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 mandible. These tumors are histologically benign but show locally aggressive behavior. Without complete resection, frequent local recurrence occurs. DF in the cranium is extremely rare. Since the first report by Gardini et al. in 1978, fewer than 20 cases have been described in the literature. 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-

---

**References**

3. Byung-Kyu Cho, M.D., Ph.D.
4. Sung-Mook Jung, M.D.
5. Sungjoon Lee, M.D.
6. Hoon Kim, M.D.

**Address for reprints:** Hoon Kim, M.D.
Department of Neurosurgery, College of Medicine, The Catholic University of Korea, Bucheon St. Mary's Hospital, 327 Sosa-ro, Wonmi-gu, Bucheon 420-717, Korea
Tel.: +82-32-340-2259, Fax.: +82-32-340-7391, E-mail: armada1997@naver.com

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (http://creativecommons.org/licenses/by-nc/3.0) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

---


Print ISSN 2005-3711  On-line ISSN 1598-7876

online © ML Comm
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.

**References**