B117 / Poster • Posterior reversible encephalopathy syndrome in pediatric leukemia. Case report.

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AIM We describe the case of a 5-year-old boy undergoing induction for acute T-cell lymphoblastic leukemia (T-ALL) with CNS involvement. He was treated according to the protocol EORTC 58081-VHR-CNS3. During the second week on induction he developed generalized seizures with loss of consciousness. Blood pressure was elevated (150/100mmhg = 95th percentile). Computer tomography showed no abnormalities. EEG was slow with signs of cerebral distress. Magnetic resonance imaging (MRI) - T2 weight and FLAIR images - showed subcortical with matter abnormalities localized bilaterally in the parietal and occipital lobes. The diagnosis of posterior reversible encephalopathy syndrome (PRES) was suspected. Treatment with antihypertensive (calcic antagonist) and antiepileptic (levitiracetam) drugs was started and chemotherapy was transient discontinued. After two weeks, he presented a second episode of seizure associated with new hypertensive crisis. Increased brain damage was observed at MRI with involvement of the white matter of frontal and temporal lobes. Additional antihypertensive agents were started (beta-blockers and AEC inhibitor). After one month the patient showed a complete clinical recovery and MRI follow up showed significate regression of the lesions, confirming the diagnosis of PRES.

RESULTS PRES is a rare potential complication of cancer treatment that has been increasingly recognized since the apparition of MRI. Some few cases of children treated for ALL are discussed in the literature. PRES symptoms consist of seizures, headaches, altered level of mental status and cortical blindness. Chemotherapeutic and immunosuppressor agents seem to be risk factors for the development of PRES. Hypertension plays a central role in the pathophysiology of the PRES. Nevertheless, other factors also play a role as suggested by the absence of hypertension in over 20% of the cases. MRI typically shows vasogenic oedema in the posterior regions of the brain but some lesions can also be observed in other areas as parietal, temporal or even frontal areas like observed in this case. The differential diagnosis of PRES in childhood cancer consists of CNS infection, CNS development of the malignancy, methotrexate encephalopathy, metabolic causes like hyponatremia and stroke.

CONCLUSION

Symptoms and radiological finding normalize in 90% of the cases, but in 10% neurological symptoms remain. Early treatment of hypertension, control of seizure activity, and withdrawal of inciting agents can lead to rapid reversal of symptoms and return to baseline functioning. Further studies could focus on long term functioning in children after PRES with the hope to better define factors predisposing to a worse neurological evolution.

B118 / Poster • A Rare Case of Severe Thrombocytopenia with transient Pancytopenia in the neonatal period.

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INTRODUCTION We report the case of a male neonate who presented on day 1 of life with thrombocytopenia and petechiae. Until now no congenital, immunological and/or acquired etiology could be identified.

RESULTS Case Report A one-day-old boy, the first child born of non-consanguine, Caucasian parents, was transferred to the neonatal intensive care unit due to thrombocytopenia with a platelet count of 17.500/mm³. Familial history was negative. Mother was O-negative, she had one spontaneous abortion in early pregnancy before. Serology status was negative, GBS unknown, but treated. The boy was born after 36 weeks of pregnancy after spontaneous labour. Birth weight was 2,700 kilograms (P50-75), length 48 cm (P50-75) and head circumference 34.5 cm (P75-90). Pediatric examination at presentation was completely normal with exception of numerous petechiae on the trunk. Blood group was A positive, direct Coombs-test negative, as well as the tests for neonatal allo-immune thrombocytopenia. Sepsis work-up, urine PCR for viruses and serology for parvovirus, toxoplasmosis and syphilis all were negative. Metabolic screening showed no arguments for metabolic diseases. Imaging showed no abnormalities. Genetics showed a normal male karyotype. Micro-array revealed duplication at 4q12 (inherited of the father), consisting a part of the SRP22-gene, which is associated with (adult-onset) bone marrow abnormalities and with congenital nerve deafness. The hearing of our patient however was normal. Mutation analysis of the MPL-gene was negative, which excluded CAMT (Congenital Amegakaryocytic Thrombocytopenia) and analysis of WASP-gene (Wiskott-Aldrich syndrome), was also negative. Bone marrow aspiration revealed lowered amount of megakaryocytic cells, but no amegakaryocytosis, and was suggestive for immunologic etiology. Thrombopoietin was elevated, which indicated a production problem in the bone marrow. Adamts-13 Ag and activity were normal. During his first days of life, he received several platelet transfusions but he relapsed every time. On Day 5, intravenously immunogobulines were administered but without response on the platelet count. On day 8, the boy developed pancytopenia. On day 16, oral corticosteroid therapy was started and later also cyclosporin. On day 26, he still was thrombocytopenic and we decided to taper and stop corticosteroids and cyclosporin. Until now his platelet count is stable around 20.000/mm³ without platelet transfusions since a few months. Discussion Neonatal thrombocytopenia often presents early after birth. The majority is infectious or immune in etiology. In rare cases, a congenital disorder is the cause, like congenital amegakaryocytic thrombocytopenia (CAMT). In rare cases, children fulfil clinical criteria, but lack detectable c-Mpl mutations, they might have mutations in upstream non-coding gene sequences that regulate c-Mpl expression.

CONCLUSION We reported the rare case of severe and chronic neonatal thrombocytopenia. The majority is infectious or immune of origin, but in rare cases, a congenital disorder is the cause. Until now, no definitive diagnosis could be established in our case.

B137 / Poster • Atypical bifocal intracranial tumor. A case report.

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INTRODUCTION Primary malignant central nervous system (CNS) tumors are the second most common childhood malignancies, and are the most common pediatric solid organ tumor. CNS germ cell tumors (GCT), one subtype of these tumors, usually affect the pediatric and adolescent population, with a predilection for males. They usually develop in the midline around the third ventricle, particularly in the suprasellar and pineal regions. They can be broadly divided into two major histological subtypes pure germinomas and non-germinomatous germ cell tumors (NGGCT). Around 30% of pure germinomas are bifocal, involving both the suprasellar and pineal region.

AIM We report an atypical case of intracranial bifocal tumor, involving the pituitary and epiphyseal region, in a 10-year old boy. The imaging was realized at the occasion of epileptiform events. The pituitary lesion contained 8 perfectly differentiated teeth and was well delimited, compatible with a mature teratoma. The epiphyseal lesion evoked a germinoma. The endocrinological exploration disclosed no anomaly, except for a partial deficit in growth hormone. No other lesion was vizualised in the entire brain and medullar region, and alpha feto-protein (AFP) and human chorionic gonadotropin (hCG) measured in blood and cerebrospinal fluid (CSF) were negatives. Four cures of neoadjuvant chemotherapy were administrated but there was no regression of any of the lesions. The epiphyseal lesion was removed surgically without any complication. Surpisingly, the anatomopathological examination revealed a pineal cyst without any tumoral component. Subsequently, four teeth were resected from the supra sellar region, confirmed by the histological examination. The residual pineal lesion appeared stable at the imaging. The decision was then taken to limit ensuing medical care to a radiological follow-up, without further radiotherapy or chemotherapy. Atypical movements of the head reappeared one year after the second neurosurgery, but the clinical and follow up X-ray examinations have been reassuring. A diagnosis of uncomplicated tics is retained. The lesion has been stable since the last surgery (16 months ago).

CONCLUSION

The discussed case is atypical for two reasons. Firstly, to our knownledge, an intracranial lesion containing teeth has only been described once in the literature, and there is no report of a bifocal presentation of such a lesion. Secondly, our case contradicts the prevailing consensus that the occurrence of synchronous bifocal intracranial tumors associated with normal AFP and normal or slightly hCG levels is pathognomonic of germinomas. A review of the literature shows that this is not an isolated case, suggesting that intracranian bifocal NGGCT should been included in the differential diagnosis. Accordingly, the potential benefit of a histological diagnosis should be balanced against risks of neurosurgery, keeping in mind the stereotactic limitations of a biopsy because of the potential presence of heterogeneous subtypes in a same lesion.

B140 / Poster • Choroïd plexus tumor in children

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INTRODUCTION Choroid plexus tumors (CPT) are rare since they represent roughly 4 % of all children brain tumors. They are divided into three grades according to both WHO classification and pathological characteristics. Grade Ipapilloma, grade II atypical papilloma and grade III carcinoma. The prognosis depends on tumor grade, percentage of surgical removal, and absence of metastatic location.

AIM The aim of this study was to analyze clinical data, medical work-up, and clinical evolution.

METHOD Between March 1995 and December 2014, we performed a single centre study which included 8 consecutive patients (pts) diagnosed of having CPT. The analyzed data were age at the time of diagnosis, duration between onset of symptoms and the final diagnostic procedure, clinical signs and symptoms including ophthalmological findings at the time of diagnosis, pathological analysis, evaluation of both medical and surgical treatments.

RESULTS At diagnosis median age was 19 months (0-109 months). Median interval between onset of symptoms and diagnosis was 1.5 week (0-92 weeks). Intracranial hypertension was found in 7/8 pts, hemiparesis in 2/8 pts, cognitive decline in 1/8 pt, visual loss in 1/8pt, and 2 pts had normal neurological examination. Severe optic fundus edema (OFE) was found in 6/8 pts, while 1/8 had mild OFE. Pathological findings disclosed 3 grade I papilloma, I grade II papilloma, and 4 grade III papilloma. The most frequent location was the lateral ventricle, found in 7/8 pts, while the last pt had a location at the lateral and third ventricle junction. A suspected phenotypical familial presentation of Li Fraumeni was found in 1 pt with grade II papilloma but without genotypic confirmation. All pts were surgically treated, Chemotherapy was given in all pts with carcinoma, 2 pts underwent also radiation therapy.7 out of 8 pts were alive after a 45 mo of follow-up (1-285 mo). One pt died after 2 years of tumor recurrence despite having had a second line surgery and radiation treatment.

CONCLUSION

Despite highly malignant, CPT remained rare. Early recognition of clinical signs and symptoms suggesting of tumor evolution allows early diagnosis and therefore accurate treatment. Total surgical removal associated to chemotherapy allows disease remission in most pts. The role of radiation remains controversial in CPT treatment but several studies dedicated to this point are yet in progress.