

Case Report

Desmoplastic Fibroma of the Cranium in a Young Man

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Desmoplastic fibroma, which develops predominantly in long bones and the mandible, is a rare and benign but locally aggressive tumor. Desmoplastic fibroma of the cranium is extremely rare. We report a case of desmoplastic fibroma of the frontal bone in a young man. Because of its locally aggressive behavior, complete surgical excision with a safety margin is essential.

Key Words : Desmoplastic fibroma · Frontal bone · Skull.

INTRODUCTION

Desmoplastic fibroma (DF) is an intra-osseous tumor that frequently occurs in long bones and the mandible¹⁵. These tumors are histologically benign but show locally aggressive behavior⁹. Without complete resection, frequent local recurrence occurs¹². DF in the cranium is extremely rare. Since the first report by Gardini et al.⁷ in 1978, fewer than 20 cases have been described in the literature^{5,11,14}. In this report, we present a case of DF in the cranium. Differential diagnosis and treatment strategy are discussed with a literature review.

CASE REPORT

A 20-year-old man visited our clinic with a 1-year history of worsening headache and swelling of the right frontal region. Physical examination revealed bony swelling with a smooth cortical lining. The swollen area was mildly tender, and the overlying scalp was intact. Neurological examination was unremarkable. Plain skull X-ray films demonstrated a lytic lesion with a sclerotic margin. A computed tomographic (CT) scan demonstrated a 3×3.5-cm focal calvarial thickening and expansion of the diploic space by a hypo-attenuated mass with a sclerotic margin and ground-glass appearance. Magnetic resonance imaging showed heterogeneous signal intensity on T2-weighted images and intermediate signal intensity on T1-weighted images

with multifocal enhancement (Fig. 1). There was no soft tissue or intracranial invasion. Focal hot uptake at the lesion site was observed on bone scan.

The patient underwent a craniectomy under general anesthesia. The lesion was completely excised with sufficient safety margins by inspection under guidance of intra-operative navigation. There were neither scalp nor dural invasions. The gross specimen was bulging with a pinkish color at the lesion site, which contained a round dark-brownish area with a sclerotic margin in the diploic space, slightly expanded and thicker compared to normal bone flaps. Sectioning after decalcification revealed an ill-defined fibrotic area. The lesion was accompanied by yellow necrotic foci and exhibited fibroblastic proliferation within a collagenous background, with extensive hemorrhage and fat necrosis. Tumor cells included spindle cells and lacked significant unclear atypia and mitotic figures. These findings suggested desmoplastic fibroma of the skull. Immunohistochemical stain for smooth muscle actin (SMA) was focally positive (Fig. 2), and S-100 was positive. Other markers such as EMA, CD34, CD68, vimentin, and desmin were negative. All margins were free of tumor cells.

DISCUSSION

First described by Jaffe in 1958, desmoplastic fibroma is recognized as a separate entity from bone tumors. DF is a rare be-

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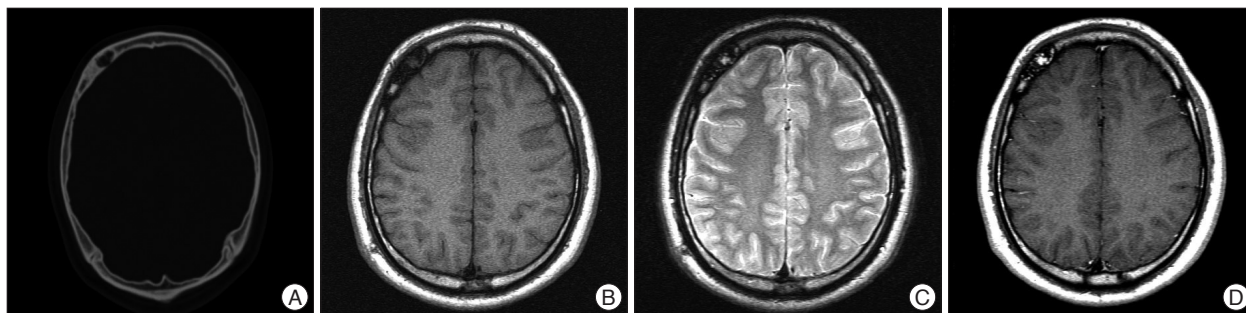


Fig. 1. A : CT scan demonstrating focal calvarial thickening of right frontal skull. B, C and D : MRI reveals intermediate signal intensity on T1-weighted images and heterogeneous intensity on T2-weighted images with multifocal enhancement.

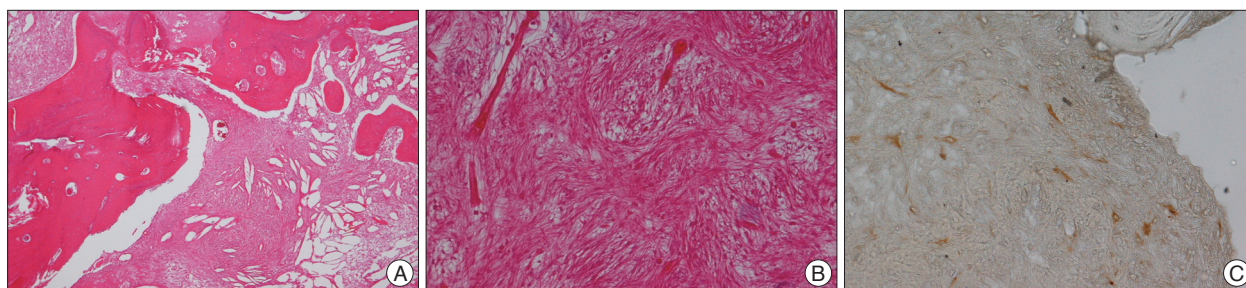


Fig. 2. A : Micrograph of the lesion showing spindle cell proliferation with hemorrhage and fat necrosis (H&E, $\times 100$). B : Note fibroblastic proliferation with collagenous background (H&E, $\times 200$). C : Smooth muscle actin staining revealed focal positivity ($\times 200$).

nign bone tumor composed of spindle cells accounting for 0.3% of benign bone tumors¹³). The World Health Organization describes DF as exhibiting “minimal cytological atypia and abundant collagen production”⁶). DF can occur at any age, but most cases occur before age 30¹). DF may involve any bone, but commonly occurs in the metaphyses of the long bones, mandible, and pelvis, and cranial DFs are exceedingly rare. DFs affect both genders, although a female preference has been suggested for cranial DF¹²). Despite benign histological appearance and slow growth, frequent local recurrence puts DF in a category between benign and malignant bone tumors³). Clinical symptoms include headache, cranial asymmetry, ear symptoms, and skull mass. Intracranial involvement was reported in one case, but brain parenchymal invasion has not been reported¹⁷).

Radiographically, cranial DFs are often solitary, honeycomb, or trabeculated lytic lesions, with expansion of the diploic space with or without marginal sclerosis⁸). CT scans typically show destruction and thinning of the cortex. Dense connective tissue and hypocellularity yields heterogeneous signal intensity on T2 and iso-signal intensity on T1-weighted magnetic resonance images with heterogeneous enhancement^{9,16}). These findings are not distinctive compared to other skull lesions such as fibrous dysplasia, hemangioma, eosinophilic granuloma, low-grade osteosarcoma, or metastasis. Given its rarity and nonspecific radiographic findings, it is very difficult to diagnose cranial DFs.

Differential histopathological diagnosis includes benign and malignant spindle cell bone tumors, including fibrous dysplasia, fibrosarcoma, low-grade intra-osseous osteosarcoma, and non-ossifying fibroma¹⁰). The crucial differential diagnosis is low-grade fibrosarcoma. Typical fibrosarcoma is more cellular with a

herringbone pattern that shows more pleomorphism and higher mitotic activity⁴). In fibrous dysplasia, recognition of C-shaped, woven bone formation within a fibrous background is an important diagnostic feature. Osteoid production is generally evident in intraosseous osteosarcoma. Non-ossifying fibroma consists of cellular masses of fibrous tissue often arranged in a storiform pattern. Foamy and hemosiderin-laden macrophages are common in non-ossifying fibroma. Hemorrhage and fat necrosis seen in our case are uncommon findings in DF. Immunohistochemistry of DF may be positive for SMA, vimentin, and desmin, while cytokeratin, S-100, EMA, and CD16 are negative¹⁸).

Surgical resection with adequate margins, with cranioplasty, is the treatment of choice. Recurrence is rare if negative margins are achieved. Despite some reports that endocrine therapy with tamoxifen can be effective, chemo- and radiotherapeutic approaches in DF have not been established²). So far, there are no reports of recurrence in cranial DF, which may be due to feasibility of wide excision in skull lesions. Since cranial DF remains poorly understood, radiographic follow-up is warranted, especially in incomplete resection cases.

CONCLUSION

DF is uncommon, but should be considered in differential diagnosis of any expansile cranial mass. Because DF may be locally aggressive, en bloc resection with adequate margin is the treatment of choice.

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