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HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS IN TWO ROMANIAN YOUNG INFANTS

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Haemophagocytic lymphohistiocytosis (HLH) is a rare disorder of early infancy, resulting from abnormal proliferation of histiocytes in tissues and organs, but potentially fatal disease. Diagnosing this disease may be difficult and is often delayed because the clinical presentation mimics other conditions like severe sepsis, hepatic failure and malignancies. HLH is characterized by fever, hepatosplenomegaly, cytopenia, hypertriglyceridemia and/or hypofibrinogenemia, hyperferritinemia with hemophagocytosis in the bone marrow, spleen or lymph nodes, which represent the diagnostic criteria.

We report two cases of HLH in infants within the first 10 weeks of life. Diagnosis of HLH was made relatively early after symptoms started, one of them - a rare case of primary HLH form (familial form), and other - the secondary form of HLH (associated infection with EBV virus) indicating that not all cases of HLH in very young infants are familial. Both patients had typical presentations with fever, anemia and thrombocytopenia, enlarged liver and spleen, hyperferritinemia, hypertriglyceridemia and hypofibrinogenemia, initially without the finding of hemophagocytosis in bone marrow. Awareness of the clinical symptoms and of the diagnostic criteria of HLH is important to start life-saving therapy with immunosuppressive/immunomodulatory agents in time. Diagnostic must be rapidly made, because the familial form is always fatal without treatment. In the absence of a specific marker, differential diagnosis may be difficult, especially in patients without familial recurrence.

When there is strong clinical suspicion of HLH, chemotherapy and immunosuppressor treatment should be started early.

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IT MAY BE MORE THAN JUST A DENTAL ABCESS

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An 11-year-old boy is brought to the Emergency Department with a history of a dental abscess in the upper jaw. He was previously treated with Amoxicillin, Clavulanate plus Clindamycin, without any improvement. He complained of limb weakness, paresthesias, abnormal gait and urinary hesitancy.

On physical examination he had swelling of the face on the right side, with slight homolateral proptosis. The neurologic examination revealed decreased muscular strength in the limbs; deep tendon reflexes were brisk and symmetric throughout and sensory function was apparently normal.

The laboratory analysis showed leukocytosis, neutrophylia, elevated erythrocyte sedimentation rate and lactate dehydrogenase.

The patient was hospitalized, keeping the antibiotics previously prescribed. On day 3 he started low-grade fever with no improvement of the clinical situation. A maxillofacial CT was performed, showing opacification of the sinuses, presence of an expansive lesion occupying the right maxillary sinus with erosion of the wall, extending to the orbit, infratemporal and buccal areas.

He underwent an Endoscopic Sinusectomy, with washing of the right maxillary sinus, and right anterior ethmoidectomy with drainage of hemopurulent liquid.

Clindamycin was suspended and he was started on Metronidazole, Netilmicin and Prednisolone. Although apyretic, he kept complaints of pain and limb weakness, accompanied by nocturnal sweating.

A Cervical-thoracic MRI was performed, showing a paravertebral mass, spinal canal infiltration, alteration of bone trabeculation and insuflation of the vertebral body at D5 level.

The diagnosis of FAB L3 Acute Leukemia was established based on more than 30% blasts in the bone marrow aspirate. The patient underwent chemotherapy.