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Case Report

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Desmoplastic Fibroma of the Skull in an Infant: A Case Report

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Abstract

Desmoplastic fibromas are rare benign bone tumors occurring primarily in long bones and mandible. In this case report, we present a desmoplastic fibroma originating from the left frontal bone. This is an exceptionally rare presentation of this pathology and the associated imaging and pathologic slides are highly educational. We discuss the relevance to the literature and how to manage these patients clinically.

Keywords: Desmoplastic Fibroma; Pediatric Skull Tumors; Clinical Pearls

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Introduction

Desmoplastic fibromas are benign bone tumors that can be locally aggressive [1]. They typically present in long bones and along the mandible [2]. Cranial desmoplastic fibromas are exceedingly rare. The tumor is histologically and biologically similar to fibro-mitosis or desmoids [3]. In the English literature, only 19 desmoplastic fibromas have been reported for cranial bones [4]. Only two of these cases were in children and only one deemed congenital [5]. In this case report we present a 10-month-old patient with a desmoplastic fibroma arising in the left frontal bone. We review previously published cases and highlight the uniqueness of a desmoplastic fibroma originating within the cranium. The aim of this report is to discuss the clinical, radiological, and histological characteristics for optimal treatment of this rare aggressive skull tumor.

Case Report

A 10-month-old female infant was brought to the pediatric office due to swelling on the left side of her head. The swelling had slowly been enlarging for 1 month. The mother of child reported no history of trauma and no deficits in neurologic exam were noted. On local exam of the region, the lesion was non-painful, non-tender, firm, and consisted of 3 by 3 cm swelling in the left frontal region. Laboratory studies were all within normal limits. Chest x-ray and abdominal ultrasound were also unremarkable (Figure 1).

The mass was in the intradiploic space of the left frontal bone. Both the outer and inner tables of the calvarium were thinned. This indicates that the mass originated from inside the bone rather from the dura. The radiographic diagnosis at time of presentation was eosinophilic granuloma (Figure 2).

The infant underwent surgery to remove the mass. A frontal craniotomy was performed with excision of tumor involving the bone. The mass was approximately 3 cm in diameter and involved the outer

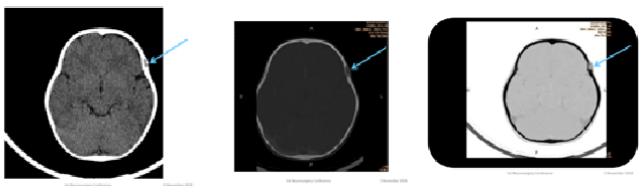

Figure 1: Panel 1, 2, and 3 show a CT scan with an expansile osteolytic mass that the thins the underlying calvarium and disrupts the outer table of the left frontal bone but does not extend cranially.

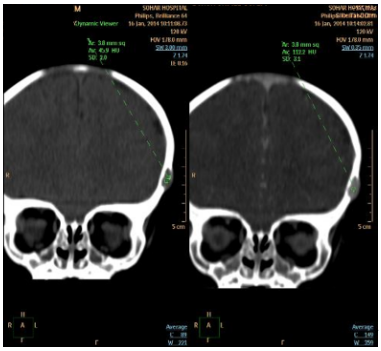

Figure 2: Coronal bone-window of the initial CT scan showed an osteolytic mass eroding the outer table.

cortex. The excision did not involve the inner cortex. The mass was completely excised with gross total resection. Hemostasis was achieved with cautery and bone wax (Figure 3).

Pathological examination of the lesion identified desmoplastic fibroma of the cranium. The lesion was sent en bloc that appeared white with measurements of 2 by 2 by 0.5 cm. Histologically, the lesion was composed of a proliferation of fascicles of spindly appearing cells with bland appearing elongated nuclei that were set in collagenous stroma. A few blood vessels were visible on histology. Occasional extravasated RBC's were noted. There was no associated mitosis, eosinophil proliferation, curvilinear bone trabeculae, or necrosis. Tumor cells were not immunoreactive to CD34, S100, or desmin. It did show positive immunoreactivity for SMA, vimentin, and β catenin as seen in Table 1 (Figure 4 and Figure 5).

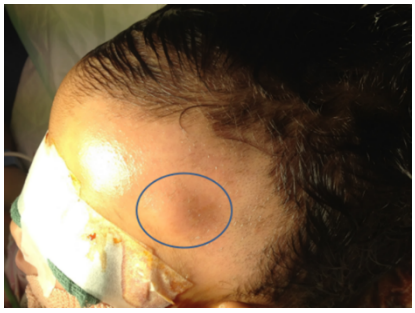

Figure 3: Gross visualization of the lesion prior to excision.

IHC	Non ossifying Fibroma	Desmoplastic Fibroma	Fibrous Dysplasia	Low grade Fibrosarcoma
SMA	-ve	+ve	-ve	-ve
Vimentin	-ve	+ve	-ve	+ve
B catenin	-ve	+ve	-ve	-ve

Table 1: Common histopathologic findings associated with bone tumors of the cranium.

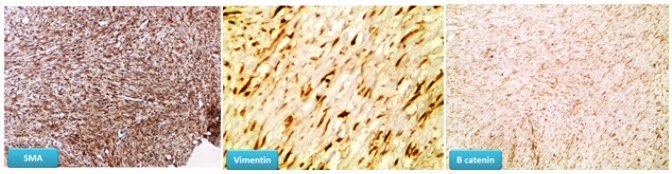

Figure 4: Positive SMA, Vimentin, and β Catenin staining consistent with desmoplastic fibroma.

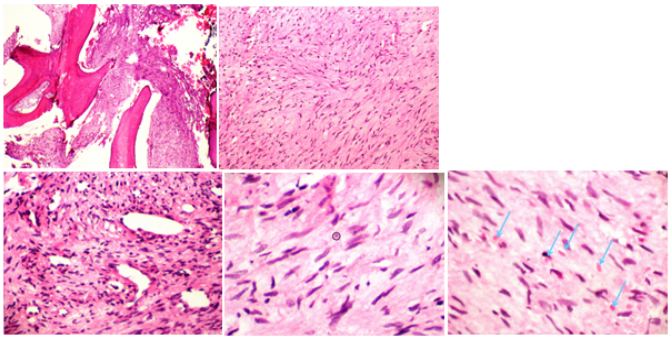

Figure 5: H&E low and high power histology of tumor.

Discussion

Initial differential diagnosis based on imaging and presentation included non-ossifying fibroma, desmoplastic fibroma, fibrous dysplasia, and low-grade fibrosarcoma [6]. A review of the characteristics of common bone pathologies is described in Table 2.

Variables	Non ossifying Fibroma	Desmoplastic Fibroma	Fibrous Dysplasia	Low grade Fibrosarcoma
Gender	M:F is 1:1	M:F is 1:1 With sight male predominance.	M:F is 1:1	M:F is 1:1
Age	2nd Decade	2nd and 3rd Decades	First 2 decades	All ages Rare in 1st decade
Location	Metaphysis of axial	Mandible Pelvis and axial	No specific site of involvement	Axial and mandible
Radiographic features	Eccentric Metaphysis	Expansible with cortical destruction and ST extension	Expansible with sclerotic rim Ground glass	Large, purely lytic metaphyseal or diaphyseal
Gross	Soft yellow to tan	Non specific	More gritty	No specific, haemorrhgae, necrosis
Microscopic	Fibrous, storiform	Fibrous, collagen, haphazard	Fibrous with irreg. bone trabeculae	Gradable malignant
Atypia and mitosis	———	———	———	G1: 1-4 G2&G3: numerous
New Bone	———	———	++++++	———

Table 2: Common characteristics associated with bone tumors.

Desmoplastic fibroma is an extremely rare neoplasm. It constitutes 0.3% of benign bone tumors and 0.06% of cranial bone tumors [7]. They usually present in patients younger than 30 years old [8]. The radiographic appearance can mimic many much more common bone neoplasms. The lesion is typically iso or hypodense [9]. 29% have cortical breakthrough with destruction of the outer table and intradiploic space [10]. Therefore these lesions can mimic other lytic pathologies than can invade the scalp or dura [11]. Typically they present as a lump on the head and occasional with hearing loss if temporal bone is involved [12].

Differential is broad and can include eosinophlic granulomas, cavernous hemangiomas, fibrous dyplasia, fibrosarcoma, chondromyxoid fibromas [13]. Other types of pathologies that present similarly on radiographic imaging include giant cell tumors, simple bone cysts, aneurysmal bone cysts, chondrosarcoma, meningioma, and metastasis [14]. Therefore, getting adequate tissue for histopathology is essential. It is histologically similar to desmoid tumors [2]. It is composed of fibroblasts and myofibroblasts with bland ovoid and spindle-shaped nuclei mixed in a collagenous matrix [15]. Immunohistochemical stains for β catenin is most useful for diagnosis and degree of cellularity is highly variable [16]. Although benign, they are fast growing tumors and rarely have been reported to metastasize [17]. They can be locally aggressive with a high rate of recurrence if gross total resection in not obtained [18]. Current literature suggests 100% recurrence after biopsy, 42-55% after curettage, 17% after subtotal resection, and 0% after gross total resection [19]. Adjuvant chemotherapy and radiotherapy is not currently recommended. This is the youngest reported case in the literature. Fortunately, this patient had gross total resection and remained without tumor recurrences out to a year follow up.

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