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CT of Eosinophilic Granuloma of the Skull with Sonographic Correlation

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Eosinophilic granuloma is a benign disorder usually of children and young adults. Although its identity is still controversial, some authors consider this disease a distinct pathologic entity to be differentiated from Letterer-Siwe and Hand-Schüller-Christian disease [1-3]. The skeleton is most commonly affected, but occasionally the skin, lung, and gastrointestinal tract are involved. The skull is the most frequent site of bony involvement, and it is occasionally accompanied by a palpable soft-tissue mass [4]. Although the conventional radiographic findings are well documented, the computed tomographic (CT) appearance has not yet been described. We present the CT and ultrasound findings of two patients with eosinophilic granuloma of the skull.

Case Reports

Case 1

A 20-month-old white male infant in good general health was noted to have a swelling over the left occipital area 2½ months before admission. There was no history of head trauma, wound drainage, fever, or developmental abnormalities. Examination revealed a 2.5 cm, firm, nontender swelling with the scalp movable over the mass. Skull films demonstrated a lytic defect with beveled edges and a questionable soft-tissue mass. The precontrast CT scan (fig. 1A) demonstrated a lytic skull defect with an accompanying extracranial soft-tissue mass. The contrast CT scan (fig. 1B) showed no evidence of enhancement of this soft-tissue mass nor evidence of intracranial involvement. Sonography (fig. 1C) demonstrated an echogenic boundary on the inner aspect of the soft-tissue mass, suggesting an intact dura and absence of intracranial extent. Surgical resection confirmed lack of dural invasion. The histology was eosinophilic granuloma. The patient is currently doing well.

Case 2

A 13-month-old white infant was noted to have a left occipital mass which gradually grew over 6 months. Physical examination revealed a 4 cm, mildly tender, soft-tissue mass. Skull films showed a large lytic lesion with a beveled appearance and a soft-tissue

mass. The CT scan (fig. 2) demonstrated a large lytic defect with a soft-tissue mass exhibiting peripheral contrast enhancement. CT showed no calcium, fat, or intracranial extension. Surgery confirmed lack of dural involvement by the pathologically proven eosinophilic granuloma. The postoperative course was uneventful.

Discussion

In 1893, Hand described a syndrome in a 3-year-old boy with diabetes insipidus, exophthalmus, bronzed skin, hepatosplenomegaly, and poor development, which later became known as Hand-Schüller-Christian disease [5, 6]. Letterer in 1924 and Siwe in 1933 described cases in children characterized by petechiae, anemia, hepatosplenomegaly, multiple osseous lesions, and lymphadenopathy (Letterer-Siwe disease) [7, 8]. In 1940, Lichtenstein and Jaffe [9] described an osseous lesion which they called "eosinophilic granuloma." In 1953, Lichtenstein [1] reviewed all previous histopathologic material and concluded that Hand-Schüller-Christian disease, Letterer-Siwe disease, and eosinophilic granulomas were all inflammatory histiocytosis. He postulated these actually represented a spectrum of the same disease which he named "histiocytosis X." More recently, authors have emphasized the distinct epidemiology and prognosis of eosinophilic granuloma [2, 3].

Eosinophilic granuloma typically occurs in children aged 3-10 years. Less commonly it has been reported in adults; Edeiken and Hodes [10] described a case in a 62-year-old woman. The lesion is usually solitary with bone pain often the presenting complaint.

Common sites of skeletal involvement include the skull, bony pelvis, and spine. In the skull, the temporal bone most frequently is involved [11]. Our two cases showed involvement of the occipital bone. Beveled edges are common on plain film and are usually due to more severe destruction of the outer rather than the inner table of the skull [10]. The CT in our two cases confirms the greater extent of the involvement of the outer table of the skull, which is characteristic but not pathognomonic of eosinophilic granuloma [12].

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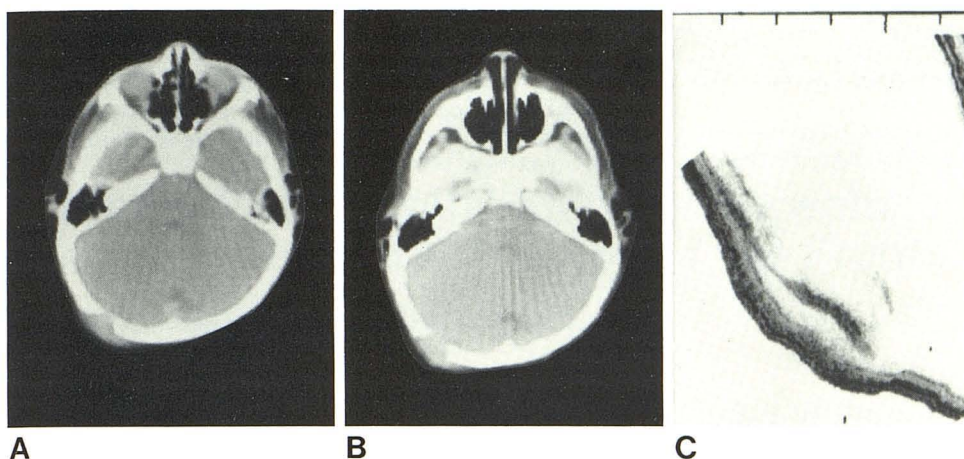


Fig. 1.—Case 1. Eosinophilic granuloma of occipital area. A and B, Base scan. Predominant destruction of outer table of skull with accompanying extracranial soft-tissue mass. C, B-scan sonogram. Anechoic soft-tissue mass with strong acoustic reflections at dural edge.

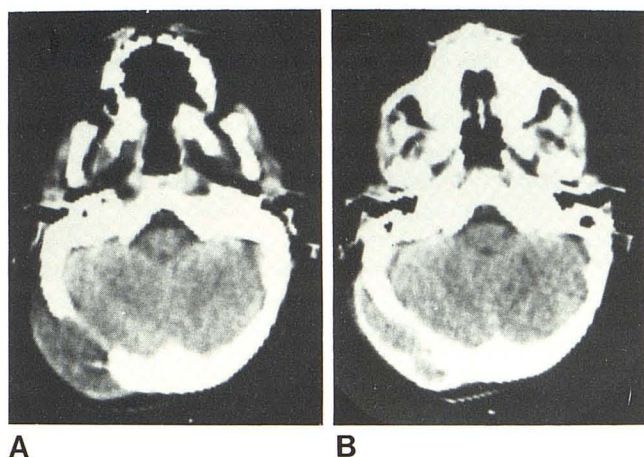


Fig. 2.—Case 2. Eosinophilic granuloma of occipital area. A, Base scan. Predominant destruction of outer table of skull and large accompanying extracranial soft-tissue mass. B, Contrast scan. Enhancement of edges of soft-tissue mass and lack of intracranial extent.

An extracranial soft-tissue mass is common in cases of eosinophilic granuloma of the skull, but is demonstrated with difficulty on plain radiographs [4, 10]. In addition, with plain films a soft-tissue mass may be appreciated with other entities such as a metastatic deposit, lipoma, encephalocele, and dermoid cyst [13–15]. On the other hand, CT is helpful in evaluation and differential diagnosis of the soft-tissue component of eosinophilic granuloma of the skull. Our cases showed the soft-tissue component to be better demonstrated on CT, using appropriate window settings, than on plain film. CT also showed lack of calcium and fat, a helpful point to exclude other lesions such as dermoids or lipomas. CT contrast enhancement of the edges of the soft-tissue component of eosinophilic granuloma may also be seen (case 2). Theoretically enhancement may occur with other soft-tissue tumors of the head, and we have personally seen it occur with metastatic lesions of the bony calvarium. This phenomenon was not described in two other series

dealing with the CT appearance of calvarial metastases [15, 16].

Eosinophilic granuloma of the skull may not always be localized to the scalp and bony calvarium, but may extend to the dura and rarely into the brain substance [2, 10, 14]. In this regard, the contrast enhancement may be helpful in outlining the extent of the isodense part of the soft-tissue and showing presence or lack of intracranial involvement. Sonography (fig. 1C) may also be helpful in showing the extent of soft-tissue mass in eosinophilic granuloma.

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