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CASE REPORT

Skull fracture mimicking eosinophilic granuloma

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Abstract
Background Delayed swelling after skull fractures is an uncommon complication following head trauma in children. Classically, growing skull fractures typically present in patients under 3 years of age with progressive subcutaneous fluid collections, or occasionally with neurologic symptoms. We present the case of a healthy 2-year-old boy with a lytic "punched-out" frontal skull lesion. The child presented 2 months after a minor forehead injury for which no medical attention was sought.
Methods The skull defect had no associated leptomeningeal cyst or brain herniation. Imaging and presentation were thought to be consistent with eosinophilic granuloma. Histologic findings demonstrated a healing skull fracture.
Results Cranioplasty was performed, and the patient had an uncomplicated postoperative course.
Conclusions In this report, we describe our experience with this atypical presentation of a healing skull fracture mimicking a typical eosinophilic granuloma.

Keywords Children · Eosinophilic granuloma · Skull fracture

Introduction

Increasing swelling arising in a delayed manner weeks after head injury is an uncommon complication pediatric skull fractures. One common mechanism for this phenomenon is a posttraumatic leptomeningeal cyst, also called a "growing skull fracture." These lesions occur most frequently in children, with an estimated incidence of 0.03–1.6 % following skull fracture [1–3]. Over 90 % of cases occur in children under 3 years of age, and greater than 50 % occur in patients under 1 year old [4]. The classic presentation in a young child includes a subgaleal fluid collection overlying a diastatic parietal skull fracture resulting from head trauma in the setting of progressive neurologic deficit and/or posttraumatic seizures [5, 6]. Plain skull films demonstrate smooth-edged ossification defects with scalloping and often sclerotic margins. Computed tomography (CT) shows similar skull abnormalities in addition to underlying parenchymal injury with cystic degeneration. A leptomeningeal cyst or brain tissue will often herniate through the skull defect [5].

Clinical presentation and imaging characteristics are generally sufficient to diagnose growing skull fractures. In this report, we describe the case of a 2-year-old boy with the appearance of a lytic skull lesion on CT that, despite presentation consistent with eosinophilic granuloma (EG), histologic findings confirmed the diagnosis of a healing skull fracture.

Case report

History and presentation

A 2-year-old healthy male presented by referral for a skull lesion. Two months prior, another child struck the patient on his left forehead. His mother noticed mild bruising in the area immediately following the incident. The family did not seek medical attention at the time and the bruising resolved within several days following the injury. No imaging of the skull was performed at the time of the trauma. Several weeks later, the child developed a bony step-off and palpable soft tissue mass

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in the general area. The patient had no history of seizure or neurologic deficits before or after the injury.

On physical examination, the patient was neurologically intact. There was a tender, intermittently pulsatile mass on the left forehead without overlying redness or swelling. The mass was palpable within a skull defect that had irregular borders.

Head CT revealed a soft tissue mass within a lytic skull defect with irregular eroded borders measuring 2 cm in diameter (Figs. 1 and 2). The lesion affected the outer table more so than the inner table, and had a beveled appearance. There was no evidence of surrounding sclerosis or periosteal reaction. Based on clinical presentation and imaging, the most likely diagnosis was thought to be a solitary calvarial EG.

Operation

The patient was taken to the operating room for surgical resection. Intraoperatively, an area of abnormal skull was identified in the left frontal bone but no linear fractures were seen. A 2-cm circumferential craniectomy using a high-speed drill and rongeurs was performed around the lesion for complete resection. Abnormal bone and underlying fibrous tissue were sent to surgical pathology. There was no dural defect under the skull lesion, and the dura was not violated during the procedure. Bone dust was collected and used to fill the craniectomy site. An absorbable plate was cut and fashioned to repair the skull defect. The surgical wound was closed in the standard layered fashion, and a sterile dressing was applied.

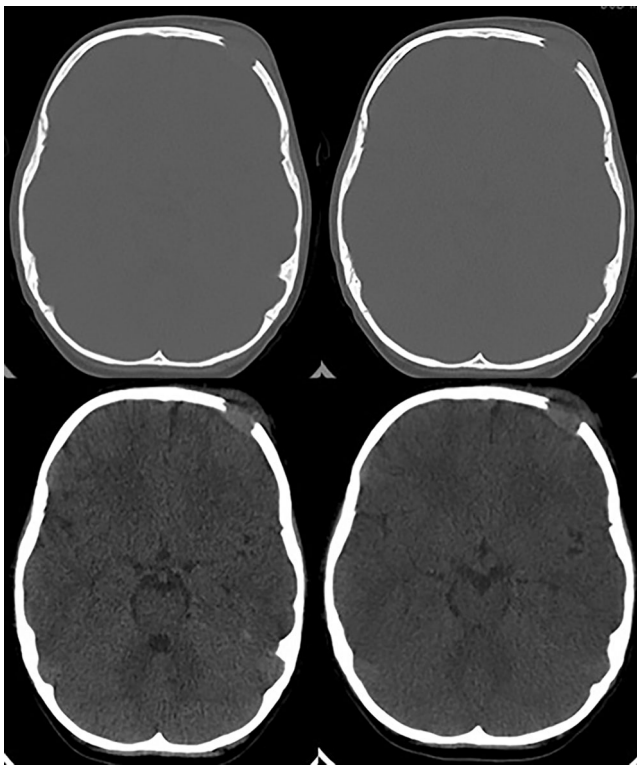

Fig. 1 Preoperative non-contrast head CTs, with axial bone (*upper*) and brain (*lower*) windows. Imaging demonstrates a lytic skull lesion with beveled borders, the typical appearance for EG. Note the fracture is non-diastatic, and there is no cyst or brain herniation through the defect

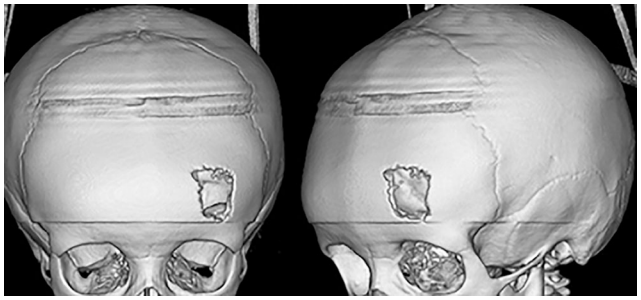

Fig. 2 Three-dimensional CT skull reconstructions. Skull defect is visible over the left frontal bone

Pathologic findings

Microscopic features was evaluated using hematoxylin and eosin (H&E) staining, and CD34 immunohistochemistry showed fragments of woven and lamellar bone with fibrosis and granulation tissue in marrow spaces consistent with healing fracture. Structural features included hemosiderin-laden macrophages, fibrous tissue, and reparative woven bone. Osteoblastic rimming was seen and is typical of bone repair, but not fibrous dysplasia. Eosinophils and Langerhans cell histiocytes, squamous epithelium, and adnexa were not present. Immunohistochemistry was negative for cluster designation 1A (CD1A), K903 cytokeratin, and S-100 protein staining; these findings ruled out EG, epidermoid cyst, and dermoid cyst (Fig. 3) [7].

Other diagnoses are associated with bone repair, but few show a focal lytic skull lesion in this age group. For example, active Paget disease makes woven bone, but it does not affect children. Fractures associated with unicameral (simple) bone cysts are rare in the skull. Features of simple bone cyst fracture include bone repair. In this 2-year-old patient, the injury preceded growth of the lesion, consistent with a growing skull fracture. Aneurysmal bone cysts have a rim of bone repair, but this case does not have other features of an aneurysmal bone cyst, such as spaces with serum or blood, septa with fibro-osteoid, or calcified matrix. Slides were reviewed by two pathologists with both coming to the same diagnosis of a healing skull fracture.

Postoperative course

Patient was discharged following surgery without complication. The incision healed well without underlying mass or bony deformity. He remained neurologically intact throughout with no seizures. Head CT at 6-month follow-up showed a healing skull defect (Fig. 4).

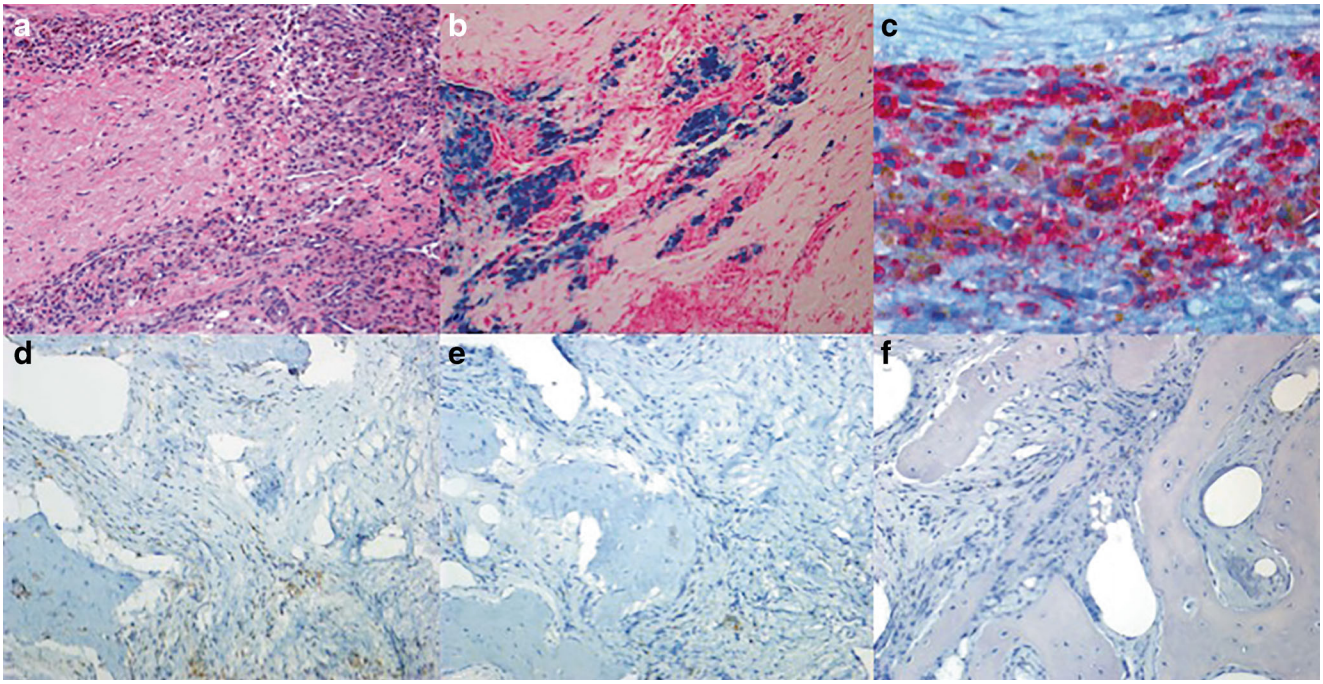

Fig. 3 **a** Lesion in the dura shows large numbers of phagocytes with plump cytoplasm containing variable amounts of brown pigment (H&E stain; original magnification, ×20). **b** The pigment in the cytoplasm stains blue, a characteristic feature of hemosiderin. While some hemosiderin granules are scattered in fibrous tissue, hemosiderin is not seen in pink nuclei (Gomori stain for iron; original magnification, ×20). **c** Immunohistochemistry for CD68 marker stains macrophages red, but does not alter the natural brown color of hemosiderin granules. Hematoxylin stains collagen blue and nuclei dark blue (CD68 with hematoxylin counterstain; original magnification, ×60). **d** Skull lesion shows no evidence of S100 protein, a marker for EG. Reparative woven bone lacks laminations of lamellar bone, and has hypertrophic osteocytes in lacunae. Reactive fibrosis and macrophages contain a few granules of hemosiderin (S100 protein with hematoxylin counterstain; original magnification, ×20). **e** There is no evidence of CD1a, another marker for EG. Reactive fibrosis and macrophages contain many granules of hemosiderin (CD1a with hematoxylin counterstain; original magnification, ×20). **f** Cytokeratin marker CK903 for epidermoid/dermoid cysts is negative (CK903 with hematoxylin counterstain; original magnification, ×20)

Discussion

To our knowledge, this is the first description of a healing skull fracture mimicking an EG in a pediatric patient. The literature contains two reports of adults with a distant history of head trauma (20 and 50 years) with growing skull fractures mimicking skull tumors [8, 9]. Our case is unique because of its atypical presentation for a growing skull fracture with multiple radiographic and clinical findings consistent with EG.

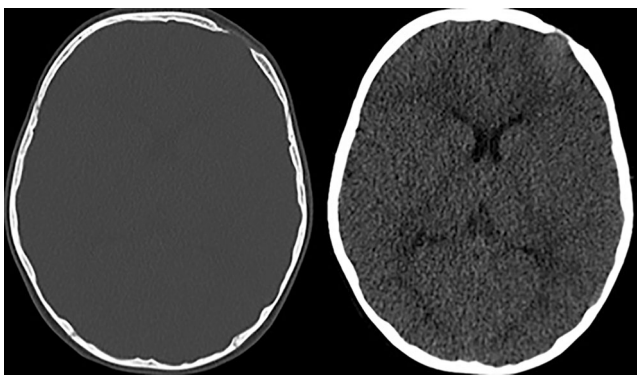

Fig. 4 Postoperative non-contrast head CTs, with axial bone (*left*) and brain (*right*) windows. Skull defect edges demonstrate new bone formation. Absorbable plate can be visualized over the defect

In some cases, the phenomenon of a "growing skull fracture" is thought to arise from a tear in the dura underlying a skull fracture that results in herniation of the arachnoid membrane. This herniation produces a leptomeningeal cyst that, through cerebrospinal fluid pulsations and continued pressure on the overlying fracture, leads to erosion of bone edges [10]. More recent evidence implicates the herniation of brain parenchyma itself through the skull defect. Continued underlying tissue injury causes brain necrosis with reactive gliosis leading to cystic degeneration [11–13]. This pathophysiology is more consistent with the progressive neurologic deficits and seizures often seen in these patients [4, 6].

Our patient was neurologically intact and had no dural defect, leptomeningeal cyst, or brain herniation through the skull fracture. The skull defect was non-diastatic with a "punched-out" lytic appearance and beveled edges located in the frontal bone, an uncommon location for growing skull fractures [5, 6]. These imaging features are typical for EG [14]. Patients diagnosed with EG have a history of trauma to the area over the tumor in 33–50 % of cases [15–18]. The strong association between local head trauma and EG has

led authors to question this as coincidental [19]. These findings indicate that a history of trauma does not significantly aide in establishing a diagnosis of growing skull fracture versus EG.

EG is a local manifestation of Langerhans cell histiocytosis (LCH), often referred to as unifocal LCH. EG constitutes 21–32 % of all primary pediatric skull tumors with epidermoid/dermoid cysts, fibrous dysplasia, and vascular anomalies being other common diagnoses [20, 21]. Our case exemplifies the fact that EG may be mistakenly diagnosed, given the greater incidence of EG compared to growing skull fractures, even in the setting of previous head trauma.

We present this case as an example of an atypical presentation of a healing skull fracture and as a cautionary note on the early surgical management of presumed EG in patients with preceding head trauma. Watchful waiting is an acceptable treatment option in solitary calvarial EG and growing skull fractures. Spontaneous resolution of both lesions has been described [22–24]. A period of observation may be indicated to make a definitive diagnosis and establish appropriate treatment.

References

1. Lende RA, Erickson TC (1961) Growing skull fractures of childhood. J Neurosurg 18:479–489
2. Arseni C, Simionescu MD (1966) Growing skull fractures of childhood. A particular form of posttraumatic encephalopathy. Acta Neurochir (Wien) 15:159–172
3. de P Djientcheu V, Njamnshi AK, Ongolo-Zogo P, Kobela M, Rilliet B, Essomba A, Sosso MA (2006) Growing skull fractures. Childs Nerv Syst 22:721–725
4. Tandon PN, Banerji AK, Bhatia R, Goulatia RK (1987) Craniocerebral erosion (growing fracture of the skull in children). Part II. Clinical and radiological observations. Acta Neurochir (Wien) 88:1–9
5. Ersahin Y, Gülmen V, Palali I, Mutluer S (2000) Growing skull fractures (craniocerebral erosion). Neurosurg Rev 23:139–144
6. Pezzotta S, Silvani V, Gaetani P, Spanu G, Rondini G (1985) Growing skull fractures of childhood. Case report and review of 132 cases. J Neurosurg Sci 29:129–135
7. Kobayashi TK, Ueda M, Nishino T, Bamba M, Echigo T, Oka H, Hino A, Fuse I, Fujimoto M, Katsumori T, Kaneko C (2007) Langerhans cell histiocytosis of the skull on cytologic squash preparations. Diagn Cytopathol 35:154–157
8. Kurosu A, Fujii T, Ono G (2004) Post-traumatic leptomeningeal cyst mimicking a skull tumour in an adult. Br J Neurosurg 18:62–64
9. Weinberg JS, LeRoux PD, Panasci D, Weiner HL (1999) Adult growing skull fracture mimicking a skull tumor. Acta Neurochir (Wien) 141:545–546
10. Taveras JM, Ransohoff J (1953) Leptomeningeal cysts of the brain following trauma with erosion of the skull; a study of seven cases treated by surgery. J Neurosurg 10:233–241
11. Ito H, Miwa T, Onodra Y (1977) Growing skull fracture of childhood with reference to the importance of the brain injury and its pathogenetic consideration. Childs Brain 3:116–126
12. Roy S, Sarkar C, Tandon PN, Banerji AK (1987) Cranio-cerebral erosion (growing fracture of the skull in children). Part I. Pathology. Acta Neurochir (Wien) 87:112–118
13. Muhonen MG, Piper JG, Menezes AH (1995) Pathogenesis and treatment of growing skull fractures. Surg Neurol 43:367–373
14. D'Ambrosio N, Soohoo S, Warshall C, Johnson A, Karimi S (2008) Craniofacial and intracranial manifestations of langerhans cell histiocytosis: report of findings in 100 patients. AJR Am J Roentgenol 191:589–597
15. Fox JD, Knighton RS (1956) Diagnosis and treatment of eosinophilic granuloma of skull. J Am Med Assoc 162:1294–1297
16. Arseni C, Dănăilă L, Constantinescu A (1977) Cranial eosinophilic granuloma. Neurochirurgia (Stuttg) 20:189–199
17. Baker GS, Fisher RG (1948) Eosinophilic granuloma of the skull; report of four cases, with 5-year follow-up. Surg Clin N Am 28:1057–1070
18. Rawlings CE, Wilkins RH (1984) Solitary eosinophilic granuloma of the skull. Neurosurgery 15:155–161
19. Mut M, Cataltepe O, Bakar B, Cila A, Akalan N (2004) Eosinophilic granuloma of the skull associated with epidural haematoma: a case report and review of the literature. Childs Nerv Syst 20:765–769
20. Yoon SH, Park SH (2008) A study of 77 cases of surgically excised scalp and skull masses in pediatric patients. Childs Nerv Syst 24:459–465
21. Gibson SE, Prayson RA (2007) Primary skull lesions in the pediatric population: a 25-year experience. Arch Pathol Lab Med 131:761–766
22. De Angulo G, Nair S, Lee V, Khatib Z, Ragheb J, Sandberg DI (2013) Nonoperative management of solitary eosinophilic granulomas of the calvaria. J Neurosurg Pediatr 12:1–5
23. Oliveira M, Steinbok P, Wu J, Heran N, Cochrane D (2003) Spontaneous resolution of calvarial eosinophilic granuloma in children. Pediatr Neurosurg 38:247–252
24. Sekhar LN, Scarff TB (1980) Pseudogrowth in skull fractures of childhood. Neurosurgery 6:285–289