An unusual pediatric case of an intradiploic epidermoid cyst with cranial bone invasion

Michael Weingarten, BA, Mary Beth Gadarowski, MD, Stephanie Zyck, MD, George Koutsouras, DO, Timothy Richardson, DO, Kalliopi Petropoulou, MD, Satish Krishnamurthy, MD, and Ramsay S. Farah, MD
Syracuse, New York

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INTRODUCTION

Epidermoid cysts are benign and slow-growing lesions that are usually less than 2 cm in diameter and do not affect the overlying skin.[1] Characterized by a stratified squamous epithelial wall, they often feature a central punctum and are freely mobile on palpation.[2] Although relatively common elsewhere in the body, they rarely arise from the central nervous system, with an incidence of 0.2% to 1.8% of all brain tumors.[3] The majority of central nervous system epidermoid cysts are observed in adults, with a mean age of presentation of 40 years.[4] These lesions are thought to arise from inadvertent inclusion of ectodermal elements during neural tube closure, trapping them in the diploe, meninges, scalp, or suture lines.[5] Intradiploic cysts may also occur secondary to trauma.[6] Regardless of the mechanism underlying their formation, central nervous system epidermoid cysts are overwhelmingly intradural and, even when located in the intradiploic space, rarely perforate bone.[7] Here, we present a pediatric case of an intradiploic epidermoid cyst with cranial bone invasion.

Abbreviation used:

CT: computed tomography

CASE REPORT

An active, well-appearing, right-handed, 7-year-old boy presented to an outpatient pediatric office for a swelling on his head. There was no trauma history aside from a distant and isolated episode in which the child had accidentally hit his head. The parents had noted an apparent swelling of the left frontal aspect of his skull that had become more prominent. They denied any pain, previous swelling, bruising, or overlying skin changes. Medical history was significant for septic arthritis of the ankle and a Colles fracture requiring surgical intervention. Developmental milestones were appropriately met, vaccinations were up to date, and family and social history were noncontributory.

Clinical examination revealed that the skin overlying the lesion was normal. The lesion was fixed to the bone and the scalp was movable over it. A radiograph of the skull and a computed tomography (CT) scan of the head with contrast were obtained (Figs 1 and 2). These revealed a lytic lesion centered in the left parietal diploic space, with scalloped and well-demarcated margins. Extensive remodeling of the outer table and a smaller area of completely remodeled inner table was noted. The lesion, measuring approximately 0.9 × 1.2 cm, invaded the epidural space where the inner table had been remodeled. There was no ventriculomegaly or abnormality of the brain parenchyma or meninges (Fig 2). A presumptive diagnosis of Langerhans cell histiocytosis was made. The boy was promptly referred to the pediatric neurosurgery clinic.

At neurosurgical consultation, no neurologic deficits were noted. The palpable left frontal lesion measured 2.5 cm in its greater diameter and appeared to be attached to the skull. No pain, tenderness, or fluctuation was elicited. No induration, erythema, or pigment changes were appreciated.

From the State University of New York Upstate Medical University, Syracuse.

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Correspondence to: Ramsay S. Farah, MD, 2058 Upstate Health Care Center, 90 Presidential Plaza, Syracuse, NY 13202. E-mail: farahrs@upstate.edu.

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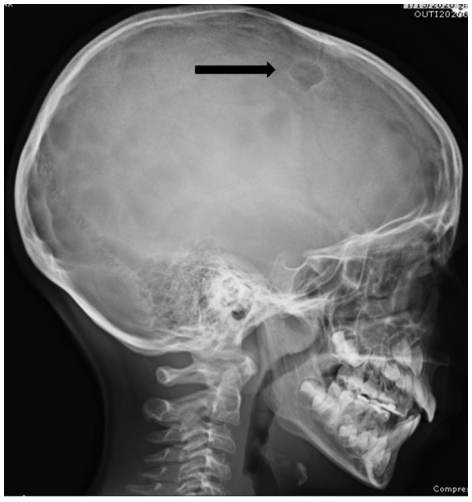

Fig 1. Lateral radiographic view of the skull, demonstrating a lytic lesion (arrow).

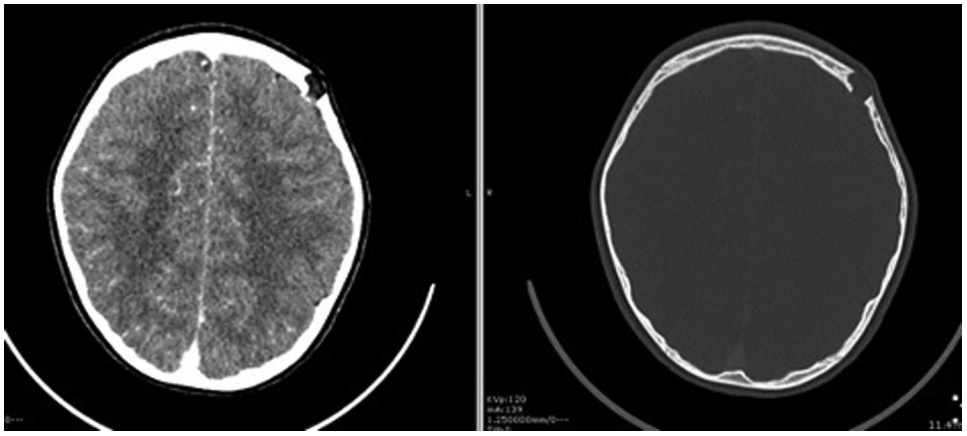

Fig 2. CT scan demonstrating a 0.9 × 1.2 cm lytic lesion with well-demarcated, scalloped margins centered in the left parietal diploic space, with remodeling of the outer and inner table of the calvarium. *CT*, Computed tomography.

The patient was scheduled for a craniectomy with excision of the skull lesion. At surgery, complete excision of the lesion was performed. It was noted to be yellow and rubbery, without invasion of the underlying dura. A titanium plate was placed to close the bony defect and the patient recovered without any complications. Gross specimen neuropathology described a 1.5 × 1.5 × 0.5 cm, tan-white, ovoid, partially firm, and partially softened portion of bone. The final neuropathology report revealed fragments of nonneoplastic bone and squamous epithelium with abundant keratin debris, most consistent with an epidermoid cyst (Figs 3 and 4). Given the benign nature of the lesion, without evidence of malignant degeneration, no additional treatment was necessary. The patient recovered without complications and, to date, no evidence of recurrence after surgery.

DISCUSSION

Epidermoid cysts compose less than 2% of all brain tumors.[3] If present in the central nervous system, 90% are found intradurally, illustrating the unusual finding of our patient's intradiploic lesion.[7] When located in the intradiploic space, they are most commonly found, in descending order of frequency, at the cerebellopontine angle, the fourth ventricle, and the middle cranial fossa.[6] Only a small subset of cases have presented as an asymptomatic bony lump with underlying erosion of the skull.[6] Our patient's intradiploic epidermoid cyst that measured 2.54 cm is one of the largest that has been described in a young child, to our knowledge.

Given the benign nature of epidermoid cysts, various theories have been offered as the mechanism

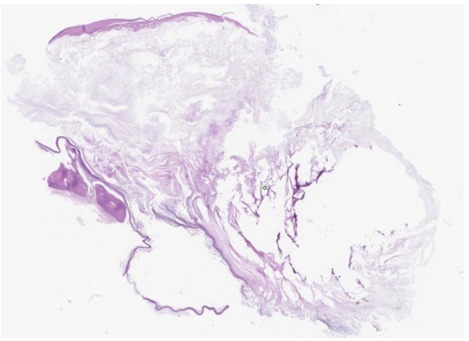

Fig 3. Histologic specimen, hematoxylin-eosin stain, low magnification. Fragments of nonneoplastic bone and squamous epithelium with abundant keratin debris, consistent with a benign epidermoid cyst.

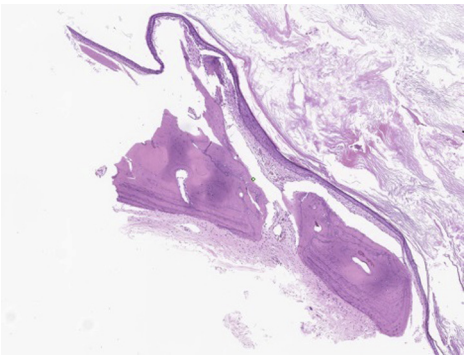

Fig 4. Histologic specimen, hematoxylin-eosin stain, high magnification. Fragments of nonneoplastic bone and squamous epithelium with abundant keratin debris, consistent with a benign epidermoid cyst.

driving any underlying bone erosion. Ambo et al[8] suggested that during a long period, the weight of a large epidermoid cyst could exert continuous pressure on the underlying bone, resulting in its degradation through a process known as scalloping. They argued that the presence of bony sclerosis bordering the bony defect on imaging studies is highly suggestive of this phenomenon.

Pediatric cases of epidermoid cysts with underlying bone erosion challenge this assertion. In the case noted by Ambo et al,[8] the patient was a middle-aged woman with a large (3 cm) epidermoid cyst that had slowly eroded the underlying bone during a 10-year period. Could a smaller lesion with similar diagnostic imaging in a pediatric patient with minimal trauma to the area, as in our case, truly be caused by scalloping? This seems unlikely, given the relatively short period of its exerting continuous pressure on the underlying bone. Although it is plausible that the scalloping process may be expedited in the pediatric patient because of lack of complete cranial bone ossification or be hastened by even minimal trauma to the area, this remains unknown and necessitates more thorough investigation.[9]

Our case highlights the diagnostic challenges of pediatric intradiploic epidermoid cysts. Given their rarity and the plethora of differential diagnoses, a keen-eyed clinician and subsequent alignment of the radiologic examination, imaging, and pathologic analysis are paramount. Although CT imaging is avoided when possible in pediatric patients because of the risk of radiation exposure, it is superior to magnetic resonance imaging for evaluating bony lesions. Radiography of the skull is useful as a screening study. However, once a bony lesion is identified, CT with contrast provides further detail to evaluate for brain parenchymal invasion, assess for meningeal enhancement, and plan surgical treatment. Surgical excision is ultimately mandated because epidermoid cysts have undergone malignant transformation in some cases.[10] Regardless of differing treatment courses, one thing is quite clear: the exact pathogenesis and behavior of epidermoid cysts in the central nervous system remain largely a mystery.

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