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A Langerhans cell histiocytosis in its rare Letterer-Siwe form in a child mimicking an otoantritis: A case report and literature review

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Langerhans cell histiocytosis (LCH) is a group of three rare diseases (annual incidence less than 4/1000.000 H) with increasing aggressiveness respectively: Eosinophilic granuloma (unifocal solitary osteolytic lesion), Hand-Schuller-Christian disease (multifocal lesion) and Letterer-Siwe disease (the most aggressive, disseminated disease with systemic manifestations). The bone is the most commonly involved with a predilection for the skull. Some cases of temporal bone localisations have been described in the literature. We report a case of an 18 month child presenting a LCH of the temporal bone misdiagnosed at the beginning as an otoantritis. In fact, the child presented a retro-auricular swelling with inflammatory characters, fever and otorrhea. The exploration before surgery revealed besides the extensive temporal lesion, hematologic perturbations, a splenomegaly and a hepatomegaly. The histopathology confirmed the diagnosis of a LCH. We describe the diagnosis challenges, the surgical difficulties and the therapeutic response of the child and give a review of what was published on LCH and Letterer-Siwe disease.

Biography

M Bouali is currently working as faculty of medicine in department of Otorhinolaryngology at Oran University, Algeria.

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