this interesting case is the first report of the occurrence of proteinuria in a child with Castleman disease.

049

PRIMARY RHABDOMYOSARCOMA OF THE BRAIN

Sule Yesil, Yasar Kandur, Neriman Sari, Inci Ergurhan Ilhan

Ankara Oncology Research and Education Hospital, Pediatric Oncology, Ankara, Turkey

Purpose: Primary intracranial rhabdomyosarcoma (RMS) is a rare tumor in childhood that is found in various locations in the central nervous system. This report presents such a case.
Method: This 6 year old boy presented with severe headache, emesis, and double vision. On examination, left 6th and 7th cranial nerve palsy was noted. MRI revealed a temporal mass extending to frontal lobe, basal ganglia, sylvian fissure and obliterating the left ventricle. The patient underwent a left frontotemporal craniotomy with gross total tumor resection. The pathological diagnosis was embryonal RMS. Postoperatif MRI revealed left hipocampal residual nodule. Computerized tomography (CT) of chest showed metastatic nodules in both lungs. Radiation therapy with a dose of 54 Gy was applied to whole brain. He concurrently received chemotherapy consisting of ifosfamide, carboplatin and etoposide. At the end of induction chemotherapy, CT revealed considerable resolution of the lung metastasis and also MRI revealed resolution of the hipocampal residual nodule. After 6 months of remission period, cranial MRI revealed recurrence of the hipocampal nodul. Total re-excision of tumor was performed and histopathological examination revealed embryonal RMS. Post-operative MRI revealed a new periventricular lesion. MRI-Spectroscopy also supported this finding. A regimen of chemotherapy including intravenous vincristine, cyclophosphamide, adriamycin and cisplatin and intrathecal methotrexate, cytosine arabinoside and prednisolon was administered. Under chemotherapy, cervical lymph node metastasis was detected. Patient's family rejected further treatment and he is still alive with progresive disease after 30 months of diagnosis.
Results: Primary RMS of the central nervous system (CNS) is rare in children. The outcome in the majority of cases is poor and many cases are associated with early mortality as in our case Conclusion: In spite of advances in the treatment of RMS, primary brain localization has still very poor prognosis. Novel approaches for treatment of special localizations are required.

050

CEREBRAL VENOUS AND RIGHT ATRIAL THROMBOSES IN A CHILD PRESENTING WITH MULTISYSTEMIC LANGERHANS CELL HISTIOCYTOSIS (MS-LCH)

Manuel Diezi, Sarah Fattet, Yves Pastore, Nicolas von der Weid, Maja Beck-Popovic

Centre Hospitalier Universitaire Vaudois, Pediatric Hematology/Oncology, Lausanne, Switzerland

Purpose: A 3 years old boy presented with headache, ocular pain, prominent frontal veins, irritability and occasional vomiting.
Method: Clinical examination revealed turgescent frontal veins, marked exophtalmos, moderate hepatomegaly and congested retinal veins on fundoscopy.
Results: Thromboses of the sagittal, transverse and sigmoid sinuses were confirmed on cranial MRI. Leukoencephalopathy compatible with either chronic venous hypertension or LCH-CNS involvement was also noted. Biology showed elevated sedimentation rate (ESR), microcytic anemia, leucopenia and slightly elevated triglycerides. A comprehensive thrombophilia work-up including antithrombinIII, protein C/S, Factor VLeiden/Prothrombin mutations, homocystein level and antiphospholipid antibodies was negative. Fibrinogen, sCD25 and TNFalpha were elevated.Radiology showed multiple osteolytic lesions of the scalp, ribs, pelvis and inferior extremities. Histology of a lesion revealed CD1a-positive dendritic cells with Birbeck granules. Bone marrow was hypercellular with focal histiocytic proliferation and hemophagocytosis. MS-LCH with secondary HLH was diagnosed. Subcutaneous LMWH and chemotherapy according to LCH-III protocol were started. Reevaluation after 6 weeks showed partial reopening of cerebral thromboses and lower ESR but otherwise persisting disease activity. After re-induction as per protocol no further clinical, biological or radiological benefit could be achieved and we proceeded to salvage therapy for refractory MS-LCH (LCH-S-2005) combining HDAra-C and 2-CDA. After the first course, the patient developed an asymptomatic thrombus of the roof of the right atrium, unrelated to the central venous catheter. Full UFH-anticoagulation remaining unsuccessful, surgical removal of the thrombus under extra-corporeal circulation was performed. Microscopic examination revealed lymphoplasmocytic-histiocytic inflammatory infiltrates negative for S100 and CD1a. A second thrombophilic work-up remained unremarkable.
Conclusion: To our knowledge, this is the first description of such significant thrombotic events in a child with MS-LCH. Common inherited and acquired predisposing factors were excluded and we hypothesize that the multiple thromboses observed might be secondary to the activation of dendritic cells and cytokine release described in histiocytoses and acute inflammatory diseases.