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Chondromyxoid Fibroma of Frontal Bone: A Case Report and Review of the Literature

Frontal Kemikte Yerleşen Kondromiksoid Fibroma: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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ABSTRACT

Chondromyxoid fibroma is an unusual benign tumor of cartilaginous tissues that may be confused with other some malign tumors. It is rarely seen in the skull. A 45-year-old female was admitted with painless bony swelling in the forehead. Computerized tomography demonstrated a well-defined expansive lesion with a sclerotic margin measuring ~ 3 cm in diameter in the right frontal bone. On MRI, T1-weighted images revealed a well circumscribed, lobulated and strongly enhancing lesion. On the T2-weighted images, the lesion showed high heterogeneous signal intensity. The patient underwent tumor excision with craniectomy, then acrylic cranioplasty. The histopathological diagnosis was chondromyxoid fibroma. There was no recurrence in a period of 22-months. In conclusion, chondromyxoid fibroma is a benign primary bone tumor that is located extremely rarely in the frontal bone. Accurate initial diagnosis of such tumors are important for appropriate treatment. En block surgical resection of the tumor is the cornerstone of treatment.

KEY WORDS: Benign cartilaginous tumor, Chondromyxoid fibroma, Frontal bone, Skull tumor

ÖZ

Kondromiksoid fibroma kıkırdak dokusunun az görülen iyi huylu tümörlerindendir ve tanı konurken diğer bazı kötü huylu tümörlerle karıştırılabilir. Kafatasında ender olarak görülür. 45 yaşındaki kadın hasta alındaki şişlik yakınması ile başvurdu. Bilgisayarlı tomografi tetkikinde sağ frontal kemikte yaklaşık 3 cm çapında, kenarları sklerotik, iyi sınırlı ve genişleyici özellikte bir lezyon saptandı. Manyetik rezonans incelemesinde kitlenin T1 ağırlıklı kesitlerde lobüle, iyi sınırlı ve kuvvetli boyanma gösterdiği, T-2 ağırlıklı kesitlerde heterojen sinyal özelliği bulunduğu görüldü. Hastaya kranyektomi ile total kitle eksizyonu ve kranyoplasti ameliyatı yapıldı. Kitlenin histopatolojik tanısı kondromiksoid tümör olarak rapor edildi. Hasta 44 aydır izlenmektedir ve herhangi bir nüks saptanmamıştır. Sonuç olarak frontal kemikte yerleşim gösteren kondromiksoid fibroma oldukça nadir görülmektedir. Bu tümörün tedavisindeki en önemli nokta tümörün tümüyle rezeksiyonudur.

ANAHTAR SÖZCÜKLER: Beningn kartilajinöz tümör, Kondromiksoid fibroma, Frontal kemik, Kafatası tümörü

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INTRODUCTION

Chondromyxoid fibroma (CMF) is a well-known but uncommon tumor that usually occurs in the long bones (2). Jaffe and Lichtenstein described it for the first time in 1948 (9). It is estimated to represent less than 1% of all primary bone tumors (6). Although CMF has been reported in numerous anatomic sites of the body (21) the involvement of the skull is extremely rare (3,5,6,8). This tumor has been defined as "a benign tumor characterized by lobules of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material separated by zones of more cellular tissue rich in spindle-shaped or round cells with varying number of multinucleated giant cells of different sizes" by the World Health Organization (7). It may be misdiagnosed as other tumors such as chondrosarcoma because of some similarities (6,12), and it is important to distinguish it by establishing the clinical, radiological and pathological features of CMF.

To our knowledge, only 12 cases involving the frontal bone have been reported until now (Table I). In this study, we describe the clinical, radiological and pathological features involving the frontal bone in a 45-year-old woman.

CASE REPORT

This 45-year-old woman presented with a local swelling that progressed in the last year in her the right front part of her head. Her medical past was unremarkable and her neurological examination was normal. Physical examination readily revealed a hard, spherical bony swelling in the right frontal area. The X-ray examination showed a round radiolucent area with sclerotic margins in the right frontal squama (Figure 1) The CT scan showed an osteolytic lesion with expansion of the inner and outer tables of the right frontal bone (Figure 2). It had irregular mixed density with some calcification in front part of the lesion. MRI with contrast enhancement revealed a well-circumscribed,

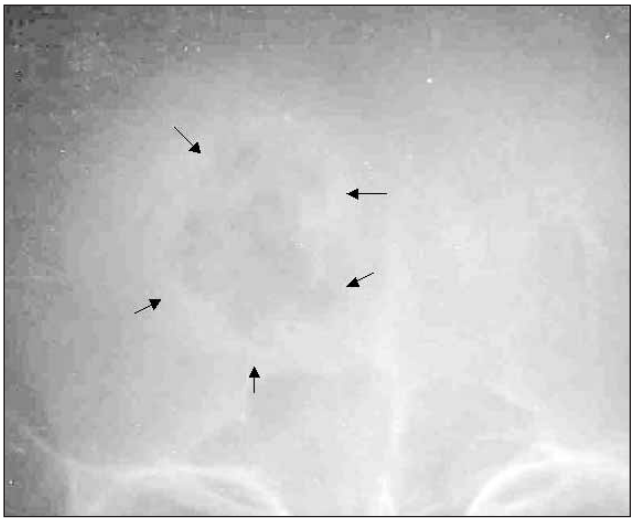
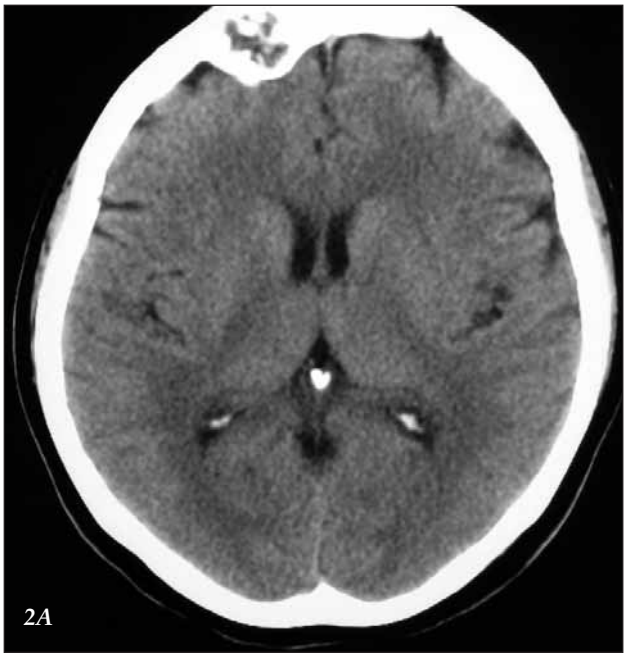


Figure 1: *A round radiolucent area with sclerotic margins (black arrows) in the right frontal squama on plain x-ray.*

Table I: List of the Chondromyxoid Fibroma Cases Involving Frontal Bone

Case no	Author & year	Age & Sex	Location
1	Chalapati et al, 1976 4	30 y / F	Right frontal bone
2	Miyamoto et al, 1981 15	15 y / F	Frontal bone
3	Thurner J & Lisanti M, 1981 17	? / ?	Frontal bone squama
4	Watanabe et al, 1985 19	20 y / M	Frontal bone
5	Kanamaru et al, 1982 10	15 y / M	Orbital roof of frontal bone
6	Morimura et al, 1992 16	41 y / M	Frontal bone
7	Wolf et al, 1997 20	35 / F	Left inferolateral frontal bone
8	Wu et al, 1998 21	? / ?	Frontal bone
9	" "	? / ?	Frontal bone
10	" "	? / ?	Frontal bone
11	Baujat et al, 2001 2	50 y / F	Nasal bone extending into the frontal and ethmoidal sinuses
12	Azorin et al, 2003 1	46 y / M	Right frontal sinus
13	Present case, 2008	45 y / F	Right frontal bone



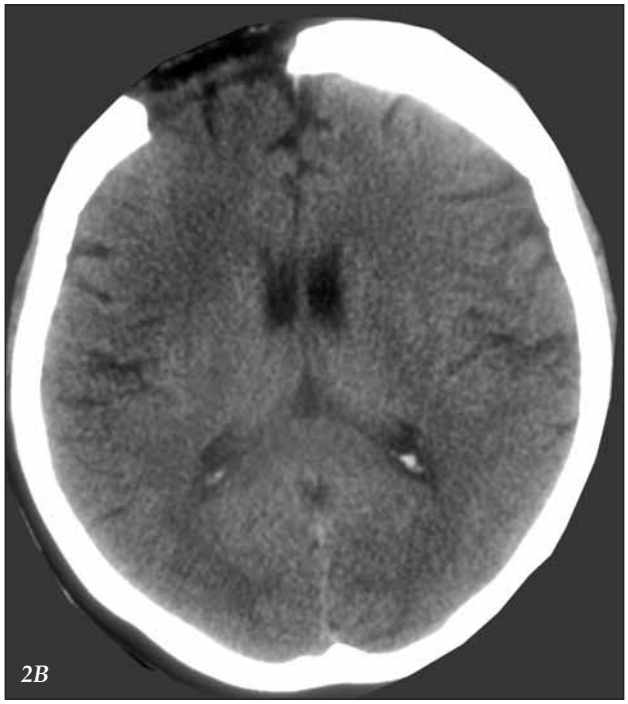
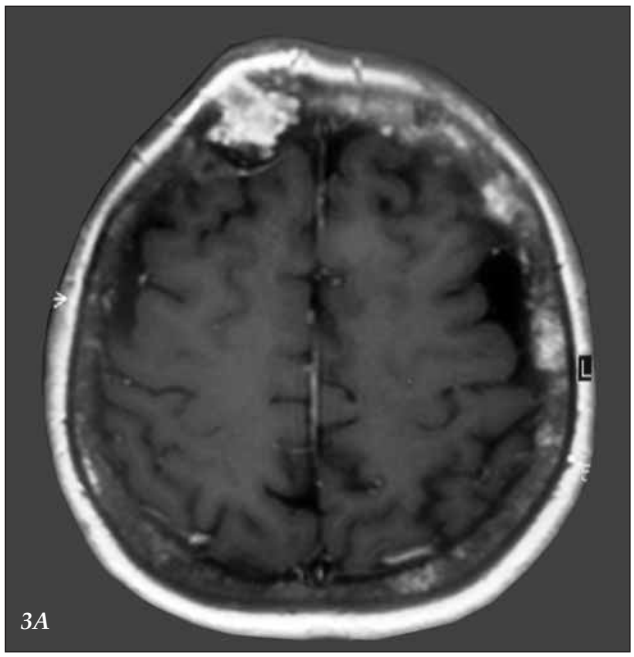


Figure 2: ***A****: The CT scan showed an osteolytic lesion with expansion of the inner and outer tables of the right frontal bone. An irregular mixed density with some calcification in front part of the lesion is easily seen.* ***B****: The postoperative CT of the patient showing the area that craniectomy and cranioplasty were performed.*



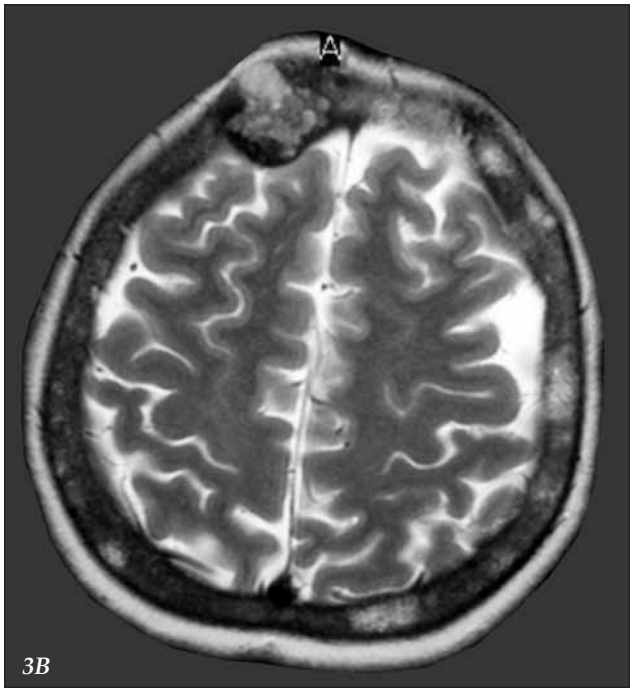


Figure 3: *MRI with contrast enhancement revealed a well-circumscribed, lobulated lesion that is approximately 3 cm in diameter located in the right frontal bone.* ***A****: T1 weighted MR scan with strong contrast enhancement and* ***B****: T2-weighted MR scan with heterogenous signal intensity.*

lobulated lesion that was approximately 3 cm in diameter (Figure 3) locating in the right frontal bone. Following an en bloc resection of the lesion, an acrylic cranioplasty was performed at the same operation. Histopathological examination of the tumor showed a lobular pattern with stellate shaped cells in a myxoid background. The lobules demonstrated hypo-cellular centers and hyper-cellular peripheries, and focally located multinuclear giant osteoclastic cells were seen (Figure 4). The

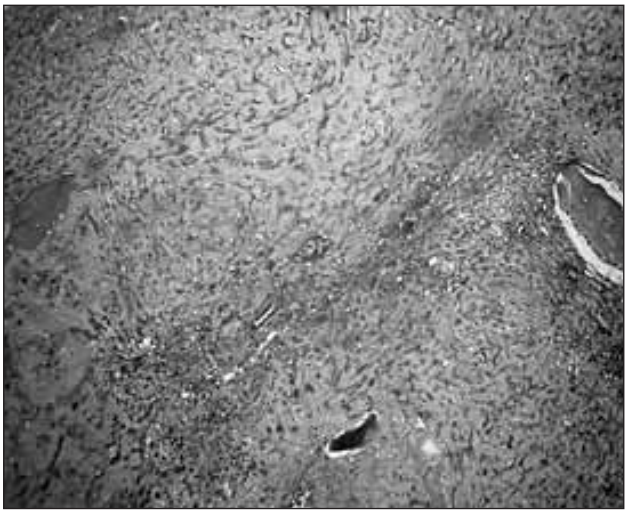


Figure 4: *Chondromyxoid fibroma: nodule of chondromyxoid tissue with poor delineation of fibroid element. (H&E x 100)*

interesting point of this pathological examination was the appearance of lesion fragments in the perilesional spongy bone tissue that had the same microscopic findings with the ones mentioned above. The postoperative period was uneventful. There was no recurrence or new lesion formation during the 20-month follow up period.

DISCUSSION

Chondromyxoid fibroma is an unusual benign tumor of cartilaginous tissues (11). Although it involves the metaphyses of major long bones (21), it has been observed in different anatomic locations (22). Skull bones develop from endochondral tissue and it is believed that CMF arises from cartilage (8) or embryonic cartilage residue (14).

CMF occurs in all age groups (5,22) but mostly in the second and three decades (5,18,21). There is a slightly predilection in males (3,18,21). Zillmer et al (22) reported that the location of chondromyxoid fibroma varies with age. It is found predominantly in long bones between 1 and 10 years, while the distribution of localization becomes equal between the long bones, flat bones and ribs in the fourth decade. According to some authors, the average age for intracranial CMF is 37 (20) – 39 (6) years. It is 32.7 years (range 15-50 years) among patients who have frontal CMF.

The duration of the symptoms before diagnosis varies between 10 days and 20 years (6,22). The clinical symptoms are mostly related to the site where the tumor arises. In case of cranial involvement, the symptoms change according to the bone that is affected. Patients can present with diplopia (6), neuralgia and dysarthria (3), facial pain (8), episodes of convulsions (11), exophthalmus (10) and headache (12,20) or with bony swelling as in our case.

Although they are rare tumors, the radiological features are well known. In plain radiographs the lesions are radiolucent with well-defined margins (5,19). The CT scan shows osteolytic lesions with a sclerotic margin (11). The tumor may show foci of calcification, with mineralization that is obvious radiologically in about 13 % (6,8,21). Hypointensity can be seen on T1-weighted MRI images and hyperintensity that indicates water content on T2-weighted images; the intensive contrast enhancement is classic for CMF (6,16). Most of these radiological findings such as radiolucency with sclerotic margin on plain films, mineralization on CT scans, hypo-intense areas on T2-weighted MRI with intensive contrast enhancement on T1-weighted MR were found in the presented case.

Chondrosarcoma, fibrous dysplasia, chondroblastoma (5,6,8,11,12,13), chordoma (6,8,11) and enchondroma (11) are listed in the differential diagnosis. Some chondrosarcomas may demonstrate peripheral lobularity. This feature is what usually causes confusion when making a distinction between CMF and chondrosarcoma. This kind of chondrosarcomas, however, are usually of grade 2 or higher and thereby demonstrate a high-grade sarcoma with plenty mitoses. Mesenchymal chondrosarcoma produces islands of chondroid to chondrosarcomatous blue hyaline cartilage. A hemangiopericytoma-like pattern is often evident. Mitoses are not difficult to find. The presence of "chondroblasts" of a chondroblastoma each of which is surrounded by a honeycomb, reticulin sheath on silver stains is distinctive; the fairly large numbers of osteoclast-like giant cells distributed throughout the lesion and "chicken wire" calcifications should distinguish these two entities.

The main therapy of CMF is surgery, especially "en block" resection. Taking care to include a rim of normal bone is recommended because of probability of leaving small tumor islands (5,6,8). An en block resection may also avoid the recurrence and the possible spontaneous malignant formation of the tumor that is reported as extremely low (2). We did an en block resection in the presented case and it has

not recurred in a follow-up period of 20 months. The rate of recurrences vary between 3 % and 26 % with an interval of 5 months to 10 years (5,21,22). The high recurrence rate may be due to incomplete resection of tumor with curettage (11). The severe functional and cosmetic morbidities after wide resection of the tumor can be problem (6,13). Acrylic cranioplasty was performed in this presented case especially for prevention of cosmetic bone defect in frontal area.

Postoperative high-dose irradiation is recommended for incomplete resections due to the location of tumor near critical structures such as the skull base (12,18). According to Feuvret et al (6), radiotherapy is now part of standard treatment of CMF. They combined treatment with proton beam to minimize acute and late side effects. Radiotherapy was not used in the presented case but may be considered if recurrence occurs.

In conclusion, chondromyxoid fibroma is a benign primary bone tumor that is located extremely rarely in the frontal bone. The accurate initial diagnosis of such tumors is important for appropriate treatment. En block surgical resection of the tumor is the cornerstone of treatment. Cranioplasty may be done in the same session to prevent cosmetic morbidity.

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