introduction/Objectives: Despite endovascular thrombectomy (EVT) having become the standard of care for largevessel occlusion acute ischemic strokes (LVO-AIS), some uncertainty remains regarding the best acute endovascular treatment for LVO-AIS with concomitant extracranial internal carotid artery (eICA) steno-occlusive disease (tandem lesions). The aim of this study is to compare the clinical impact of eICA stenting versus performing balloon angioplasty (BA) alone in the setting of patients with tandem lesions submitted to EVT.

Methods: Single-centre, retrospective, observational study reviewing all patients undergoing EVT at our centre from January 2018 to October 2022, that had AIS due to anterior circulation LVO, steno-occlusive eICA disease, and were submitted to either eICA stenting or BA in the acute setting. Demographic, clinical, and procedure-related data was withdrawn. Primary outcomes were mortality rate (MR) and functional outcomes, defined according to the modified Rankin scale (mRS) in good (0-2) and poor (3-6).

Results: Thirty-seven patients were treated with eICA stenting and 17 patients with BA. Doppler ultrasonography showed re-occlusion of the eICA in 41%(N=7) of the angioplasty group and in 11%(N=4) of the stenting group. Restenosis of >70% occurred in 29%(N=5) of the angioplasty group and not in the stenting group. During in-hospital stay, two patients have died in the BA group due to malignant infarct and cerebral haemorrhage and five patients died in the stenting group, three due to cerebral hematomas after procedure. The stenting group showed better outcomes (mRS 0–2) compared to BA alone (p=0.02) and there was no difference in mortality between treatment groups (p=0.10).

Conclusion: Mortality did not occur more frequently after stenting than after BA of the cervical carotid lesion. Patients with tandem occlusions treated with emergent stenting of the eICA in conjunction to thrombectomy showed better mRS scores.

ID47 | THE CHALLENGE OF DIFFERENT CLINICAL AND IMAGIOLOGICAL PRESENTATIONS IN MOYAMOYA VASCULOPATHY

Henrique Coimbra Queirós - henriqueclq@gmail.com
Centro Hospitalar Universitário de Coimbra, Praceta Professor Mota Pinto, 3004-561 Coimbra, Portugal
Rui Duarte Armindo, Ana Carolina Chaves, Ricardo Veiga
1 Hospital Beatriz Ângelo, Avenida Carlos Teixeira, 3 2674-514 Loures, Portugal
2 Centro Hospitalar Universitário de Coimbra, Praceta Professor Mota Pinto, 3004-561 Coimbra, Portugal
3 Centro Hospitalar Universitário de Coimbra, Praceta Professor Mota Pinto, 3004-561 Coimbra, Portugal

Introduction: Moyamoya vasculopathy, which is divided into Moyamoya disease (MMD) and Moyamoya syndrome (MMS), is a progressive steno-occlusive process involving mainly the supraclinoid segments of the internal carotid arteries (ICA) and their proximal branches, with development of compensatory collaterals. MMS is secondary to an underlying disease process, whilst MMD is idiopathic.
Methods: Description of two pediatric-onset cases and brief literature review.
Results:
Case 1:
An 11-year-old female patient presented with sudden-onset headache, neck stiffness, vomiting and confusion. Additionally, she had low-grade fever. Head CT showed an extensive intraventricular hemorrhage, without subarachnoid hemorrhage or parenchymal hematomas. She was admitted for further investigation, including digital subtraction angiography (DSA), that revealed stenosis of supraclinoid ICA bilaterally, with occlusion of the proximal middle cerebral artery (MCA), and prominent collateral lenticulostriate and perisylvian branches. There was also a sub-occlusive stenosis of the right anterior cerebral artery (ACA). There were no evident signs of atherosclerosis, vasculitis, or fibromuscular dysplasia.
Case 2:
A 14-year-old female patient with previous history of headaches was submitted to a brain MRI to investigate a vascular cause of migraine. Because of the findings, a brain CT angiography was done and confirmed the presence of severe stenosis of both supraclinoid ICAs, with multiple tortuous collaterals in the region of the basal ganglia and leptomeningeal collaterals in the ACA territory from the patent posterior cerebral arteries. All these findings were posteriorly confirmed by DSA and a genetic panel was performed.
Conclusion: Although Moyamoya vasculopathy usually presents with ischemic vascular events, some patients may show different symptoms, such as headaches, vomiting or meningeal signs. It may be challenging to evaluate the different clinical and imagiological findings, especially in the pediatric population, but an early diagnosis is essential to the correct management of these patients.

ID18 | PEDIATRIC HYDROCEPHALUS: A RETROSPECTIVE ANALYSIS OF THE CLINICAL AND RADIOLOGICAL FEATURES IN CHILDREN

Carolina Chaves - anacbchaves@gmail.com

Unidade Funcional de Neurorradiologia, Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra

Pedro Barradas[1], Henrique Queirós[1], César Nunes[1], Sílvia Carvalho[1]

Unidade Funcional de Neurorradiologia, Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra

Introduction: Hydrocephalus is a disorder of CSF physiology resulting in abnormal expansion of cerebral ventricles. Advances in MRI and better understanding of the pathophysiology of hydrocephalus have organized the etiopathogenic concepts into obstructive (ventricular and extraventricular) and nonobstructive, as well as chronic and acute hydrocephalus. It is a common disorder in children.

Study design: In this study we performed an unicentric retrospective analysis of pediatric patients with hydrocephalus diagnosed on neuroimaging, between 4/2011 and 10/2022, from Hospital Pediátrico de Coimbra's neuroimage database.

Imagiological and clinical charts were reviewed to find patients' demographic, pathologic and neuroimaging features.

Results: We selected a large number of cases representative of the broad spectrum of pediatric hydrocephalus, which included a total of 114 pediatric patients. MRI was extensively used in diagnosis and signs of hydrocephaly were found in all of our cases, with more than a half having acute evolution. The mean age was approximately 8 years-old and 62% of patients were males. Intraventricular hemorrhage was the leading cause of pediatric hydrocephalus, representing 24% of cases. Tumors comprise a broad group of pathologies that can cause obstruction of any portion of the ventricular system, such as intraventricular, brainstem, pineal region, suprasellar and posterior fossa tumors, and caused 37% of hydrocephalus in our study. Aqueductal stenosis and congenital brain malformations, such as Chiari II malformation and Dandy-Walker malformation, represented 30% of the cases.

Conclusion: Hydrocephalus is not a single disease but a manifestation of many different ones, and although it is a common topic, developments in this field provided us with new insight. MRI can establish a diagnosis and differentiate obstructive from nonobstructive, as well as acute versus chronic or compensated hydrocephalus.

ID51 | PEDIATRIC ORBITAL VASCULAR ANOMALIES

Tiago Lorga - tiagomlorga@gmail.com
Serviço de Neurorradiologia do Centro Hospitalar Lisboa Ocidental, Rua da Junqueira 126, 1349-019 Lisboa, Portugal
Mariana Portela, Carolina Bruxelas, João Costa, Pedro Soares
1, 2, 3 Serviço de Oftalmologia do Centro Hospitalar Lisboa Ocidental, Rua da Junqueira 126, 1349-019 Lisboa, Portugal
4 Serviço de Neurorradiologia do Centro Hospitalar Lisboa Ocidental, Rua da Junqueira 126, 1349-019 Lisboa, Portugal

Introduction/Purpose: Pediatric orbital vascular anomalies (OVA) represent an important and diverse spectrum of lesions encountered within the orbit. They can present early in life, and some are unique to this particular age group. OVA may affect ocular motility and vision, as well as cause proptosis, pain and disfigurement.
Over the years, disagreement about the nomenclature and classification of vascular lesions arose. Nowadays, the ISSVA classification offers an updated system that classifies OVA as vascular malformations and vasoproliferative tumours.
Accurate diagnosis is essential for proper management, and imaging plays a crucial role in narrowing down a vast list of differential diagnoses.
This work aims to present pediatric OVA cases, review current literature, and describe distinctive imaging findings.
Methods: We describe several cases of pediatric OVA from our center and highlight the main imaging findings of each different entity.
Results: Hemangiomas are the most common vascular tumours in infancy and childhood. Infantile hemangioma (IF) (formerly capillary hemangioma) is the most common type, usually presents in the first eight weeks of life and shows a rapid proliferative stage ensued by a regressive, involuting phase. Cavernous venous malformation (wrongly termed cavernous hemangioma) is rare in the pediatric population and more frequent among adults.
Venous malformations type 2, previously known as "orbital varices", demonstrate distention on dependent position or Valsalva maneuver.
Venolymphatic malformations are the most frequent vascular malformation in children. They have a propensity to bleed and tend to enlarge during upper respiratory infections.
Although rare, congenital vascular anomalies, such as Sturge-Weber and Wyburn-Mason, can occur.
Conclusions: OVA represent a significant group of lesions in the pediatric population, in which imaging plays a crucial role in both diagnosis and management. Being acquainted with pediatric OVA typical imaging findings is paramount to narrowing down a vast list of differential diagnoses.

ID48 | NEUROIMAGING FINDINGS IN SICKLE CELL DISEASE

Francisca Sena Batista - franciscafsenabatista@gmail.com
Hospital Garcia de Orta, Avenida Torrado da Silva, Almada, Portugal
Diogo Marques, Filipa Castelão, Rosa Couto, Mariana Diogo, Lígia Neves, Cristina Rios
Hospital Garcia de Orta, Avenida Torrado da Silva, Almada, Portugal

Introduction: Sickle Cell Disease (SCD) is an autosomal recessive hemoglobinopathy caused by a point-mutation in B-Globin gene that leads to abnormal protein - Hemoglobin S (HbS) - highly prone to sickling in vessels. Multisystemic ischaemia (vaso-occlusive crisis) and haemolytic anaemia will develop. Brain, lungs, spleen, bones, kidney, liver, retina, heart and skin are the affected organs.
Stroke if the most common neurologic manifestation. Carriers of the mutation in homozygous form are at 250-fold increased risk of developing clinical or silent strokes.
We propose a pictorial neuroradiology review of classical and less known findings in SCD, with illustrative cases from our department, explanation of mechanisms and differential diagnosis.
Material & Methods and Results: SCD is a major cause of stroke in children. In addition to vasculopathy, the brain pro-coagulant and inflammatory state also contributes to increase incidence of neurologic manifestations. These include gradual decrease in cognitive and motor function (repeated silent strokes), acute focal neurological deficit (acute stroke), meningismus and headache (subaracnoid haemorrhage). Haemorrhagic stroke occur in 25% of cases, majority in adulthood. Skeletal manifestations of this entity are also relevant, associated with exacerbation of haematopoesis and replacement of yellow to red bone marrow, ischaemic insult to bones, and an increased risk of espondilitis/espondilodiscitis, osteoporosis and fractures.
Neuroimaging modalities, including CT, MRI, Transcranial Doppler US, and under certain indications and conditions, DSA, are crucial for diagnosis, management and follow up of this pathology. Contrast administration should be avoided, and if needed measures have to be undertaken.
Conclusion: Knowledge of the array of imaging findings in SCD is paramount for neuroradiologists, and these encompass not only brain and vascular imaging, but also the spine and head and neck.
We present an illustrative collection of cases to provide a structured approach to imaging and findings in these patients.

ID44 | UNILATERAL MICROBLEEDS IN PEDIATRIC POPULATION: A CLUE TO THINK IN SCLERODERMA

Mariana Ribeiro dos Santos - marianaribsantos@hotmail.com
Serviço de Neurorradiologia do Hospital de Braga, Braga, Portugal
Sofia Almeida Xavier, Vasco Sousa Abreu, Carla Conceição
1 Serviço de Neurorradiologia do Hospital de Braga, Braga, Portugal
2 Serviço de Neurorradiologia do Centro Hospitalar Universitário do Porto, Porto, Portugal
3 Serviço de Neurorradiologia do Hospital Dona Estefânia, Lisboa, Portugal

Introduction: Scleroderma is a rare pediatric disease characterized by collagen deposition and fibrosis of the skin and soft tissues. Central nervous system involvement in scleroderma has been increasingly recognized and brain MRI may reveal cerebral findings ipsilateral to the skin abnormality.
Clinical cases: A 10-year-old girl, from central America, was referred to the emergency department due to a 2-month progressive headache associated with nausea and photophobia. The patient also reported two previous suspected episodes of seizures with recovery after 5 minutes. On the physical examination, facial asymmetry was evident with right hemifacial atrophy. Neurological examination showed no abnormalities. Brain MRI revealed multiple microbleeds in the right cerebral hemisphere with ipsilateral subcortical white matter lesions, temporal leptomeningeal enhancement and effacement of parieto-occipital sulci. In the extracranial compartment, a right hemifacial atrophy was also notorious. Clinical presentation and MRI findings were compatible with Parry-Romberg syndrome. An 8year-old girl, caucasian, was referred for consultation in an outpatient setting to study the etiology of a previous stroke episode, which resulted in only slight face asymmetry. A small linear hyperpigmented cutaneous lesion was observed in the parasagittal left frontal region with associated alopecia. Brain MRI depicted a chronic left striatum-capsular infarction and additional ipsilateral cerebral multiple microbleeds and subcortical white matter lesions. Furthermore, it was confirmed a slight reduction on the left frontal parasagittal subcutaneous tissue thickness. The patient was positive for Scl-70 antibodies. Clinical presentation, laboratory and MRI findings were suggestive of linear scleroderma "en coup de sabre", with an associated stroke episode potentially related to vasculitis.
Conclusion: These cases depict a rare entity that should be remembered in the differential diagnosis of headache, seizures or focal neurological deficits, especially in pediatric female patients. Presence of multiple cerebral microbleeds ipsilateral to atrophic cutaneous lesions should prompt the inclusion of scleroderma in the differential diagnosis.

ID27 | UNDER PRESSURE: ABORDAGEM CLINICO-RADIOLÓGICA DA HIPERTENSÃO INTRACRANIANA IDIOPÁTICA EM IDADE PEDIÁTRICA

Liliana Igreja - lilianaigreja@gmail.com

Centro Hospitalar Universitário do Porto (CHPorto) - Largo do Prof. Abel Salazar, 4099-001, Porto, Portugal

João Tarrio[1], José Sá Silva[1], André Almeida[2], Joana Martins[3], Inês Carrilho[3], Sónia Figueiroa[3], José Eduardo Alves[1], Cristina Garrido[3]

1 Departamento de Neurorradiologia, Centro Hospitalar Universitário do Porto (CHPorto) - Largo do Prof. Abel Salazar, 4099-001, Porto, Portugal

2 Serviço de Pediatria e Neonatologia, Centro Hospitalar de Trás-os-Montes e Alto Douro (CHTMAD) Rua dos Lagoeiros 43, Vila Real, Portugal

3 Unidade de Neuropediatria, Departamento de Pediatria, Centro Materno Infantil do Norte (CMIN), Centro Hospitalar Universitário do Porto (CHPorto) - Largo da Maternidade de Júlio Dinis 45, 4050-651 Porto, Portugal

Introdução/Objetivos: A Hipertensão Intracraniana Idiopática (HII) é uma entidade rara em idade pediátrica e definida por pressão intracraniana elevada sem evidência clínica ou imagiológica de causa secundária, sendo a sua exata fisiopatologia pouco compreendida. O objetivo deste trabalho foi rever o estudo por neuroimagem dos doentes com diagnóstico de HII seguidos em consulta de neuropediatria.

Material e Métodos: Foi realizada uma análise retrospetiva de doentes com 10% no seguimento. 17% apresentaram diminuição de acuidade visual sequelar e 25% recidivaram da sintomatologia após suspensão do tratamento.

Conclusões: Os sinais imagiológicos indiretos de HII revelaram elevada especificidade diagnóstica que, quando combinados com os achados clínicos e oftalmológicos, podem permitir uma identificação eficaz destes doentes.

ID41 | THIS SHOULDN'T BE HERE! – A PICTORIAL REVIEW OF FOREIGN BODIES IN NEURORADIOLOGY

Gonçalo Gama Lobo - goncalogamalobo@gmail.com
Neuroradiology Department, Hospital de São José, Centro Hospitalar Lisboa Central, Lisbon, Portugal
Sara Rosa, Pedro Brandão, Isabel Fragata
Neuroradiology Department, Hospital de São José, Centro Hospitalar Lisboa Central, Lisbon, Portugal

Introduction: A foreign body represents a non-physiological material found in the body introduced from the outside. Foreign bodies are frequently retained in traumatic wounds and lacerations (7-15%), occurring mainly at the extremities, with the head and neck following, and the hand and wrist as the most frequent locations. Intracranial and intraspinal foreign bodies usually arise as a complication of invasive procedures. The imaging characteristics and material density/signal together with a thorough clinical history can lead to the correct diagnosis and help planning the best approach.
Material and methods: We retrospectively collected cases of foreign bodies in neuroimaging studies and provide a pictorial review depicting key clinical and imaging distinctive findings as well as the main complications.
Results: In our series, we describe several cases of foreign bodies in neuroimaging studies, including a variety of different materials, resulting from different mechanisms. Our cases comprise the principal foreign body materials such as air, glass, metal, plastic, wood and stone, as well as the principal routes of entry – trauma and invasive procedures. We also present the main complications associated with the presence of these non-physiological materials.
Conclusions: A wide variety of foreign bodies, from different materials and resulting from different mechanisms, can appear in neuroimaging studies. Up to 38% are missed at the initial physical evaluation. Therefore, the neuroradiologist plays a crucial role in identifying the material, the involved structures and possible complications. Knowing the main imaging features as well as potential complications, combining them with a clinical history, can guide the neuroradiologist to a correct diagnosis and help clinicians plan their approach.

Posters

ID1 | LAMINATION, SULCATION AND GYRATION: THE TIMELINE OF CORTEX DEVELOPMENT IN UTERO

Francisca Sena Batista - franciscafsenabatista@gmail.com
Hospital Garcia de Orta, Avenida Torrado da Silva, Almada, Portugal
Diogo Marques, Filipa Castelão, Rosa Couto, Mariana Diogo, Lígia Neves, Cristina Rios
Hospital Garcia de Orta, Avenida Torrado da Silva, Almada, Portugal

Introduction: Cerebral cortex development, sulcation and gyration are complex processes that occur in intrauterine life, particularly in second and third trimester and go on after birth. These processes are so precise in time, that correspond to milestones in CNS development and can predict gestational age, and can be visualised using fetal MRI.
Materials/Methods & Results: The primitive cerebral hemispheres are first formed at 5 GW (gestational week) while layering of cortex begins at 7 GW with waves of neuroblasts migrating to subpial position, long before current MRI capabilities can be used. By 16 GW, 7 layers can be identified, and will mature and change thickness and signal until birth although the brain surface remais smooth.
Gyri and sulci develop in response to arrival of sequential layers of cells for cortex formation. Folding will increased the surface area and accommodate an increase number of cells.
During second half of pregnancy, primary, secondary and tertiary sulcation confer the brain its complex anatomy, and are an integral part of evaluation of normal and pathological brain development.
In this paper we will present through MRI images the main milestones of fetal brain development of lamination, sulcation and gyration, using the best planes and sequences, in order to provide an illustrative atlas of paramount fetal brain development.
Conclusions: Fetal brain evolves through massive changes in structure and morphology, particularly in the second and third trimesters, when MRI is a valuable tool in CNS evaluation.
Knowledge of normal patterns is essencial for paediatric neuroradiologists in order to identify accurate gestational age, variations of the normal and recognise early neurodevelopment abnormalities.
We propose a review of the timeline of normal cerebral cortex development and patterns of sulcation and gyration with MRI imaging correlation.

ID2 | MORE THAN AN ACUTE OTITIS: A PICTORIAL REVIEW OF ACUTE MASTOIDITIS IN CHILDREN AND ITS COMPLICATIONS

Joana Castro Freitas - joanaccfreitas@gmail.com
Centro Hospitalar Universitário Lisboa Norte, Av. Prof. Egas Moniz MB, 1649-028 Lisboa
Hugo Loureiro Cadilha, Maria Inês de Sá, Carla Guerreiro
Centro Hospitalar Universitário Lisboa Norte, Av. Prof. Egas Moniz MB, 1649-028 Lisboa

Objectives: Review the imagological stages of acute mastoiditis and its complications, describing its imaging findings on computed tomography (CT) and magnetic resonance (MR) imaging.
Background: Acute otitis media (AOM) is an inflammation of the middle ear and a common disease in the pediatric population, usually with a benign course. However, occasionally, when an infection persists for more than a few days, it can spread to nearby structures, producing a suppurative infection of the mastoid air cells, acute mastoiditis. Mastoiditis is the most common complication of AOM, which can be effectively managed with antibiotics; however, incomplete treatment can potentially result in worsening infection with contiguous and hematogenous spread leading to severe complications. Radiographic findings of mastoiditis range from subtle changes of mastoid air cell opacification to more profound osseous destruction, with coalescent mastoiditis and complicating intracranial and/or extracranial subperiosteal abscesses. In severe cases, there may be intracranial complications, including epidural empyema, subdural collection, meningitis, cerebritis and dural venous sinus thrombosis. Usually, AOM is a clinical diagnosis and imaging studies are not recommended, unless a complication is suspected - especially in the pediatric population, where symptoms may be vague and non-specific. Beyond that, it has a key role to guide treatment and management of complications. CT should be performed to classify the mastoiditis as incipient or coalescent and to detect intracranial complications, with MR being particularly useful when intracranial complications are suspected. Methods: A pictorial review of complications of mastoiditis was performed, illustrating them with imaging examples from the Neurological Imaging Department of Centro Hospitalar Universitário Lisboa Norte.
Results: We illustrate with CT and MR images the most common complications of mastoiditis.
Conclusions: Acute mastoiditis may cause intratemporal and intracranial complications and it is crucial that radiologists accurately identify them, as early detection can help prevent serious sequelae.

ID3 | INNER EAR HEMORRHAGE – ANALYSIS OF TWO CLINICAL CASES

Inês Carneiro - inescarneiro93@gmail.com

Serviço de Imagiologia Neurológica do Centro Hospitalar e Universitário de Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

David Berhanu, Daniela Serras, Rita Sousa

1 Serviço de Imagiologia Neurológica do Centro Hospitalar e Universitário de Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

2 Serviço de Otorrinolaringologia, Hospital Dr. Nélio Mendonça, Avenida Luís de Camões 57 9004-514 Funchal, Portugal

3 Serviço de Imagiologia Neurológica do Centro Hospitalar e Universitário de Lisboa Norte, Avenida Professor Egas Moniz 1649-035 Lisboa, Portugal

Introduction: Inner ear haemorrhage is a rare cause of sudden sensorineural hearing loss (SSNHL). It may be underdiagnosed due to its rarity and difficulty to diagnose without magnetic resonance imaging (MRI). Conventional treatment is not very effective.

Methods: We report two cases of patients that were diagnosed with SSNHL in our centre and that displayed inner ear haemorrhage and review the clinical and neuroimaging aspects.

Results:

Case 1- A 43 year-old male presented with transversal right petromastoid fracture with cerebrospinal fluid otorrhea. He had a right positive head-impulse test (HIT), the audiogram revealed a profound right sensorineural hearing loss, and he was proposed to cochlear implantation. The diagnosis of intracochlear haemorrhage was made with MRI, which showed increased signal intensity on T1-weighted (T1W) and T2 fluid-attenuated inversion recovery (FLAIR) images in the right cochlea. No other abnormalities were found, in particular no enhancement after gadolinium. There was a failure to position the implant and the patient has not recovered from the hearing loss.

Case 2- A 63 year-old woman presented with sudden onset of dizziness, vomits, imbalance and tinnitus. She displayed left gaze horizontal rotary nystagmus with left rapid phase, without any other neurologic abnormalities. Peripheral vertigo was assumed, and she underwent brain and ear MRI which showed T1W and FLAIR hyperintensity in the right vestibule, semi-circular canals and, to a lesser extent, the cochlea, with lower signal on T2-weighted images, suggestive of right labyrinthitis with intra-labyrinth haemorrhage. During in-stay she developed right-ear hearing loss.

On the evaluation, she had a right positive HIT and the audiogram showed sensorineural hearing loss in the right ear.

She underwent vestibular rehabilitation with progressing improvements.

Conclusion: Intralabirynthine haemorrhage is a rare cause of SSNHL, may be present in trauma or inflammatory conditions and can be reliably identified with MRI.

ID4 | ENLARGED VESTIBULAR AQUEDUCT – ESSENTIALS

Hugo Loureiro Cadilha - hcadilha@gmail.com
Centro Hospitalar Universitário Lisboa Norte, 1649-035, Portugal
Joana Freitas, Maria Inês de Sá, Rita Sousa
Centro Hospitalar Universitário Lisboa Norte, 1649-035, Portugal

Enlarged vestibular aqueduct (EVA) is the most common radiologic malformation of the inner ear associated with sensorineural hearing loss (SNHL). The abnormality is often bilateral, may be asymmetric, and is slightly more common in females. EVA is associated with non-syndromic and syndromic forms of sensorineural hearing loss. In more than 80% of cases, an enlarged vestibular aqueduct is accompanied by other inner ear anomalies, such as cochlear dysplasia, modiolar deficiency, vestibular enlargement, or semicircular canal dysplasia. Those affected typically experience a sudden onset of hearing loss that fluctuates in severity and that may be aggravated by minor head trauma, barometric pressure changes, or activity that causes increased intracranial pressure. We describe two recently diagnosed pediatric cases of EVA from our center, displaying sudden SNHL post minor trauma, and a semicircular canal anomaly in association with EVA. We leverage these to display standard EVA measurement guidelines in the axial plane according to Cincinnati criteria (>1mm mid-point and >2mm opercular width), and in the Pöschl plane (>0,9mm mid-point width), in CT and MR scans. Early identification of EVA may warrant close audiological monitoring for cochlear implant candidacy and associated inner ear anomalies may influence electrode selection and surgical planning.

ID5 | RETINOBLASTOMA DIAGNOSIS IN A TRAUMATIC SETTING: THE IMPORTANCE OF DWI/ADC IMAGING

Vasco Sousa Abreu - vrlsa1993@gmail.com
Department of Neuroradiology, Centro Hospitalar Universitário do Porto, Portugal
Eduarda Pinto[1], José Silva[1], André Ferreira[2], Sofia Maia[2], Catarina Mendes Pinto[1]
1 Department of Neuroradiology, Centro Hospitalar Universitário do Porto, Portugal
2 Department of Ophthalmology, Centro Hospitalar Universitário do Porto, Portugal

Introduction: Retinoblastoma (RB) is the most common intraocular neoplasm of the childhood, commonly sporadic, but may be inherited due to a germline mutation of the tumoral suppressor RB1 gene. MRI is fundamental for diagnosis, staging and treatment planning.
Aim: To review RB imaging findings throughout a histological-proven case with an atypical clinic-radiological presentation.
Clinical case: A 3-year-old boy presented to the emergency department due to a blunt trauma to the right eye 2 weeks before. Physical examination was difficult to perform due to major eyelid swelling. Ultrasound evaluation revealed posterior lens dislocation while fundoscopy was impossible to perform. During hospitalization, he developed preseptal cellulitis of the same eye.
Cross-sectional imaging showed an enlarged and exophthalmic right eye. Its posterior segment was heterogenous, with a non-rhegmatogenous retinal detachment and associated subretinal space-occupying lesion. The later was characterized by low-T2 signal, restricted diffusion, and calcified component, but no appreciable contrast enhancement. Nevertheless, there was enhancement of the optic nerve head (also with restricted diffusion), of the periocular fat and of the uvea-scleral contours anteriorly.
Immediately, RB hypothesis was considered, although doubtful, with the possibility of intraocular traumatic/inflammatory lesional changes alternatively been raised. Exploratory vitrectomy confirmed a subretinal mass, whose histological analysis revealed an RB (staging: group E). Enucleation was subsequently performed, with actual tumoral-free 1-year follow-up.
Discussion: Main RB differential usually includes other pediatric causes of leukocoria, such as persistent hyperplastic primary vitreous or exudative retinitis. However, our main challenge was the differential with traumatic/ inflammatory changes, which masked the final diagnosis. In fact, lack of valuable contrast enhancement of the intraocular lesion may jeopardize the diagnosis. Thus, we emphasize the importance of DWI/ADC imaging, once RBs, due to their high cellularity, are characterized by restricted diffusion, which may be a clue in these rare cases with no/poor solid contrast enhancement.

ID6 | MULTIFOCAL MEDULLOBLASTOMA IN FANCONI ANEMIA

José Manuel Sá Silva - jose.msas.silva@gmail.com
Centro Hospitalar Universitário do Porto, Porto, Portugal
João Tarrio, Liliana Igreja, José Eduardo Alves
1, 2, 3 Centro Hospitalar Universitário do Porto, Porto, Portugal

We present the case of a two-year-old female with known history of Fanconi Anemia (FA) with a PALB2 germline biallelic pathogenic variant; in the context of her disease she had anemia, radial ray anomalies, unilateral renal agenesis and skin hyperpigmented spots.
She was brought to the ER for a two-week-long history of progressive irritability, vomiting and ataxia. MR imaging showed a large well-demarcated expansile lesion in the cerebellar vermis, with intermediate T2 and low T1 signal, restricted diffusion and mild contrast enhancement; it had significant mass effect with effacement of the fourth ventricle and basal cisterns, and obstructive hydrocephalus. In addition, three other smaller lesions with similar characteristics were found in the cerebellar hemispheres. There were no spine lesions.
Partial surgical excision of the largest vermian lesion was accomplished. Histologic and genetic testing confirmed the diagnosis of medulloblastoma, SHH-activated TP53-wildtype with nodular desmoplastic histology.
Although chemotherapy was started, the residual tumor and the smaller lesions still grew. She deceased after four months with respiratory failure in the context of severe pneumonia.
FA is characterized by predisposition to malignancies. Medulloblastoma has been described in cases of FA with biallelic inactivation in the tumor suppressor gene BRCA2/FANCD1 or its associated gene PALB2/FANCN; typically, these medulloblastomas are SHH-activated.
We hypothesize that, in our patient, the multiple cerebellar lesions represented synchronous primary medulloblastomas rather than metastatic dissemination, because the smaller lesions exhibited intraparenchymal location without other more typical leptomeningeal metastases. Intraparenchymal metastases are considerably rare in medulloblastoma and multifocal medulloblastoma in FA has been previously described.
Further proof could be obtained if the smaller lesions had been histologically tested - distinct gene expression patterns could suggest different synchronous tumors, considering that medulloblastoma typically maintains molecular subgroup affiliation in metastatic lesions.

ID7 | THE 2021 WORLD HEALTH ORGANIZATION CLASSIFICATION OF TUMORS OF THE CENTRAL NERVOUS SYSTEM: WHAT PEDIATRIC NEURORADIOLOGISTS SHOULD KNOW

Carolina Maia - carolinamaia.areal@gmail.com
Medical Image Department - Neuroradiology Functional Unit, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal
Pedro Barradas, Joana Pinto, Sílvia Carvalho, Teresa Garcia, Rui Pais
Medical Image Department - Neuroradiology Functional Unit, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

The fifth edition of the World Health Organization Classification of Tumors of the Central Nervous System (WHO CNS5) published in 2021 builds on the 2016 edition and includes output from the Consortium to Inform Molecular and Practical Approaches to CNS Tumor Taxonomy (cIMPACT-NOW).
WHO CNS5 presents fundamental changes to brain tumor classification, especially in the pediatric population, through the introduction of newly recognizing entities, elimination and renaming of multiple specific tumor types, and by adjusting the taxonomic structure. Indeed, the updates to the pediatric classification are protean, ranging from the introduction of new types to establishing separate tumor families for pediatric-type gliomas. Thus, pediatric neuroradiologists must familiarize themselves with the updated WHO Classification of CNS neoplasms to be informed and effective at multidisciplinary tumor boards and in interactions with colleagues in neuro-oncology, neurosurgery, radiation oncology and neuropathology.
We aimed to present, summarize, and illustrate with cases from our Centre the most salient aspects of the new 5th edition in the pediatric population.

ID8 | SCHILDER'S DISEASE – WHERE DO WE STAND?

Tiago Lorga - tiagomlorga@gmail.com
Serviço de Neurorradiologia, Centro Hospitalar Lisboa Ocidental, Rua da Junqueira 126, 1349-019 Lisboa, Portugal
Marta Magriço, Manuel Salavisa, Pedro Soares
1 Serviço de Neurologia, Centro Hospitalar Lisboa Ocidental, Rua da Junqueira 126, 1349-019 Lisboa, Portugal
2 Serviço de Neurologia, Centro Hospitalar Lisboa Ocidental, Rua da Junqueira 126, 1349-019 Lisboa, Portugal
3 Serviço de Neurorradiologia, Centro Hospitalar Lisboa Ocidental, Rua da Junqueira 126, 1349-019 Lisboa, Portugal

Introduction/Purpose: Schilder's disease (SD), also known as myelinoclastic diffuse sclerosis, is considered to be a rare and aggressive variant of multiple sclerosis (MS) that predominantly affects children. SD is characterized by focal neurological deficits, which is an atypical clinical presentation for MS, and by a monophasic disease course.
Due to its particularly low prevalence, there are few studies about SD. It is debatable whether SD is a variant of MS or a pathological entity in its own right. Oligoclonal bands (OCBs) are only present in up to 20% of cases.
The presence of two large roughly symmetrical bilateral lesions in the centrum semiovale is the hallmark imaging finding of SD.
The mainstay of treatment of SD is high-dose corticosteroids, although there are some reports regarding the usage of disease modifying treatment options for MS.
This work aims to present an usual case of a teenager with SD, to describe typical imaging findings, and to review the literature of this rare condition.
Methods: We describe the case of a 17-year-old boy referred to the Neurology Clinic to investigate sudden-onset sensitive and motor deficits in the limbs.
Results: The patient revealed lower limbs ataxia, motor deficits of the left hand and generalized hyperreflexia. Brain-CT showed two large bilateral hypodense lesions in the centrum semiovale. In MRI these lesions presented faint peripheral enhancement, little mass effect and rough symmetry, with T2/FLAIR-hypersignal. MRS revealed a decreased NAA peak. The patient was treated with high-dose corticosteroids. 6-month follow-up MRI showed lack of enhancement and slight reduction of the lesions.
Conclusions: SD is a rare disease that might be a distinct pathological entity. Its imaging hallmark of two large symmetrical lesions in the centrum semiovale plays a critical role in diagnosis. Further research is paramount to define this entity and to establish treatment guidelines.

ID9 | SICKLE CELL DISEASE: CEREBRAL AND SKELETAL MANIFESTATIONS

Inês Freire - ines.sf@live.com.pt

Neuroradiology Department - Hospital de Egas Moniz, Centro Hospitalar Lisboa Ocidental (CHLO), Rua da Junqueira 126, 1349-019, Lisbon, Portugal

João Gonçalves[2,3], Filipa Proença[4], Carla Conceição[2]

2 Neuroradiology Department - Hospital de São José, Centro Hospitalar Lisboa Central (CHLC), Lisbon, Portugal

3 Neuroradiology Department - Hospital Central do Funchal, Funchal, Portugal

4 Neuroradiology Department - Hospital de Santa Maria, Centro Hospitalar Lisboa Norte (CHLN), Lisbon, Portugal

Introduction and Purpose: Sickle cell disease (SCD) is an autosomal recessive hemoglobinopathy characterized by the presence of hemoglobin S, which leads to abnormally shaped (sickled) red blood cells. The main manifestations of this disease are related to hemolytic anemia and vaso-occlusive crises, the latter being clinically threatening.

The purpose of this work is to illustrate the findings that can be found in neuroimaging in patients with SCD.

Methods: The authors present several CT and MR images performed at CHLC with the cerebral and skeletal imaging findings most frequently found in patients diagnosed with SCD.

Results and Discussion: Sickle cell disease can have a wide variety of clinical presentations, usually manifesting in early childhood. Over their lifetime, approximately 25% of these patients will have a neurologic complication. Vasculopathy is the underlying cause of many radiological manifestations, including territorial infarction, "silent ischemia" and intracranial hemorrhage (intraparenchymal and subarachnoid hemorrhage). Territorial infarcts are mostly seen in the anterior circulation, and "silent ischemia" typically involves the "watershed" border zones. Occlusion or stenosis of the distal internal carotid artery may result in the development of moyamoya syndrome. There is a greater propensity for aneurysm formation in adults with SCD, which tend to be multiple and to appear in the posterior circulation. Less frequent complications include posterior reversible encephalopathy syndrome (PRES) and cerebral fat embolism secondary to the breakdown of infarcted bone.

Extramedullary hematopoiesis may be especially evident in the skull due to diploic space widening ("hair-on-end" appearance). Skeletal complications include infarction and osteomyelitis. Spontaneous epidural hemorrhage and subperiosteal hemorrhage can be seen with calvarial bone infarction, and microvascular endplate infarction may result in H-shaped vertebrae.

Conclusions: The neurological and skeletal complications of SCD are relatively common, and the neuroradiologist needs to be familiar with their neuroimaging presentation.

ID10 | NEUROTOXOPLASMOSIS: AN OVERLOOKED COMPLICATION OF BIOLOGIC TREATMENT FOR RHEUMATOID ARTHRITIS

Gonçalo Almeida - goncalo.almeida@campus.ul.pt
Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal
Miguel Cristóvão, Carlos Pontinha, Clara Ribeiro
1 Department of Pathology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal
2 Department of Pathology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal
3 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

Introduction: Toxoplasmosis is one of the most common opportunistic infections, mainly reported in patients with acquired immunodeficiency syndrome (AIDS). Patients with rheumatoid arthritis (RA) have been linked to reactivation of toxoplasmosis due to immunosuppressive treatment, although biologic drugs have seldom been implicated. The aim of this work is to present a case of cerebral toxoplasmosis in a patient with RA following initiation of adalimumab, a tumor necrosis factor α (TNF-α) inhibitor.
Material and Methods: Clinical, imaging and pathologic data were retrospectively obtained from the patient's case files.
Results: A 62-year-old female presented to our hospital due to worsening temporal disorientation and unsteady gait over a two-week period. She had been diagnosed with RA 10 years before and was on chronic treatment with oral prednisolone, leflunomide and methotrexate. Due to progressive joint pain, adalimumab was added to her regimen 2 months before admission. Blood tests revealed relative neutrophilia (8120 μL, 89%) and relative lymphopenia (600 μL, 7%). The patient was seronegative for human immunodeficiency virus type 1 (HIV-1) but had detectable serum IgG antibodies to toxoplasma gondii. A CT scan of the brain showed a space-occupying hypodense lesion in the right basal ganglia, surrounded by edema. On MRI the lesion showed heterogeneous signal intensity on the T2/FLAIR sequence, marginal enhancement following gadolinium injection and significant decrease of cerebral blood flow (CBV). A brain biopsy was performed and revealed not only cysts containing bradyzoites but also free tachyzoites across the sample, confirming the diagnosis of toxoplasmosis.
Conclusions: This case portrays reactivation of toxoplasmosis after initiation of adalimumab in a patient already on chronic treatment with other non-biologic immunosuppressive drugs. Since the use of biologic drugs is increasing, each case must be carefully evaluated prior to treatment. A high-index of suspicion in seropositive patients is warranted.

ID11 | INTRACRANIAL NEUROENTERIC CYST: CASE REPORT

Maria Inês de Sá - ines_fs_6@hotmail.com
Serviço de Imagiologia Neurológica, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Av. Professor Egas Moniz, 1648-028 Lisboa, Portugal
Guilherme Martins, Joana Freitas, Carlos Morgado, Carla Guerreiro
Serviço de Imagiologia Neurológica, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Av. Professor Egas Moniz, 1648-028 Lisboa, Portugal

Introduction: Neuroenteric (NE) cysts are rare benign endodermal lesions of the central nervous system that results from incomplete resorption of the neurenteric canal during the third week of embryogenesis.
The majority appear in the intradural extramedullary spinal cord, but they can also be found intracranially, most often located in the extra-axial space of posterior fossa, anterior to the brainstem or in the cerebellopontine angle.
Presentation: A 10-year-old girl with a four-year diagnosis of brainstem cavernoma with frequent episodes of headaches and vomiting underwent a follow-up magnetic resonance imaging (MRI) examination at our Hospital.
MRI revealed an extra-axial lobulated lesion located anteriorly to brainstem, with mostly high T1 and T2/FLAIR signal suggestive of high protein content, no restriction diffusion, no gadolinium enhancement, and foci of low signal on T1, T2 and T2*. On the perfusion study, rCBV was not increased. The lesion had no surrounding edema and had a moderate mass effect on the brainstem.
Conclusion: The lesion's lobulated morphology and its location are typical of a NE cyst. Signal intensity on MR sequences varies with the protein content, with most being T1 and T2 hyperintense. Also, intralesional hypointense foci on T2-weighted images have been suggested to be due to elevated protein concentration within the cyst, squamous metaplasia, and/or keratinous debris. Differential diagnosis includes arachnoid cyst, epidermoid cyst, Rathke cyst and colloid cysts. Epidermoids are often located along the midline and are T1 hyperintense, but typically show restriction diffusion. On the other hand, arachnoid cysts have the same signal intensity as CSF on all sequences. Rathke and colloid cysts can be distinguished from NE cysts based on location. Cavernomas are more likely supratentorial, intraaxial and demonstrate a "popcorn" appearance containing areas of bleeding at different stages and change in signal intensity due to aging of blood products.

ID12 | CHEMICAL MENINGITIS DUE TO RUPTURED EPIDERMOID CYST

Carolina Chaves - anacbchaves@gmail.com
Unidade Funcional de Neurorradiologia, Serviço de Imagem Médica, CHUC
Pedro Barradas[1], César Nunes[1], Joana Sousa[2], Ana Brett[2,3], Fernanda Rodrigues[2,3], Rui Pedro Pais[1]
1 Unidade Funcional de Neurorradiologia, Serviço de Imagem Médica, CHUC
2 Serviço de Urgência e Unidade de Infeciologia, Hospital Pediátrico, CHUC
3 Faculdade de Medicina, Universidade de Coimbra
4 Serviço de Pediatria Médica, Hospital Pediátrico, CHUC

Introduction: Aseptic meningitis (AM) is defined as a serous inflammation of the meninges. Rarely, AM can occur due to the rupture of an epidermoid cyst (EC) into the subarachnoid space, causing chemical irritation. ECs are rare benign intracranial tumours that account for approximately 1% of all intracranial tumours. Most are congenital, arising from epithelial cells that are retained during closure of the neutral tube.
Methods: We report a case of aseptic meningitis due to rupture of an epidermoid cyst.
Results: A 2-year-old female presented to the emergency department with a history of headache, fever, sleepiness and neck stiffness. Cerebrospinal fluid (CSF) showed elevated cell count (6302 cells/mm3), increased proteins (273 mg/dL), decreased glucose (15mg/dL), with negative PCR and cultures. The child was started on antibiotics for acute bacterial meningitis. Later, a brain magnetic resonance imaging (MRI) showed an intracranial extra-axial tumour, involving the right prepontine cistern, with isosignal to CSF in T1 and T2, restriction in diffusion-weighted imaging (DWI), and dimensions of approximately 35x22 mm in the axial and sagittal planes. Additionally, a small foci of restriction on DWI was found in the left occipital horn, which was perceived as purulent material or blood. The mass was compatible with an intracranial EC and was interpreted as an incidental finding. The child improved gradually. However, 3 weeks after discharge she returned to the emergency department with a similar clinical picture and CSF changes. Follow-up brain MRI showed a significant reduction of the EC, with decreased mass effect upon the brainstem, and chemical meningitis was considered, with good outcome.
Conclusion: Acute meningitis usually has an infectious aetiology. However, in recurrent cases without microorganism identification, other diagnosis should be considered. The follow-up MRI study, in combination with clinical findings, helped to diagnose chemical meningitis due to spontaneous rupture of the EC.

ID13 | THE PRODUCTS OF OUR IMAGINATION: A JOURNEY THROUGH THE PAEDIATRIC NEURORADIOLOGY SIGNS AND APPEARANCES

Carolina Chaves - anacbchaves@gmail.com

Unidade Funcional de Neurorradiologia, Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra

Pedro Barradas, Sílvia Reigada, Bruno Gomes, Cristina Moura, César Nunes

Unidade Funcional de Neurorradiologia, Serviço de Imagem Médica, Centro Hospitalar e Universitário de Coimbra

Introduction: A number of particular appearances in neuroimaging studies gave inspiration for the description of various classic signs and appearances allusive to common objects or patterns that we come across in our daily routine in life. The goal behind the association between such common patterns or objects and the corresponding pathologies is to make the learning process easier, by increase retention of information and enhance memory recall. Consequently, these signs are very popular among neuroradiologists.

Methods: We make a revision of various neuroimaging classic signs and appearances described in paediatric neuroradiology.

Results: In this essay, we describe various classic, important and frequently seen neuroradiological signs and appearances in children. We present these cases with suitable examples and illustrations, using mainly computed tomography (CT) and magnetic resonance (MR) images. Some signs can be quite specific while others are often indicative of a group of similar pathologies. Additionally, we aimed to review the causes for their particular appearance, significance and differential diagnosis when faced with them. Study of these classical signs would be extremely helpful in routine clinical practice, not only for neuroradiologists, but also for neuropediatricians, paediatric neurosurgeons and other relatable medical specialties.

Conclusion: The use of analogies in the description of imaging findings is a common practice in Neuroradiology. As a matter of fact, the use of signs is a process of conjuring up associations between imaging findings and themes or objects, and has the particular advantage of bounding memory with creativity, reinforcing information recall and making the process of learning easier and funnier. Therefore, neuroradiologists should be aware of the diversity of neuroimaging signs and use them as a tool.

ID14 | X-LINKED HEREDITARY PERIVENTRICULAR NODULAR HETEROTOPIA: A CASE REPORT

Guilherme Ribeiro Martins - guilhermermartins2@gmail.com

Serviço de Imagiologia Neurológica, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Lisboa, Portugal

Maria Inês de Sá[1], Teresa Loureiro[2], José Carlos Ferreira[2], Mariana Tomásio Neves[3], Oana Moldovan[3], Graça Sá[1]

1 Serviço de Imagiologia Neurológica, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Lisboa, Portugal

2 Unidade de Ecografia e Diagnóstico Pré-Natal, Departamento de Obstetrícia, Ginecologia e Medicina da Reprodução, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Lisboa, Portugal

3 Serviço de Genética Médica, Departamento de Pediatria, Hospital de Santa Maria, Centro Hospitalar Universitário Lisboa Norte, Lisboa, Portugal

Introduction: Periventricular nodular heterotopia (PNH) is a malformation of cortical development caused by impaired neuronal migration, resulting in abnormal periventricular nodules of ectopic grey matter that frequently lead to cognitive delay and refractory epilepsy. Filamin A gene (FLNA) is responsible for cell stability and motility. Its deficiency is inherited in an X-linked manner, with a phenotypic spectrum that includes FLNA-related PNH and also cardiovascular and pulmonary disease, among other abnormalities.

Case Description: We describe the case of a 28-year-old pregnant woman (G1P0) with history of asthma, mild obstructive ventilatory defect and left ventricular and aorta root dilatation. Her mother and sister have asthma and her father has epilepsy. At 21 weeks of gestation, she underwent a neurosonography, confirming the ultrasound findings of diffuse periventricular irregularities and mega cisterna magna. Fetal MRI was performed at 22+5 weeks and confirmed a diffuse nodular irregularity in the contour of the lateral ventricles, hypointense on T2-weighted images, suggestive of PNH. It was also evident the presence of mega cisterna magna and atrial and occipital horns enlargement. Brain MRI on the mother was performed and diffuse PNH and mega cisterna magna was found. She was referred to the Department of Genetics, and the diagnosis of X-linked hereditary PHN was proposed. Further evaluation was initiated, such as the arrayCGH analysis which was negative and the whole exome sequencing (WES) for which results are still pending.

Conclusion: We presented a case of X-linked hereditary PNH, yet to be confirmed, a disease with unknown prevalence due to its rarity. MRI imaging of the fetus allowed confirmation of the diagnosis of periventricular nodular heterotopies first found on ultrasound. The diagnosis of diffuse PNH and mega cisterna magna on the mother's brain MRI, made the suspicion of this disease more likely, ending with genetic studies to confirm.

ID15 | A PROTOTYPICAL ENTITY OF THE PEROXISOMAL DISEASE SPECTRUM

Denil Champac Tribovane - deniltribovane@hotmail.com
Hospital Beatriz Ângelo
Sandra Matias, Pedro Vilela
Hospital Beatriz Ângelo

Introduction: Disorders involving biogenesis of peroxisomes cause severe neurometabolic diseases inherited in an autossomal recessive manner. Cerebrohepatorenal or Zellweger syndrome (ZS), is a rare disorder of peroxisomal function that presents in the neonatal period with involvement of multiple organ systems. The prognosis is dismal and the average lifespan is of approximately 1 year.
Material and methods: We present two pre-term neonates with clinical, laboratory and imaging findings consistent with Zellweger Syndrome.
Results: Both patients (1 female and 1 male) were admitted to the neonatal intensive care unit after birth for generalized hypotonia and poor feeding. Clinical examination revealed dysmorphic facial features and laboratory results noted elevations in multiple very long chain fatty acids (VLCFA). MRI was performed in both patients and revealed reduced myelination, bilateral abnormal gyration in the perisylvian regions, and one patient had coexistent bilateral germinolytic cysts in the caudothalamic groove.
Discussion: The diseases of the Zellweger spectrum comprise group A of the peroxisomal biogenesis disorders, in which the peroxisomes appear abnormal and exhibit generalized loss of function, with ZS designated as the prototype disease in this family of disorders. Group B refers to diseases with normal-appearing peroxisomes but loss of multiple peroxisomal functions. Group C have normal-appearing peroxisomes with only a single loss of function.
ZS is characterized by an accumulation of VLCFA and presents with distinctive facial stigmata, hypotonia, seizures, poor feeding, hepatic dysfunction and bone abnormalities.
MRI findings are virtually pathognomonic, demonstrating delayed myelination and extensive areas of polymicrogyrialike changes in the perisylvian regions. Subependymal germinolytic cysts along the frontal horns of the lateral ventricles and incomplete opercularization are also occasionally present.
Early neurologic imaging findings of peroxisomal disorders may be subtle and MRI may provide critical information that prompts a more specific diagnostic approach to the patient.

ID16 | WHERE DO ARACHNOID CYSTS COME FROM? - TWO CASES OF FETAL SUBARACHNOID SPACE ENLARGEMENT PROGRESSING TO ARACHNOID CYSTS IN INFANCY

Sara Rosa - saradbrosa@gmail.com

Departamento de Neurorradiologia do Centro Hospitalar Universitário de Lisboa Central, R. José António Serrano 1150-199 Lisboa, Portugal

Gonçalo Gama Lobo, Alexandra Rodrigues, Ana Antunes Martins, Álvaro Cohen, Carla Conceição

1 Departamento de Neurorradiologia do Centro Hospitalar Universitário de Lisboa Central, R. José António Serrano 1150-199 Lisboa, Portugal

2 Departamento de Neurorradiologia do Centro Hospitalar Universitário de Lisboa Central, R. José António Serrano 1150-199 Lisboa, Portugal

3 Departamento de Neuropediatria do Centro Hospitalar Universitário de Lisboa Central, R. Jacinta Marto 8A Lisboa, Portugal

4 Centro de Responsabilidade Integrado de Medicina e Cirurgia Fetal do Centro Hospitalar Universitário de Lisboa Central, R. Viriato 1, 2890-495 Lisboa, Portugal

5 Departamento de Neurorradiologia do Centro Hospitalar Universitário de Lisboa Central, R. José António Serrano 1150-199 Lisboa, Portugal

Introduction: Primary arachnoid cysts are benign intra-arachnoidal accumulations of clear cerebrospinal fluid, that are most commonly found in the middle cranial fossa, anterior to the temporal lobes (50-60%). Although their etiology is thought to be congenital, most cases of arachnoid cysts diagnosed prenatally are found in different locations. This poses the question: do middle cranial fossa arachnoid cysts, the most frequent in the adult and pediatric population, arise in utero or form later in life through different mechanisms?

Methods: We present two cases of a child and an infant studied using fetal and postnatal MRIs. Fetal MRIs were performed at 23 and 27 weeks of gestation for the first case, and at 24, 28 and 32 weeks of gestation for the second one. Postnatal MRIs were performed at 5 months and 4 years of age for the first case, and at 4 and 12 months for the second case.

Results: Our patients presented with diffuse subarachnoid space enlargement diagnosed prenatally through fetal MRI, which postnatally evolved to middle cranial fossa arachnoid cysts and benign enlargement of the subarachnoid space in infancy. Additionally, one of the cases also presented with a mega cisterna magna and the other with a frontal arachnoid cyst.

Conclusion: To our knowledge, these are the first cases in the literature reporting the progression from diffuse subarachnoid space enlargement to arachnoid cyst development. Considering the latter, we propose that some middle cranial fossa arachnoid cysts might result from subarachnoid space enlargement during the fetal period, although further evidence is needed to support this hypothesis.

ID17 | EVERY CLOUD HAS A SILVER LINING: SPONTANEOUS DISAPPEARANCE OF ARACHNOID CYST AFTER HEAD TRAUMA

Ângelo Dias - angelondias@gmail.com

Department of Neurological Imaging, Hospital de Santa Maria, Centro Hospitalar Lisboa Norte, 1649-028 Lisboa, Portugal

Maria Inês de Sá, Joana Castro Freitas, Carlos Morgado

1 Department of Neurological Imaging, Hospital de Santa Maria, Centro Hospitalar Lisboa Norte, 1649-028 Lisboa, Portugal

2 Imaging University Clinic, Faculdade de Medicina da Universidade de Lisboa, 1649-028 Lisboa, Portugal

Introduction: Intracranial arachnoid cysts (AC) are common congenital cerebrospinal fluid filled sacs that are usually detected incidentally on brain imaging. They often remain stable in size and rarely involute or disappear over time. The mechanisms of the disappearance of the arachnoid cysts are yet not fully understood.

Objective: We present two cases of post-traumatic reduction of anterior temporal AC in children and adult and review current literature focusing on possible causes of AC reduction as well as discuss the underlying pathophysiological mechanisms.

Results: In most cases, AC resolve spontaneously but in rare cases it may be triggered by certain events. Among the triggering events that have been correlated with regression of AC head trauma and infection accounts for most cases. The exact mechanism underlying AC disappearance after a triggering event is not fully understood. The most likely mechanism relies on cyst wall rupture with the consequent communication between the cyst and normal cerebrospinal fluid pathways. The rupture may also occur into the subdural space as in one the cases presented here.

Conclusions: ACs disappearance is a rare phenomenon but must be kept in mind as it can occur spontaneously or after an inciting event like in these two cases of head trauma.

ID18 | CNS- IMMUNE RECONSTITUTION INFLAMMATORY SYNDROME: A CHALLENGING DIAGNOSIS

Sofia Bettencourt - sofiar@campus.ul.pt
Neuroradiology Department, Centro Hospitalar de Lisboa Central, 1150-199 Lisbon, Portugal
Pedro S. Brandão, Teresa Morais
Neuroradiology Department, Centro Hospitalar de Lisboa Central, 1150-199 Lisbon, Portugal

Introduction: Immune reconstitution inflammatory syndrome (IRIS) describes a paradoxical clinical deterioration following sudden improvement from an immunosuppressed condition, most commonly in HIV-infected patients. In those patients IRIS usually develops within weeks or months after the introduction of highly active antiretroviral therapy (HAART), representing a heightened immune response to an antigen. CNS-IRIS has been described in relation to several opportunistic pathogens, most frequently John Cunningham (JC) virus, Cryptococcus neoformans and Mycobacterium tuberculosis (TB), but other viruses, mycobacteria, and fungi have also been associated with this syndrome. It can affect 9-47% of HIV patients with a CNS opportunistic infection who start HAART and is usually selflimiting. However, it can be fulminating, with a mortality rate of 13-75%.
Despite being a diagnosis of exclusion, an history of an CNS-IRIS should be suspected in immunocompromised patients, with the onset of new or progressive clinical symptoms, regardless of medical treatment with improved immune laboratory findings.
Methods and Results: We retrospectively collected cases of CNS-IRIS in HIV-infected patients and reviewed their clinical data and imaging findings, depicting the key clinical and imaging findings that should raise the suspicion for CNS-IRIS.
Conclusion: Early recognition and intervention are essential to contain inflammatory response in these patients. The knowledge of the clinical background and the distinct MR imaging characteristics of this entity namely increased high FLAIR signal abnormalities, leptomeningeal and/or parenchymal lesions contrast enhancement and mass effect, which are not commonly found in the untreated HIV-infected patients, should raise the suspicion for CNS-IRIS.
Although this is a diagnosis of exclusion, often challenging and with limited treatment options, the neuroradiologist may play a critical role in alerting the clinician to this diagnosis.

ID19 | MEROSIN DEFICIENT CONGENITAL MUSCULAR DYSTROPHY: A CASE REPORT WITH MAGNETIC RESONANCE IMAGING FINDINGS

Sofia Bettencourt - sofiar@campus.ul.pt
Neuroradiology Department, Centro Hospitalar de Lisboa Central, 1150-199 Lisbon, Portugal
Pedro Brandão, Teresa Morais
Neuroradiology Department, Centro Hospitalar de Lisboa Central, 1150-199 Lisbon, Portugal

Introduction: Merosin-deficient congenital muscular dystrophy, also known as congenital muscular dystrophy type 1A (MDC1A), is a rare autosomal recessive condition caused by deficient merosin expression. It results from variants in the LAMA2 gene on 6p22-23 leading to partial or complete absence of laminin α2 chains that compose merosin. This structural protein of the basement membrane is expressed in the striated muscle, the Schwann cells, in the astrocyte foot processes at the glia limitans and blood-brain barrier. The deficiency of this protein leads to a loss of structural integrity and function of these cells, ultimately resulting in severe muscle weakness with onset within the first 6 months of life, accompanied by elevated serum creatinine kinase (CK) levels, and less frequently in seizures, intellectual disability, and neuropathy. Patients with less severe forms of the disease present later in childhood.
Methods and Results: We report a case of a 31-year-old woman with a personal history of neonatal hypotonia and muscle contractures, with elevated serum CK, and seizures since the age of 7. A muscle biopsy was performed revealing a merosin-deficient muscular distrophy and subsequent genetic test confirmed LAMA2 gene mutation. An inter-ictal brain MRI showed bilateral and symmetric occipital cortical thickening, with a slightly irregular inner surface and a paucity of sulcations cobblestone lissencephaly. Bilaterally symmetrical deep white matter T2 hyperintensity was also seen with associated lateral ventricles enlargement.
Conclusion: Imaging studies, particularly MRI, play an important role in the diagnosis of MDC1A patients. White matter signal abnormalities have been largely described in these patients, but rarely structural brain abnormalities. We describe a rare case of a patient with genetically proven MDC1A and cobblestone lissencephaly malformation, with emphasis on characteristic neuroimaging features.

ID20 | DIFFERENTIAL DIAGNOSIS OF PONTINE LESIONS WITH RESTRICTED DIFFUSION IN THE ADULT

Pedro Sousa Brandão - pedrodsbrandao@gmail.com
Department of Neuroradiology, Centro Hospitalar Universitário de Lisboa Central, E.P.E., Lisbon, Portugal
Sofia Bettencourt, Teresa Morais
Department of Neuroradiology, Centro Hospitalar Universitário de Lisboa Central, E.P.E., Lisbon, Portugal

Introduction: Magnetic resonance (MR) is the most appropriate imaging modality for evaluating the posterior fossa. However, imaging findings are often nonspecific, and diagnosis of brainstem lesions, either isolated or associated with supratentorial or cerebellar lesions, can be challenging.
Selective pontine involvement occurs in numerous disorders, and differential diagnosis usually dictates a review of imaging features in the context of the overall clinical picture.
In adults, evidence of restricted diffusion in the pons significantly narrows the diagnostic hypotheses among vascular, toxic-metabolic, inflammatory-infectious, neoplastic, and degenerative processes.
Methods & Results: We present three cases of pontine lesions with restricted diffusion, including acute ischemic stroke due to a paramedian perforating basilar branch occlusion, early-phase osmotic demyelination syndrome (ODS), and adultonset dentatorubral-pallidoluysian atrophy (DRPLA). Medical records and MR images were reviewed.
Conclusion: Differential diagnosis of pontine lesions with restricted diffusion in the adult population is limited, with pontine stroke being the most frequent.
Location and morphology of the lesions (vascular distribution in pontine infarction and sparing of the corticospinal tracts in ODS) and associated clinical features (presentation, onset, comorbidities, and family history) are the key to diagnosis.
Other less frequent lesions include abscesses, rhombencephalitis (infectious, autoimmune, or paraneoplastic), central nervous system vasculitides, active demyelinating disease, lymphoma, and some rare neurodegenerative disorders, such as autosomal-dominant DRPLA.
In each case, the integration of the imaging, clinical, epidemiological, and laboratory findings are indispensable to guide the differential diagnosis.

ID21 | HYPERTROPHY OF THE CLAVA, AN USEFUL IMAGING FINDING IN INFANTILE NEUROAXONAL DYSTROPHY

Pedro Sousa Brandão - pedrodsbrandao@gmail.com
Department of Neuroradiology, Centro Hospitalar Universitário de Lisboa Central, E.P.E., Lisbon, Portugal
Filipa Alveirinho[1], Eulália Viveiros[1], Ana Luísa Rodrigues[1], Carla Conceição[2]
1 Department of Pediatrics, Hospital do Divino Espírito Santo de Ponta Delgada, E.P.E.R., Azores, Portugal
2 Department of Neuroradiology, Centro Hospitalar Universitário de Lisboa Central, E.P.E., Lisbon, Portugal

Introduction: PLA2G6-associated neurodegeneration (PLAN) comprises a group of rare autosomal recessive neurodegenerative disorders caused by loss-of-function mutations in PLA2G6 gene.
Although clinical and imaging features frequently overlap, three main phenotypes are described primarily based on the age of onset and disease progression, namely infantile neuroaxonal dystrophy (INAD), atypical NAD, and PLA2G6-related dystonia-parkinsonism.
INAD patients present during the first 2 to 3 years of life, and rapidly progressive neurological deterioration and fatal outcome in the first decade are characteristic.
Diagnosis was previously only possible by histopathologic demonstration of widespread distal axonal degeneration with spheroid bodies (hallmark for neuroaxonal dystrophy), but detection of biallelic PLA2G6 pathogenic variants has become the diagnostic gold standard.
Methods & Results: We report two cases of genetically confirmed INAD with focus on the key role of MRI as part of the diagnostic workup and in the patient selection for PLA2G6 genetic testing.
Both male patients presented with psychomotor delay and regression under the age of 2. Symptoms progressed over time leading to severe disability (truncal hypotonia, hyperreflexia and spasticity), and one of the patients developed epilepsy.
Brain MRI showed bilateral cerebellar and optic nerves/chiasm atrophy, and hypointensities of globi pallidi and substantia nigra on susceptibility-weighted imaging. Though less conspicuous, hypertrophy of the clava (gracile tubercles on dorsal medulla) was also noted in both cases.
Conclusion: Clava hypertrophy is a known early imaging finding highly suggestive of PLAN/INAD. As in the cases reported, it is useful in the differential diagnosis of symmetrical cerebellar atrophy and iron deposits in the basal ganglia. Therefore, radiologists should be aware of this feature that can help guide genetic testing allowing prompt confirmation of the diagnosis.

ID22 | CEREBRAL ATROPHY - A CASE OF VITAMIN B12 DEFICIENCY

Filipa Castelão - fcastelao@gmail.com
Serviço de Neurorradiologia, Hospital Garcia de Orta, 2805-267, Portugal
Diogo Marques, Francisca Sena Batista, Cristina Marques, Ana Cristina Rios
Serviço de Neurorradiologia, Hospital Garcia de Orta, 2805-267, Portugal

Introduction/Goals: Vitamin B12 is essential for development of the central nervous system. In developed countries vitamin B12 deficiency usually occurs in infants who are exclusively breastfed, whose mothers have unrecognized pernicious anemia or are vegetarians, causing low stores of vitamin B12 in the infant at birth. Neurological manifestations include: lethargy, developmental delay, seizures, hypotonia, hypothermia, and coma. Brain MRI typically demonstrates thinning of the corpus callosum, brain atrophy, sylvian fissures enlargement, ventricular dilatation, and retardation in myelination.
This paper aims to alert to this entity, that has an excellent prognosis after vitamin replacement.
Material and Methods: We report 8-month-old girl, exclusively breast-fed for up to 6 months, who had been referred due to progressive hyptonia, apathy and developmental delay. The girl was ex-premature (35 weeks). Laboratory findings revealed normocytic anemia and vitamin B12 deficiency (<100pg/ml). Brain MRI was performed and revealed diffuse cerebral atrophy, thinning of the corpus callosum, sylvian fissures enlargement, slight retardation in the myelination and bilateral, chronic subdural hematomas, with no relevant mass effect. Vitamin B12 supplementation was initiated with excellent neurological improvement. The mother was diagnosed with pernicious anemia.
Results and Conclusion: Vitamin B12 deficiency is an easily treated clinical entity. Early diagnosis and therapy seem to be crucial for the prevention of permanent neurological squeal. Although brain MRI is not essential in the diagnosis, it is an excellent contribution to the evaluation of the repercussion of the effects of vitamin deficiency on the brain parenchyma.

ID23 | MITOCHONDRIAL LEUKODYSTROPHIES – A LYRM7-MUTATION CASE REPORT

Hugo Filipe Loureiro Cadilha - hcadilha@gmail.com
Centro Hospitalar Universitário Lisboa Note, 1649-035, Portugal
Joana Freitas, Guilherme Martins, Graça Sá
Centro Hospitalar Universitário Lisboa Note, 1649-035, Portugal

A male child presented with psychomotor developmental delay, noticed during the first year of life, due to delayed gait acquisition and hypotonia. At 20 months-old he had a distinct magnetic resonance imaging (MRI) pattern of bilateral symmetrical T2 hyperintense white-matter changes, with a preferential periventricular distribution and multiple cavitating lesions with peripheral increased restricted diffusion. Initial β-Galactosidase, galactocerebrosidase, arylsulfatase A and plasma very long chain fatty acid testing were negative. Next-generation sequence testing of 76 genes associated with leukodystrophies and leukoencephalopathies was also negative. By 3 years of age, he had clinically progressed to spastic diplegy, ataxic broad-based gait, dysmetria, genu recurvatum, delayed speech and language development, and progressively fell to percentile 3-15 of weight and height for age. Repeat MRI at 55 months-old showed worsening of the previously described lesion pattern, with more marked bilateral symmetric deep and periventricular white-matter T2-hyperintensity, larger well-defined cystic/cavitated lesions with foci of diffusion restriction at their edges, distributed along the periventricular white-matter, corpus callosum and medulla. Subsequent whole exome sequencing with mitochondrial genome analysis showed mitochondrial complex III deficiency nuclear type 8 (MC3DN8). MC3DN8 is caused by pathogenic homozygous mutations in the LYRM7 gene, encoding a protein which is part of the mitochondrial respiratory chain. There are multiple phenotypic presentations of LYRM7 gene mutations reported in the literature. We leverage this case report to illustrate MRI features that suggest mitochondrial leukodystrophies, and that may facilitate the diagnosis in future patients.

ID24 | CEREBRAL FOLATE DEFICIENCY: A CASE REPORT

João Nuno Oliveira - joao70610@gmail.com
Hospital Beatriz Ângelo
Catarina Brito, Ana Maria Braz, Joana Graça, Sandra Matias
Hospital Beatriz Ângelo

Introduction: Folate is a vitamin essential for brain metabolism and important for the synthesis of myelin. 5-methyltetrahydrofolate (5-MTHF) is the main transport folate form that crosses the blood-brain barrier through binding with the folate receptors α (FRα). Cerebral folate deficiency (CFD) is a neurological syndrome caused by a disorder of Fra with specific inability to transport 5-MTHF across the blood–brain barrier, associated with low concentrations of 5-MTHF in the cerebrospinal fluid, usually with normal serum folate levels.
Methods: We describe a rare case of CFD that presented in our institution and review key features.
Results: A 22-month-old female, with a history of developmental delay and hypotonia of lower limbs, was referred to our hospital because of altered mental status and seizures. The initial laboratory demonstrated a lower level of serum folate, while a brain CT showed multiple bilateral focal white matter and basal ganglia calcifications and corticosubcortical hypodensity of the right frontal, parietal and temporal lobes. A brain MRI was performed and demonstrated cerebellar vermian atrophy, bilateral frontal hypomyelination and restricted diffusion and T2 hyperintensity of white-matter and left head of caudate nucleus. A few weeks later a mutation in the FOLR1 gene was detected. Treatment with folate supplementation was initiated but seizures were refractory, and only ceased with a ketogenic diet.
Conclusion: Mutations in the FOLR1 gene, encoding for the FRα, represent a rare genetic cause of CFD and assay of cerebrospinal fluid should detect 5-MTHF depletion in the nervous system. Patients typically present with developmental delay, seizures, and cerebellar ataxia, with onset in early infancy. Neuroimaging findings include disturbed myelination, cerebral and cerebellar atrophy, T2 hyperintensities and parenchymal calcifications. In this case, it's not clear if there is concomitant defective malabsorption, resulting in systemic folate deficiency or if is due to malnutrition that exacerbated the underlying disease.

ID25 | NEUROSCHISIS OF THE CERVICOMEDULLARY JUNCTION – A CASE REPORT

Filipa Proença - filipa.proencaa@gmail.com
Centro Hospitalar Universitário Lisboa Norte - Hospital Santa Maria, Av. Prof. Egas Moniz MB 1649-028 Lisboa, Portugal
Diogo Marques, Filipa Castelão, Carla Conceição
2 Hospital Garcia de Orta, Av. Torrado da Silva 2805-267 Almada, Portugal
3 Hospital Garcia de Orta, Av. Torrado da Silva 2805-267 Almada, Portugal
4 Hospital Dona Estefânia, R. Jacinta Marto 8A Lisboa, Portugal

Introduction/Objectives: Neuroschisis is a rare embryonic malformation described when clefts remain in the neural plate or tube, that can range from minor cleft formation to complete neuroschisis of the spinal cord. There is a strong association between neuroschisis and patients with various malformations of musculoskeletal structures and internal organs with mesodermal origin. These malformations are labelled as axial mesodermal dysplasia complex malformations and includes many syndromes such as Goldenhar, Klippel-Feil and VACTERL.
Our purpose is to highlight, through a case report, that clefts seen on the cervicomedullary junction/spinal cord could signify neuroschisis and this abnormality could help in the diagnosis of mesodermal malformation syndromes.
Material and Methods: Here in, imaging findings of a 1-year-old female patient with Pakistan origin that was referred for evaluation of musculoskeletal abnormalities (vertebral malformations and varus feet), short neck with reduced head mobility, low hair line implantation and abnormal hearing having failed the neonatal hearing screening tests.
Results: In the imaging studies various abnormalities such as cleft in the cervicomedullary junction, inner ear malformations (cochlear incomplete partition type I and aberrant common origin and course of cranial nerves V and VII/VIII), craniovertebral and vertebral malformations were detected.
Conclusions: When clefts are present in the cervicomedullary junction/spinal cord neuroschisis should be evoked. This embryonic abnormality can be found in a few entities and axial mesodermal dysplasia complex malformations should be high on the differential diagnosis, especially if there are abnormalities with mesodermal origin. This will help to prevent a delay in diagnosis or avoid misdiagnosing the patients.

ID26 | CLINICAL AND IMAGING FEATURES OF CHILDHOOD ARTERIOPATHIES: FROM GENETIC PREDISPOSITION TO ENVIRONMENTAL FACTORS

Carolina Maia - carolinamaia.areal@gmail.com

Medical Image Department - Neuroradiology Functional Unit, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Pedro Barradas, Joana Pinto, Sílvia Carvalho; Teresa Garcia, Rui Pais

Medical Image Department - Neuroradiology Functional Unit, Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Childhood arteriopathies are increasingly recognized as a prevalent cause of childhood stroke, a strong predictor of recurrence and a predictor of poor short-term outcome.
Arteries are maintained by dynamic processes responsive to genetic and local signals. Genetically determined arteriopathies could reflect abnormal arterial development, whereas acquired arteriopathy could arise from disruptions in vascular homeostasis, for example, endothelial injury, repair, and angiogenesis. Increasingly, it seems that, in many of these, the interaction between genetic predisposition and environmental factors is essential to generate the ultimate disease phenotype.
Prompt diagnosis of arteriopathies is essential to limit injury and prevent recurrent stroke. However, challenges to diagnosis include lack of standardized diagnostic criteria, technical limitations of imaging studies, dependance on magnetic resonance angiography (MRA) over conventional angiography, and rare prevalence and heterogeneity of childhood arteriopathies.
We aimed to describe the clinical and imaging features of large medium and small vessel childhood arteriopathies that may aid neuroradiologists to produce a suitable differential diagnosis.

ID27 | FETAL AND POSTNATAL MR IMAGING OF DURAL SINUS MALFORMATION: 2 CASES FROM OUR DEPARTMENT

João Pedro Freitas Gonçalves - joao.gonc94@gmail.com

1 Neuroradiology Department, Hospital de São José, Centro Hospitalar Lisboa Central, R. José António Serrano, 1150-199 Lisboa, Portugal

2 Unidade de Neurorradiologia, Hospital Central do Funchal, Av. Luís de Camões 6180, 9000-177 Funchal, Portugal

Inês Freire[3], Filipa Proença[4], Carla Conceição[1]

1 Neuroradiology Department, Hospital de São José, Centro Hospitalar Lisboa Central, Lisbon, Portugal

3 Neuroimaging Department, Centro Hospitalar Lisboa Ocidental, Lisbon, Portugal

4 Imagiologia Neurológica, Centro Hospitalar Universitário Lisboa Norte, Lisbon, Portugal

Introduction: Dural sinus malformation (DSM) is a very rare condition that is defined by the presence of a dilated dural sinus pouch that presents with mural arteriovenous shunts, either torcular or non torcular. Torcular DSM are generally diagnosed during fetal or neonatal/infantile periods.

Methods: Two cases are presented, both with fetal and postnatal MRI. In the first case has two fetal MRI at 23 and 31weeks of gestation and postnatal MRI at birth, 6 days old and 20 months old. In the second case 5 MRI were performed at 22, 26, 31 and 35 weeks of gestation, as well as post nataly at 4 days old.

Results: Both cases had reduction in size of the DSM with different evolution throughout pregnancy. The first fetal MRIs demonstrate a dilation of the torcula, with partial thrombosis in the first case. In utero follow-up MRIs of this case show partial resolution of the thrombosis and reduction of the torcular ectasia. In the second case, only in the second fetal MRI did the partial thrombosis appear, with some complications in the follow-ups, namely choroidal plexus hemorrhage, venous aneurysm/varicose vein and signal changes in periventricular white matter, probably due to venous congestion. In the afterbirth MRI of both cases we can confirm the involution of the dural sinus malformation with a slight enlargement of some venous structures, without clear AV shunts in the TOF 3D sequences (only seen in the second case).

Conclusion: Prenatal MR imaging is a good tool to assess the whole extension of the dural sinus ectasia, as well as complications such as venous aneurysms, cortical or deep vein involvement and brain parenchyma lesions.

ID28 | LIMITED DORSAL MYELOSCHISIS: AN UNCOMMON FORM OF SPINAL DYSRAPHISM IN AN ELDERLY PATIENT

Gonçalo Almeida - goncalo.almeida@campus.ul.pt
Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal
João Pedro Gonçalves, Rui Carvalho, Carla Conceição
1 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal
2 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal
3 Department of Neuroradiology, Centro Hospitalar e Universitário de Lisboa Central, 1150-199 Lisbon, Portugal

Introduction: Limited dorsal myeloschisis (LDM) is an uncommon form of spinal dysraphism, characterized by the presence of a closed focal neural tube defect and a fibroneural stalk conneting the skin lesion to the spinal cord, which is thought to result from incomplete disjunction between cutaneous and neural ectoderms. While the majority of spinal dysraphisms are diagnosed at birth or in infancy, some cases are only recognized in adulthood. The aim of this work is therefore to present the neuroimaging findings of LDM in an elderly patient.
Material and Methods: Clinical and imaging data were retrospectively obtained from the patient's case files.
Results: A 70-year-old female with hypercholesterolemia presented with long-term low back pain radiating to the right groin. Physical examination of the patient revealed the presence of a tumefactive lumbar subcutaneous mass. A CT scan of the lumbar spine showed moderate spinal degenerative changes, as well as a heterogeneous solid-cystic subcutaneous mass connected to the spinal canal by a fibrous stalk through a posterior neural arch bone defect at the L4-L5 level. On MRI, the ventral aspect of the fibroneural stalk was connected to a low-lying conus medullaris (tethered cord). In addition, split cord malformation and a small syrinx were seen at L3-L4 and L2-L3, respectively. Taken together, the imaging features were consistent with the diagnosis of LDM. The patient underwent surgery with fibroneural stalk resection to correct the tethering effect on the spinal cord. Mild clinical improvement was achieved as the patient also exhibited spinal degenerative changes with radicular compression.
Conclusions: LDM is a distinct form of closed spinal dysraphism with specific imaging features. The presence of a fibroneural stalk connecting the spinal cord to a subcutaneous mass and additional findings (such as split cord malformation and syringomyelia) should suggest this diagnosis, even in an elderly patient.

ID29 | SCRATCH MY BACK: A CASE REPORT OF VERTEBRAL OSTEOMYELITIS IN CAT—SCRATCH DISEASE

Joana Freitas - joanaccfreitas@gmail.com
Centro Hospitalar Universitário Lisboa Norte, Av. Prof. Egas Moniz MB, Lisboa Portugal
Maria Inês de Sá, Filipa Proença, Sofia Reimão
Centro Hospitalar Universitário Lisboa Norte, Av. Prof. Egas Moniz MB, Lisboa Portugal

Objectives: To report a case of cat-scratch disease with vertebral osteomyelitis.
Methods: We describe imaging findings, clinical features, diagnostic procedures and treatment options in one patient with vertebral osteomyelitis due to Bartonella henselae.
Results: A 32 years-old man, living in a rural environment with contact with several animals, presented with adynamia, asthenia, cervicalgia and a 2cm painless supraclavicular adenopathy. He was treated with analgesics without improvement but also with appearance of fever, left sacrococcygeal pain, and difficulty in initiating urination. Complete blood cell count was normal and there was a slight elevation in inflammatory markers; blood cultures were negative and serology for Bartonella henselae values were on the upper limit; he underwent a spinal MRI with identification of atlanto-axial joint effusion, lytic lesions in D3, D9, L3 and L4 with signs of arthritis, as well as a lytic lesion on the left iliac bone; abdomen CT showed several hypodense infracentimetric hepatic and splenic nodular lesions, with discrete peripheral contrast uptake. The most likely diagnostic hypothesis was disseminated disease by Bartonella henselae with hepatosplenic and spinal involvement, thus doxycycline and rifampicin were initiated.
Bartonella henselae infection was confirmed by positive IgM on Western Blot.
Conclusion: Cat-scratch disease should be considered in differential diagnosis of localized lymphadenitis. Since it has variable clinical pictures, with insidious onset and non-specific symptoms, radiology often plays an important role in suggesting the diagnosis. Osteomyelitis is an unusual manifestation but should be considered in the case of vertebral lesions with negative blood cultures, recent history of local lymphadenopathy and cat exposure, or if the patient does not respond to standard therapies. In fact, osteoarticular pain or limitation should always be investigated in the setting of this disease looking for bone spreading with MRI being the best radiographic technique to define bone and surrounding tissue involvement.

ID30 | DIASTEMATOMYELIA: A MARRIAGE WITH DIVISION OF ASSETS

João Nuno Oliveira - joao70610@gmail.com
Hospital Beatriz Ângelo
Catarina Brito, Sandra Matias
Hospital Beatriz Ângelo

Introduction: Diastematomyelia is a split-cord malformation, where the spinal cord is split into two hemicords, believed to occur during the gastrulation stage of development. This malformation accounts for 4% of all closed spinal dysraphisms, that are more common in lower cord and is divided into type I and type II. Type I diastematomyelia is characterized by two individual dural sacs separated by an osteocartiliginous/bony spur, while in type II there is a single dural sac that contains both hemicords, without a septum.
Methods: We describe two cases of diastematomyelia in childhood and review key features.
Results: In these two cases, patients had no neurologic symptoms/signs and were investigated for other deformities. A 4-yearold male, with down syndrome and sprengel deformity, performed a cervical and dorsal MRI that demonstrated hydrosyringomyelia in some levels, diastematomyelia apparently split by cartilaginous septum in D12 level and a thick filum terminale without tethered spinal cord. A 7-month-old female, with neurofibromatosis type I, followed in our hospital to investigate progressive scoliosis performed a cervical and dorsal MRI. This exam revealed D5 hemivertebra, right dorsal scoliosis, diastematomyelia at L3 level without osteocartilaginous/bony septum, conus medullaris reaching L4 vertebral level with posterior deviation of filum terminale and signs of tethered spinal cord.
Conclusion: Diastematomyelia may be isolated or associated with other segmental anomalies of the vertebral bodies, like bifid lamina, hemivertebra or bifid vertebra, more frequently in type I. The symptoms, more frequent in type I, can include gait disturbance with mild atrophy or weakness of lower extremities, progressive paraparesis, and bladder or bowel dysfunction. Skin pigmentation, hemangioma, and hypertrichosis are common signs that can suggest this malformation. MRI is the modality of choice for assessing children with split cord malformations, but CT can better image the bony septum. The role of surgery before neurological dysfunction is controversial.

Notas

Patrocinadores