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Visual Diagnosis in Emergency Medicine

LANGERHANS CELL HISTIOCYTOSIS

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CASE REPORT

A 6-year-old healthy boy presented to the Emergency Department for evaluation of forehead swelling. The patient stated that he had been hit on the forehead while lifting a chair off of a desk 2 weeks prior and subsequently the area had remained swollen. Physical examination revealed a 2.5 × 3.5-cm, well-circumscribed, palpable, nontender, soft-tissue mass of the forehead (Figure 1). A computed tomography scan of the head was subsequently obtained (Figure 2).

Figure 1. Palpable, nontender, soft-tissue mass of the forehead.

DISCUSSION

Langerhans cell histiocytosis (LCH) is a rare condition, caused by an excessive accumulation of immature Langerhans cells and resulting in granuloma formation. The etiology and pathogenesis of LCH are unclear. The incidence of LCH is about one in 200,000 children and is most common in the 2–3-year age range, but can be found in all ages, including adults (1). LCH may affect single or multiple organs. Therefore, the presentation is widely variable and depends on the areas of the body affected. In 78% of cases, one or more locations of bone are involved; most commonly the skull (49%), pelvis (23%), femur (17%), and ribs (8%). Other frequent locations include skin, lung, or lymph node lesions, but LCH can affect every organ (2). The differential diagnosis for bony lesions includes bone cysts, tumors, and trauma. Computed tomography scan of the head will typically demonstrate oval lytic lesions, involving the full thickness of the calvarium, with circumscribed margins and characteristic beveled edges, representing the unequal destruction of the outer and inner tables of the skull. Biopsy confirms the diagnosis of LCH. Because LCH is very rare and the extent and severity of the disease may range from benign to lethal, treatment is widely variable. In some cases, LCH will resolve without any intervention. When treatment is required, resection is the primary therapy for most bony presentations, with excellent results. Bony

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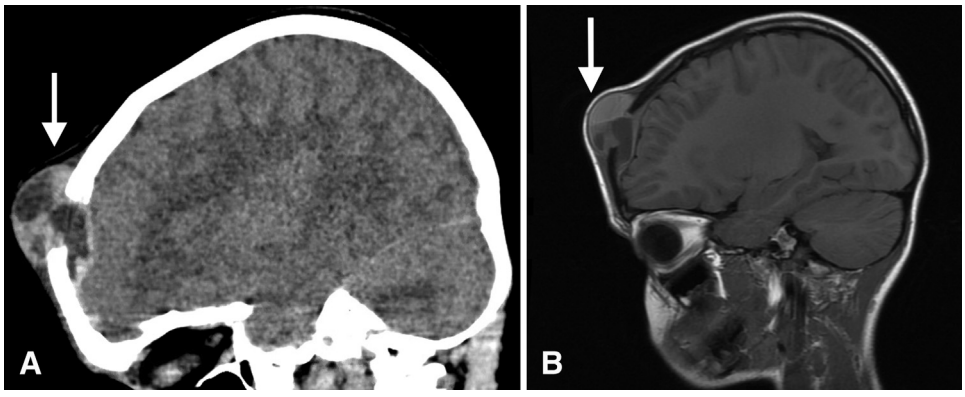

Figure 2. (A) Computed tomography scan of the head demonstrating a lytic lesion of the calvarium, with circumscribed margins, with a soft-tissue component extending intracranially and into the soft tissues extracranially. (B) Magnetic resonance T1 flair image, demonstrating calvarial lesions and isointense gray matter.

recurrence after resection is rare (3). Chemotherapy may be needed for multisystem disease.

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