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Clinical Image

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Extensive cranial langerhans cell histiocytosis revealed by acute mastoiditis

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Clinical image description

Langerhans cell histiocytosis is a group of diseases characterized by an inflammatory neoplasia of myeloid precursor cells driven by mutations in the mitogen-activated protein kinase pathway leading to the proliferation and accumulation of Langerhans cells [1,2]. Clinical manifestations range from isolated disease to multifocal lesions and systemic involvement with a worse prognosis [2,3]. The otologic features are rare and may include otorrhea, otalgia, subjective hearing loss and vertigo misleading the diagnosis to an inflammatory process [3]. The CT scan shows destruction of the skull with characteristic "punched out" lytic lesions that can be multiple and multifocal. Tissue biopsy confirms the diagnosis [3]. We report the case of a 4 years old boy who presented to the E.R with a 2-week left-sided otalgia and otorrhea without any improvement under local antibiotics, associated to superficial temporal tenderness. The physical examination revealed an acute otitis media, with an inflammatory swelling in the mastoid area with no facial palsy or pain. The CT scan showed destructive "punched out" lytic bilateral lesions of the temporal bones with signs of mastoiditis and significant soft tissue thickening in the left side, associated to multiple lytic lesions in the Diploë with destruction of the cortical outer layer of the skull vault, suggesting an extensive cranial Langerhans cell histiocytosis. The diagnosis was confirmed by histology. Regarding the extensive nature of the disease process, the patient was referred to haematology for further treatment. **Citation:** El Bouhmadi K, Loudghiri M, Oukessou Y, Rouadi S, Abada R, et al. Extensive cranial langerhans cell histiocytosis revealed by acute mastoiditis. J Clin Images Med Case Rep. 2022; 3(9): 2039.



Figure 1: 3D reconstructions of cranial CT scan showingmultiple "punched out" lytic lesions in the Diploë with destruction of the cortical outer layer of the skull vault.



Figure 2: Coronal sections of cranial CT scan showing lytic destructive bilateral lesions of the temporal bones with mastoiditis and significant soft tissue thickening in the left side.

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