

# Desmoplastic Fibroma of the Pediatric Cranium: An Aggressive Skull Tumor with Local Recurrence

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## Abstract

**Cranial desmoplastic fibroma (DF) is extremely rare and only 20 cases, including only 7 pediatric cases, have been reported previously. We describe the first case of a child with cranial DF that increased in size over a short-term and recurred after resection. The aim of this case report was to discuss the clinical, radiological, and histological characteristics and optimal treatment for this rare and aggressive skull tumor.**

Key words: skull tumor, pediatric, desmoplastic fibroma

## Introduction

Desmoplastic fibromas (DFs) are rare bone tumors that present predominantly in the long bones and the mandible. Cranial DFs are extremely uncommon, and only 20 cases have been reported to date, including only 7 pediatric cases.<sup>1–18)</sup>

A palpable soft mass on the scalp (“a lump on the head”) is a common symptom of pediatric DFs. Computed tomography (CT) scans show an iso- or low-density mass lesion at the intradiploic space with local bone thinning or complete destruction of cortical bone, whereas magnetic resonance imaging (MRI) shows hypointensity on T<sub>1</sub>-weighted images, heterogeneous hyperintensity on T<sub>2</sub>-weighted images, and well-defined lesions on gadolinium contrast-enhanced MRI. Cranial DF is difficult to diagnose by radiographic findings alone.

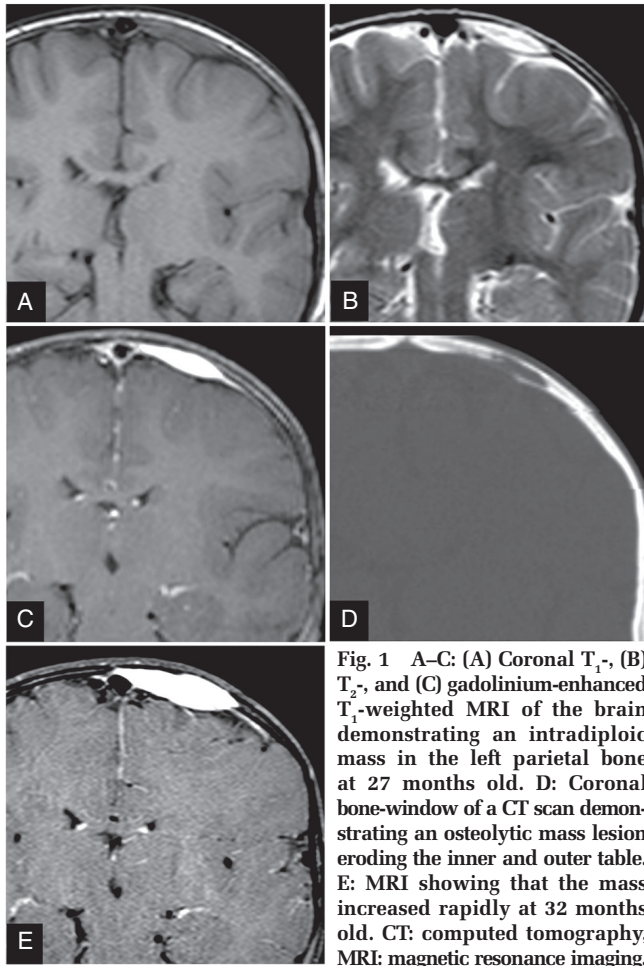
A histological examination is essential to achieve a definitive diagnosis of DF. Immunohistochemical staining for β-catenin, which is strongly expressed in the nuclei and cytoplasm of DF tumor cells, is useful for diagnosis.<sup>19,20)</sup> Compared with other bone tumors, DFs are benign but locally aggressive tumors, and therefore it is important to achieve total resection of the lesion including a sufficient margin. Here, we describe the case of a child with cranial DF that increased in size over a short-term and recurred after complete resection.

## Case Report

A 33-month-old girl who had suffered from recurrent afebrile seizures at 25 months, 30 months, and 31 months of age

was prescribed antiepileptic medication. On examination, her speech development was mildly retarded. Physical and neurological examinations revealed no remarkable abnormalities, and electroencephalogram and laboratory studies were normal. Chest radiography and abdominal sonography revealed no lesions.

At 27 months old, an MRI revealed a hypointense 40 × 10 × 35 mm spindle-shaped mass in the left parietal region that was hypointense centrally on T<sub>1</sub>-weighted images, and hyperintense peripherally on T<sub>2</sub>-weighted images (Fig. 1A, B). Contrast-enhanced MRI showed a well-defined homogeneous mass (Fig. 1C). A CT scan showed a mass located in the intradiploic space of the left parietal bone (Fig. 1D). The outer and inner tables of the calvarium were thinned, indicating that the mass originated from inside the bone rather than from the dura mater. The radiographic diagnosis was eosinophilic granuloma or cavernous hemangioma. At 32 months, an MRI showed that the mass had increased in size to 43 × 10 × 40 mm (Fig. 1E), and the child was admitted to our hospital for surgery. At 33 months old, she underwent surgery to remove the mass. The mass was elastic, hard, and bled easily. The mass infiltrated and replaced the inner table of the adjacent calvarium and was strongly adhered to the dura mater. After total lesion resection, there was no macroscopic evidence of residual tumor, and the adjacent dura was coagulated but not removed. A histological examination revealed that the tumor consisted of fibroblasts with a few myxomatous satellite cells within a collagenous background matrix (Fig. 2A, B). There were a few atypical or mitotic nuclei. Immunohistochemical analysis revealed that both the nucleus and cytoplasm of fibroblasts were strongly positive for β-catenin (Fig. 2C). Ki-67 positive cells were



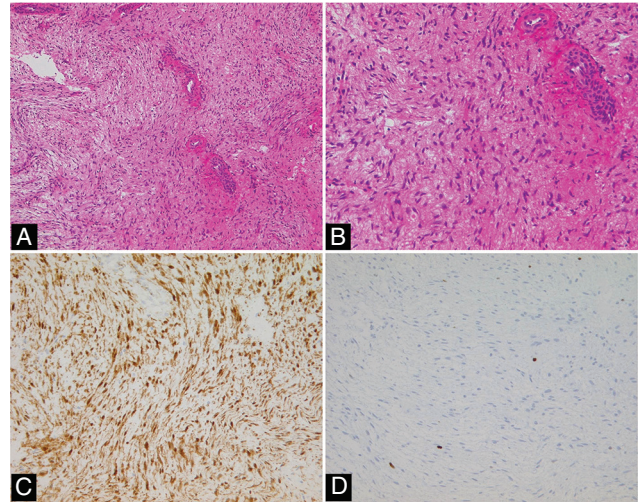
**Fig. 1** A–C: (A) Coronal T<sub>1</sub>-, (B) T<sub>2</sub>-, and (C) gadolinium-enhanced T<sub>1</sub>-weighted MRI of the brain demonstrating an intradiploic mass in the left parietal bone at 27 months old. D: Coronal bone-window of a CT scan demonstrating an osteolytic mass lesion eroding the inner and outer table. E: MRI showing that the mass increased rapidly at 32 months old. CT: computed tomography, MRI: magnetic resonance imaging.

rare (Fig. 2D), and cells were negative for S-100 protein. These findings were consistent with a diagnosis of DF.

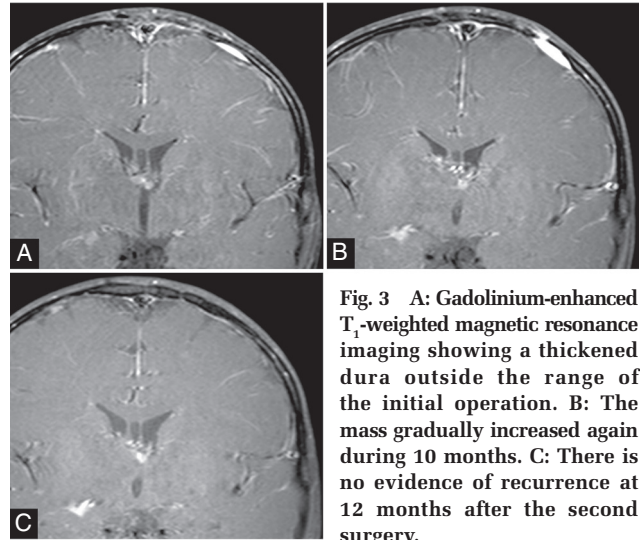
MRI 4 months after surgery revealed a thickened dura outside the range of the initial craniotomy (Fig. 3A) and a mass that had grown progressively (Fig. 3B). At 10 months postoperatively, we decided to perform a second and larger craniotomy to expose the tumor margin. The recurrent mass was removed completely, and the surrounding dura mater attached to the mass was coagulated enough. Since undergoing the second craniotomy 12 months ago, there have been no signs of recurrence on MRI (Fig. 3C) but she has repeated seizures and continues to need medical attention.

## Discussion

DFs are rare bone tumors that constitute 0.3% of benign bone tumors and 0.06% of all bone neoplasms.<sup>16)</sup> DFs usually occur in people < 30 years.<sup>21)</sup> They are found most commonly in the metaphysis of the long bones (58%) and less commonly occur in the pelvis, maxilla, sternum, and vertebrae.<sup>9)</sup> In the long bones, pathologic fracture is a common sign of DF, whereas DF in the mandible is



**Fig. 2** Photomicrograph showing fibroblasts and spindle-shaped cells with elongated nuclei within a collagenous background matrix. Hematoxylin and eosin staining, original magnification (A)  $\times 100$  and (B)  $\times 200$ . C: Photomicrograph obtained after immunostaining for  $\beta$ -catenin showing strong nuclear and cytoplasmic staining in fibroblasts. Original magnification  $\times 100$ . D: Ki-67 staining showing a few cells with positive nuclei. Original magnification  $\times 100$ .



**Fig. 3** A: Gadolinium-enhanced T<sub>1</sub>-weighted magnetic resonance imaging showing a thickened dura outside the range of the initial operation. B: The mass gradually increased again during 10 months. C: There is no evidence of recurrence at 12 months after the second surgery.

occasionally characterized by gradual swelling, asymmetry of the face, and pain or functional impairment. On CT scans, approximately 29% of DFs showed cortical breakthrough.<sup>22)</sup>

As mentioned earlier, DFs in the cranium are extremely uncommon. All previous cases of cranial DFs have presented as a palpable “lump on the head” as the sole symptom, although two cases of DFs at the temporal bone presented with hearing loss.<sup>15)</sup> In our case, the patient was evaluated for recurrent seizures and developmental speech delay but lacked a palpable lump on the head.

Previously, there has only been a single case of DF with speech delay,<sup>18)</sup> and no DF cases with seizures have been reported. In our case, it is not clear whether such symptoms were related to the mass, and the lack of superficial changes might indicate an early stage DF.

The preoperative radiographic diagnosis of DF remains a challenge because cranial DFs and other skull tumors are typically solitary lytic lesions. Studies of DF employing CT scans have revealed destruction and thinning of the cortical bone with expansion into the diploic space, and in all previous cases the lesion destroyed the outer table and protruded to the surrounding scalp, which was not the case in the present study. The prominence of the external surface leads to the suspicion of a skull tumor, but in our case, the scalp was intact, and the outer table alone showed thinning. In addition to the characteristic findings of MRI mentioned earlier, in some cases, internal flow voids of the intratumoral vessels have been noted on T<sub>2</sub>-weighted images,<sup>16,18)</sup> and in two cases the lesion had extended beyond the dura.<sup>5,11)</sup> However, these radiological findings are not specific for DFs, and this fact makes diagnosis difficult. The differential diagnosis includes eosinophilic granuloma, cavernous hemangioma, fibrous dysplasia, fibrosarcoma, chondromyxoid fibroma, giant cell tumor, simple bone cyst, aneurysmal bone cyst, chondrosarcoma, invasive meningioma, en-bloc meningioma, and metastasis.

Because of the difficulties associated with physical and radiological diagnosis, a histological examination is mandatory. DFs are composed of fibroblasts and myofibroblasts with bland ovoid or spindle-shaped nuclei within a collagenous matrix, presenting with many histologic patterns of various cellularity.<sup>23)</sup> Differential diagnosis from other spindle cell tumors is often problematic.<sup>23)</sup> However, positive nuclear  $\beta$ -catenin staining has been reported in most cases of desmoid-type fibromatosis and DFs. About 70–80% of DFs are positive for nuclear  $\beta$ -catenin staining.<sup>19,20)</sup>  $\beta$ -catenin, regulated by the adenomatous polyposis coli (APC) gene, is a cytoplasmic protein that is normally located beneath the cell membrane rather than in the nucleus. Mutations in both the  $\beta$ -catenin and APC genes have been reported to underlie the pathogenesis of desmoid-type fibromatosis, causing  $\beta$ -catenin expression in the nuclei.<sup>19,20)</sup> Thus,  $\beta$ -catenin staining is considered to be mandatory for a diagnosis of DF and desmoid-type fibromatosis. DFs are also positive for vimentin and smooth muscle actin, whereas they are typically negative for CD1a, CD34, desmin, cytokeratin, S100, epithelial membrane antigen, estrogen, and progesterone.<sup>4,12,18)</sup> In addition, the Ki-67 index range is < 5% in DFs, but much higher in malignant entities.<sup>23)</sup>

DFs are benign but have locally aggressive features; they have not been reported to metastasize,<sup>5,11)</sup> but tend to recur locally as in the present case. The Second Edition of the World Health Organization histologic classification of bone tumors designated DFs as intermediate group tumors because of this characteristic.<sup>24)</sup> It has been suggested that

DFs can be managed by complete lesion excision,<sup>5,13)</sup> and that near-total excision may prevent recurrence.<sup>25)</sup> However, other reports have suggested the need for wide excision.<sup>11,26)</sup> The recurrence rates have been reported as 100% after biopsy,<sup>27)</sup> 42–55% after curettage,<sup>26,27)</sup> 72% after excision,<sup>27)</sup> 8.3–17% after partial resection,<sup>27,28)</sup> and 0% after wide resection.<sup>14,27)</sup> The potential benefit of adjuvant therapy, including radiotherapy or chemotherapy, has not yet been established. If the tumor extends beyond the intracranial space, it should be excised together with the involved bone and the dura.<sup>11)</sup> In the present case, we noted recurrence following near-total excision, suggesting that aggressive total excision with a sufficient margin is necessary to prevent recurrence. Long-term follow-up with CT or MRI is recommended for at least 3 years because the median time to recurrence has been reported as approximately 2.7 years.<sup>17,26)</sup>

In conclusion, pediatric cranial DFs are extremely rare. Patients often, but not always, presented with a prominence of the skull that is rarely discovered incidentally. Because of the difficulties associated with diagnosis, a histological examination in combination with physical and radiological examinations should be mandatory.

Immunohistochemical staining for  $\beta$ -catenin proved useful for a definitive diagnosis of DF. If this tumor is suspected, complete resection with wide margin should be performed to prevent recurrence.

## Conflicts of Interest Disclosure

None of the authors have any financial interest in the materials and devices described in this article.

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