Fluid–fluid level in Langerhans cell histiocytosis of the skull

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ABSTRACT

We present a case of solitary eosinophilic granuloma in the skull of a 6-year-old Saudi boy. This osteolytic lesion has fluid-fluid level on CT and MRI. We are presenting a rare radiological finding of eosinophilic granuloma.

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Eosinophilic granuloma (EG), Hand-Schüller-Christian disease and Letterer-Siwe disease are diseases characterized by idiopathic proliferation of histiocytes producing focal or systemic manifestations. Collectively, we call them Langerhans cell histiocytosis (LCH).[1] The etiology and pathogenesis of these diseases are still unknown.[1] Langerhans cell histiocytosis is a rare disorder that affects the pediatric population.[2] The localized form of which is commonly referred to as eosinophilic granuloma (EG); the term is reserved for cases where the disease is limited to bone or lung.[1] Eosinophilic granuloma is a relatively rare disease typically presenting in children, and accounts for most of the cases of LCH.[3] We consider this localized form the least aggressive expression of the disease, and has the best prognosis. The most common site of involvement is the skull; especially prevalent in the frontal and parietal bones.[4] Herein, we report a rare radiological finding in a 6-year-old boy with right temporal osteolytic lesion due to EG. The aim of this report is to draw attention to a rare cause of fluid-fluid level within a skull lesion on imaging.

Case Report. A 6-year-old boy was admitted to the hospital with a 2 week history of rapidly enlarging right temporal scalp swelling, which was initially tender then became pain-free. He had no associated symptoms and had no other relevant clinical history. No previous history of head trauma was present. Physical examination showed a large, cystic, mildly tender mass in the right temporal region. The overlying skin was normal and at the edges of the mass, a skull defect could be palpated. No fever or neck stiffness was present, and neurological examination was unremarkable. Laboratory evaluation demonstrated normal hematological and biochemical values. The skull x-ray demonstrated a single osteolytic lesion at the temporal bone with lobulated outline without sclerosis (**Figure 1**). A CT scan demonstrated calvarial defect and a subgaleal mass, which have fluid-fluid level (**Figure 2**). The isoattenuating component was presumed to indicate the presence of tumor tissue or liquid blood. The MRI demonstrated a moderate sized bony defect filled with a mass lesion with extradural extension (**Figure 3**). This lesion has low signal intensity on T1WI with fluid-fluid level. There was no evidence of intraparenchymal extension or infiltration, but there was a dural tail enhancement in the post contrast study. A bone scan demonstrated a photopenic area with no uptake of the radiotracer, and no other abnormalities could be detected in the rest of the body. During surgery, an extradural, well encapsulated cystic mass was found underneath the

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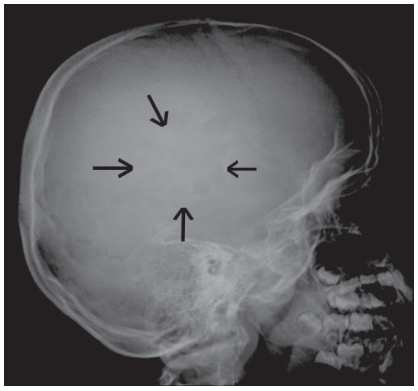

Figure 1 - Skull lateral x-ray demonstrating an osteolytic calvarial defect without sclerosis at the temporal bone.

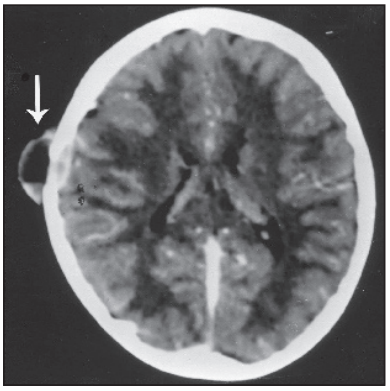

Figure 2 - Brain CT with contrast showing a calvarial defect with bevelled edge, associated with an adjacent subgaleal soft tissue mass and fluid-fluid level.

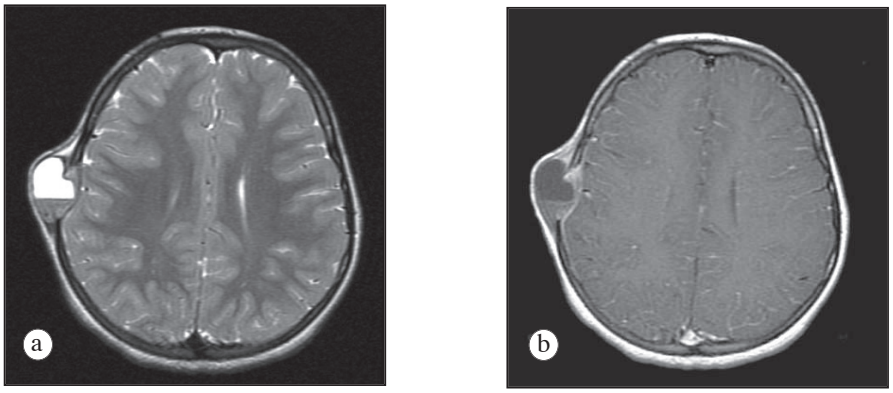

Figure 3 - Axial section MRI brain **a)** T2WI illustrating a cystic lesion at the temporal area, with fluid-fluid level. **b)** Enhanced T1WI illustrating dural tail sign.

periosteum, filling a skull defect and adherent to the underlying dura, however, this could be dissected from the dura and removed totally with ease, and when the mass was opened, altered blood came out. The bone edges of the skull defect were abnormally thinned out. Pathologic examination of the blood materials obtained from the lesion revealed the proliferation of mononuclear cells embedded in the blood pool. These cells had abundant acidophilic cytoplasm and oval indented nuclei. These mononuclear cells were confirmed to be Langerhans cells by the positive immunohistochemistry staining with S-l00 protein. The patient made an uneventful recovery and is on regular outpatient clinic follow up.

Discussion. Eosinophilic granuloma is a disease of unknown etiology, grouped under the term LCH, also known as histiocytosis X, idiopathic inflammatory histiocytosis, or reticuloendotheliosis.[3] Abnormal proliferation of histiocytes in various parts of the reticuloendothelial system such as the bone, lungs, central nervous system, skin, and lymph nodes characterize these diseases.[3] The precise cause and pathogenesis of LCH remains unclear, with several origins suggested; infectious, neoplastic, genetic, metabolic and more recently thought to be a clonal non-neoplastic disorder.[2] Eosinophilic granuloma represents approximately 70% of cases of LCH.[1,4] It most frequently involves the skull, and the natural history is thus far not completely defined.[2,4] It affects children in the first decade of life, with male predominance, and usually presents with aching pain, swelling, low grade fever, elevated sedimentation rate, and peripheral eosinophilia.[3]

The radiographic features of LCH of the skull depend on the phase of the lesion. In the incipient phase, it consists of an osteolytic area with poorly delineated borders and lamellar periosteal reaction, mimicking malignant tumor. Later on, the lesion becomes sharply delineated and a ring of sclerosis may appear, giving it a more benign appearance.[1,5] Uneven destruction of the outer and inner cranial tables may result in a bevelled edge or double contour, that we call, "button sequestrum" or "bull's-eye," representing residual bone, and better seen on CT.[4,6] It is very rare for EG to have a fluid-fluid level in

CT or MRI scans. Beltran[3] once described fluid-fluid level in EG, and ours is the second case report of an EG in the skull with fluid-fluid level on CT scan and MRI. In our case, we found a cystic lesion containing altered blood within its cavity. We suggest that, this could be attributed to spontaneous hemorrhage within the lesion, and possibly explained by the sudden onset of appearance of the scalp swelling, and later on, CT scan showed fluid-fluid level, which is likely to be due to double density with a sedimentation level, or heterogeneity of the hematoma.

Chen et al[2] reported spontaneous hemorrhage of Langerhans cell histiocytosis of the skull complicated with an epidural hematoma. Our patient's head lump presented suddenly and had no history of trauma. We also found his coagulation profile within normal limits. All these findings suggest spontaneous bleeding within the EG of the skull and the radiological appearance of fluid-fluid level.

Davies et al[7] reported the demonstration of a fluid-fluid level within an osseous lesion on CT as suggestive of an aneurysmal bone cyst, although we also rarely find fluid-fluid levels in association with other lesions such as telangiectatic osteosarcoma and a conventional osteosarcoma following chemotherapy. The MRI signals of the lesion are not specific; either for the diagnosis or for the pathological phase of the lesion.[1] The treatment of LCH depends on the extent of the disease. Surgical curettage is the best treatment for single bone lesions. Generally, the prognosis is excellent in limited disease.[2]

In conclusion, solitary EG is uncommon in the skull, and awareness of a rare presentation on CT and MRI with fluid-fluid level is important to know, and to include EG in the list of differential diagnosis. This finding is not a pathognomonic sign of EG; although, we consider that it is a very important sign for spontaneous hemorrhage within the lesion.

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