Intraosseous Dermoid Presenting as an Expansile Lytic Lesion

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Abstract
Cranial dermoids have the tendency to occur in the midline, especially near fontanelles and sutures early in the life of a patient. Here we present an unusual case of an intraosseous dermoid that presented initially as a lytic lesion, off of the midline and not associated with cranial sutures or fontanelles. The diameter of the lesion grew to approx 15 mm over time, thus the decision was made to take the child to surgery for removal of dermoid with the use of neuronavigation and cranioplasty. A dermoid cyst was confirmed on histopathologic analysis.

Introduction
Dermoid cysts are uncommon soft tissue lesions composed of squamous, epithelium-lined sacs.1 The cyst develops during gestation and is generally characterized by a midline location. The wall of the cyst contains sweat glands and sebaceous glands that secrete fluid into the cyst cavity. This causes a hyperintense signal in T1-weighted imaging.2,3 Clinically, it presents as a raised lump or mass. We report an atypical case of a patient with an intraosseous dermoid cyst, presenting as a lesion noticed as a raised lump and located off the midline.

Case Report
A 2-month-old female presented with a fluid collection within the deep scalp appearing to overlie a calvarial defect with possible intracranial extension visualized on ultrasound. At approximately 7 months of age, the patient underwent a head computed tomography (CT), which revealed a chronic appearing, small calvarial defect in the inferior right frontal region immediately anterior to the coronal suture (►Figs. 1 and 2). It measured approximately 6 mm in greatest dimensions. There were beveled sclerotic margins, and an associated defect appeared to extend through the inner table of the right frontal bone. At 18 months, a repeat head CT revealed the lesion had expanded, demonstrating increased bony remodeling (►Figs. 3 and 4). The diameter of the lesion grew to approximately 14 to 15 mm. It was decided to excise the lesion due to progression in size. The patient underwent resection of the lesion by supratentorial craniectomy with use of neuronavigation and cranioplasty. The mass was removed in a piecemeal fashion until the entire visible portion was removed. The presence of cottage cheese-like material emanated from the diploic spaces of bone, a finding generally consistent with a dermoid cyst, which was confirmed on histopathologic examination. The craniotomy defect measured approximately 12 × 17.5 × 20 mm after the mass was removed. Cranioplasty was then performed by applying autologous bone chips to the surface of the dura and overlaying them with a moldable demineralized bone putty.
Discussion

Dermoid cysts are uncommon, slow-growing congenital lesions. Their differential diagnosis includes dermoid cyst, cavernous hemangioma, eosinophilic granuloma, Langerhans cell histiocytosis, and fibrous dysplasia. Pediatric patients are the main population affected by dermoid cysts, as most develop during gestation and become symptomatic early in life. This occurs in response to entrapment of the ectoderm surface along the lines of embryonic fusion. The lesion commonly appears superficially along the cranial sutures or anterior fontanelle. They are benign but produce keratin and sebaceous material that is deposited into the cyst cavity. This results in the formation of an oily mixture inside the cyst. As secretion continues, increasing intracystic pressure can result in mass effect or rupture. Consequently, this heightens the risk of cranial erosion and expansion in the epidural space.

Dermoid cysts can occur in many locations of the body, but cysts of the head and neck are commonly located at the frontotemporal or brow region. They often present as a benign, painless lump under the scalp. Occasionally, they are noticed as a palpable bony defect, as in this case. Pryor et al performed a retrospective review of pediatric patients presenting with dermoid cysts of the head and neck and found that 61% of the cysts were located in the periorbital region. Due to its close proximity to the orbital region, cysts found in this area can cause symptoms of headache, proptosis, visual disturbances, and development of extradural hematomas. Only 10% of patients presented with cysts located in the...
skull. This cyst presented in the right frontotemporal region of the patient’s calvarium and was unusual because it was not in the midline. Additionally, it was intradiploic, a rare subtype of frontotemporal dermoid cysts accounting for only 0.4 to 0.7% of all cranial masses. An indication that the cyst arose from the diploe is if the local dura remains unaffected in addition to eroding inner and outer tables of the skull.

Discloser
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References