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285 POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES) – A RARE, SEVERE AND NEW COMPLICATION DURING THE TREATMENT OF ACUTE LEUKEMIA IN CHILDREN

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Aim To present seven cases of Posterior Reversible Encephalopathy Syndrome (PRES), diagnosed between 2014-2019, as a complication of acute leukemia treatment in children. PRES was cited for the first time in 1996 by Hinkey and collaborators as a posterior leukoencephalopathy syndrome.

Methods These patients were admitted and treated for Acute lymphoblastic leukaemia during a 5 years period. From the total number of 162 patients with leukemia only 7 cases developed PRES. They were 3 to 12 yo, 5 males and 2 females. None of them had a preexisting neurologic disorder. PRES occurred during the induction chemotherapy. All patients underwent clinical evaluation, ECG, echocardiography and laboratory tests. At the onset of the neurological symptoms emergency head CT-scan was performed, followed by head MRI which confirmed the diagnosis.

Results Neurologic manifestation of PRES consisted of upper or lower limb muscle spasm, marked agitation, aggressive behavior, generalized seizures followed by coma, right upper limb hemiparesis and facial paresis. The first patient developed PRES as early as the 9-th day of treatment after receiving high dose Prednison that induced hypertension. In the rest of the children PRES occurred simultaneously with sepsis (due to severe bone marrow aplasia), SIADH (Syndrome of inappropriate antidiuretic hormone secretion) or uncontrolled high blood pressure due to corticotherapy. Blood pressure was controlled only with combined antihypertensive agents at maximal dose.

The PRES diagnose was consistently based on cerebral MRI neuroimages that showed typical lesions of vasogenic edema with hyperintense T2 and FLAIR signals with a characteristic location for PRES. Prompt anticonvulsant and antihypertensive therapy was started, and the patients made a full neurological recovery. The MRI was repeated with minimal ischemic lesions after 1 mo and almost normal after 3 mo. The treatment protocol for leukemia was interrupted from 12 to 43 days.

Conclusions PRES is a rare, severe and almost new described complication in children with leukemia. We have to recognize and to treat it promptly in order to save the patient. A multi-disciplinary team (pediatric oncologist, cardiologist, neurologist, radiologist and intensive care specialists) is necessary to manage such cases. A positive diagnosis was established after performing cerebral MRI. Severe acute hypertension was controlled only with a combination of for anti-hypertensive drugs administered at a maximum dose.

The question regarding the etiology of PRES is still present, because only patients with sepsis, SIADH and hypertension due to corticotherapy developed the syndrome from all leukemia patients.

286 MALIGNANT PERIVASCULAR EPITHELOID CELL TUMOR IN A TEN-YEAR-OLD GIRL

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Perivascular epitheloid cell tumors (PEComas) represent a group of mesenchymal neoplasms characterized by the presence of epitheloid cells of mixed myo-melanocytic differentiation with uncertain malignant potential. The tumors are very rare with a female preponderance and median age of 45 years. The most common location is retroperitoneum, but almost every site has been reported. PEComas have an unpredictable clinical behavior.

We present a 10-year old girl with a 3-week history of progressive swelling in the left infraclavicular region. At examination, a painless, well circumscribed, firm mass measuring 3 cm in diameter was palpable, with no other abnormal clinical findings. Laboratory tests were within normal limits. The ultrasound showed well defined, echogenic heterogeneous soft tissue mass with discrete vascularization. Fine needle cytology was suspicious of a malignant mesenchymal tumor. The girl was referred to the pediatric surgeon, and complete resection was done. Pathology finding established the diagnosis of PEComa with malignant morphology and peculiar immunophenotype of tumor cells (negative for melanocytic markers HMB-45 and Melan A, but positive for MITF). Two weeks later, two painless nodules, 5 and 10 mm in diameter, were noticed at the site of the excised tumor.

Subsequent thorough examination, including magnetic resonance imaging of the primary site, chest X-ray, chest and abdominal computed tomography, and positron emission tomography-computed tomography, was without evidence of disease. The girl underwent re-excision with clear resection margins. She was treated with adjuvant chemoradiotherapy for non-rhabdomyosarcoma pediatric soft tissue tumors. At 2-year regular follow-up, the girl is well and with no signs of recurrence.

To the best of our knowledge, this is the first reported case of malignant PEComa in a young patient. Our case highlights the challenges with regard to preoperative diagnosis and treatment. Due to the uncertain prognosis, close clinical surveillance accompanied by radiological imaging is mandatory.

287 FREQUENCY OF FACTOR II, FACTOR V LEIDEN AND MTHFR MUTATIONS IN CHILDREN WITH CANCER

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Thrombosis is an increasingly recognized complication of malignancy, occurring in up to 20% of patients with cancer. Cancer-associated thrombosis is linked with poor prognosis, being the second leading cause of death in cancer patients. The pathogenesis is complex, and includes multiple genetic and acquired factors. There is significantly less knowledge