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

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-WHR-CNS3. During the second week on induction he developed generalized seizures with loss of consciousness. Blood pressure was elevated (150/100mmHg). On day 28, he still was thrombocytopenic and therefore had MRI with involvement of the white matter of frontal and temporal lobes. Additional antihypertensive agents were started (beta-blockers and ACE inhibitor). After one month the patient showed a complete clinical recovery and MRI follow-up showed significant 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 appearance of MRI. Some few cases of children treated for ALL are discussed in this literature. PRES symptoms consist of seizures, headaches, altered level of mental status and cortical blindness. Chemotherapeutic and immunosuppressive agents seem to be risk factors for PRES. Hypercortisolism and Hypertension play 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 as observed in this case. The differential diagnosis of PRES in childhood cancer consists of CNS infection, CNS development of the malignancy, metronidazole encephalopathy, metabolic causes like hyponatraemia and stroke.

CONCLUSION Symptoms and radiologic findings normalize in 90% of the cases, but in 10% neurological symptoms remain. Early treatment of hypertension, control of seizure activity, and withdrawal of inducing 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.
PEETERS E., MAES PH., WOJCECHOWSKI M. / UZA

INTRODUCTION We report the case of a male neonate who presented on day 1 of life with thrombocytopenia and pancytopenia. 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-consanguineous, 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, syphilis and hepatitis was in the neonate's history. The boy was born after 36 weeks of pregnancy after spontaneous labour. Birth weight was 2.700 kilograms (PSD-75), length 48 cm (PSD-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. Gesipno work-up, urino PCR for viruses and serology for parvovirus, toxoplasmosis and syphilis was all negative. Microscopic 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 SPG22-gene, which is associated with (idiot-onset) bone marrow abnormalities and with congenital bone deformities. The hearing of our patient however was normal. Mutation analysis of the MPL -gene was negative, which excluded CMT1 (Congenital Amegakaryocytic Thrombocytopenia) and analysis of WASP-gene (Wiskott-Aldrich syndrome), was also negative. Bone marrow aspiration revealed lowered amount of megakaryocytes, but no amegakaryocytosis, was suggestive for immunologic thrombocytopenia. Thrombocytopenia was elevated, which indicated a production problem in the bone marrow. Atadis-13q4 and activity were normal. During his first days of life, he received several platelet transfusions but he relapsed every time. On Day 5, intravenous immunoglobulines were administered but without response on the platelet count. On Day 8, the boy developed pancytopenia. On Day 10, oral corticosteroid therapy was started and later also cyclosporin. On Day 25, 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 origin. In the case of congenital disorder is the cause, like congenital amegakaryocytic thrombocytopenia (CMT). In rare cases, children fulfill 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.
PETTE C., SEPULCHRE E. / CHR Citadelle Liège

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 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 pineal region, in a 10-year-old boy. The imaging was realized at the occasion of epilepticiform events. The pilonidal lesion contained 8 perfectly differentiated teeth and was well delimited, compatible with a mature teratoma. The epiphelial lesion evoked a germinoma. The endocrinological exploration disclosed no anomaly, except for a partial deficit in growth hormone. No other lesion was visualized in the entire brain and medullary region, and alpha-feto-protein (AFP) and human chorionic gonadotropin (hCG) measured in blood and cerebrospinal fluid (CSF) were negatives. Four curves of neoadjuvant chemotherapy were administered but there was no regression of any of the lesions. The epiphelial lesion was removed surgically without any complication. Surprisingly, 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 taken to limit ensuing medical care to a radiological follow-up, without further radiotherapy or chemotherapy. A follow-up of the head reappread 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 age).

CONCLUSION The discussed case is atypical for two reasons. Firstly, to our knowledge, an intracranial concomitant 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 intracranial bifocal NGGCT should be 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.

INTRODUCTION Choroid plexus tumors (CPT) are rare since they represent roughly 4% of all childhood brain tumors. They are subclassified in WHO grades according to both WHO classification and pathologic characteristics. Grade I pappioma, grade II atypical pappioma 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 2/8 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 pappioma, 1 grade II pappioma, and 4 grade III pappioma. 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. A suspected choroidal familial predisposition. Preoperatively, a formal diagnosis of a 1 pt with grade II pappioma but without genotypic confirmation. All pts were surgically treated. Chemotherapy was given in all pts with carcinoma, 2 pts underwent also radiation therapy. 7 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 tumor evolution is of utmost importance. Early surgical treatment will have the best 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.