Case Report / Olgu Sunumu

Fibromyxoma of the frontal sinus

Frontal sinüsün fibromiksomu

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Fibromyxomas are uncommonly encountered pathologies in the head and neck region. They are slow growing, which result in expansion of the surrounding bony cortices. In this article, we report an extremely rare case of frontal sinus fibromyxoma which occluded the frontal sinus and expanded anterior ethmoid cells and nasal dorsum. Initially, it was suspected to be a mucocele. Surgical resection with a wide excision was performed and it was diagnosed as fibromyxoma, histopathologically.

Key Words: Fibromyxoma; frontal sinus; nasal bone.

Fibromiksomlar, baş ve boyun bölgesinde nadir rastlanan patolojilerdir. Yavaş büyüyerek, etrafı çevreleyen kemikli kortekslerin genişlemesine neden olur. Bu yazıda, frontal sinüsü tıkayan ve anteriyor etmoid hücreler ve nazal dorsumu genişleten, çok nadir görülen bir frontal sinüs fibromiksom olgusu sunuldu. Başlangıçta mukosel olabileceğinden şüphe edildi. Geniş eksizyon ile cerrahi rezeksiyon yapıldı ve histopatolojik incelemede fibromiksom tanısı kondu.

Anahtar Sözcükler: Fibromiksom; frontal sinüs; burun kemiği.

Fibromyxomas are rare tumors of the head and neck region. They usually occur in the second and third decades of life, rarely in children or adults over 50 years of age.[1,2] Although these tumors are slow-growing benign lesions, they may cause extensive local destruction with a tendency to recurrence after surgical excision. The posterior region of the mandible and maxilla are mostly affected.[3,4] The origin of an odontogenic myxoma is still not clear; origin from the dental follicle seems to be the most reasonable explanation.[1] They are benign but locally aggressive and cause facial deformity, loosening of teeth and extend to the orbit. Also, fibromyxomas are reported in the petrous apex, lacrimal sac, ethmoidal sinus and frontal sinus.[5-8] But these are extremely rarely localized. We report a female case with pain in the right frontal region and expansion at the root of the nose. She was treated with surgical excision of the lesion, which was reported as fibromyxoma. To our knowledge, only one case of frontal sinus fibromyxoma has been reported in the literature.[8]

Received / *Geliş tarihi:* January 14, 2012 Accepted / *Kabul tarihi:* February 20, 2012

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CASE REPORT

A 71-year-old female complained of pain on the right frontal and orbital regions. On examination, the patient was found to have an enlarged nasal dorsum. Anterior rhinoscopy did not indicate any abnormality. The medical anamnesis of the patient did not reveal anything in relation to the pathological condition. Other head and neck examinations did not reveal any other abnormality. A computed tomography (CT) scan of the paranasal sinuses revealed a 29x25 mm low density lesion extending from the right frontal sinus to the right and left anterior ethmoid cells. Also, CT showed expansion of the walls of the adjacent sinuses. Intravenous administration of contrast agent showed no enhancement of the lesion (Figure 1a-c). The lesion was approached by a brow incision. The lesion had destroyed the upper nasal bone and nasal processes of the frontal bone. It was removed with enucleation and curettage. The mass was grey-yellowish, encapsulated, rubbery and fragile. The histopathological examination revealed randomly stellate, oval and spindle-shaped cells in a myxoid stroma. Immunohistochemical examination by means of vimentin and smooth muscle actin (SMA) labeling index revealed a low rate of cell mitosis (Figure 2a-c). One year postoperatively, the patient showed no signs of recurrence.

DISCUSSION

Fibromyxoma is classified as a specific type of myxoma with a higher fibrous/myxoid tissue ratio than myxoma. Previous theories claimed that the lesion derives from the neural sheath or results from degeneration of fibromas or lipomas due to degenerative processes following tissue anoxemia and chronic irritation.[9] Maxillary and mandibular myxomas are locally aggressive neoplasms that are thought to arise from fibroblasts of the dental papilla or sinonasal mucosa. The precise origin of these tumors, however, remains controversial.[10]

Histopathological characteristics of the fibromyxomas are hypocellularity and presence of stellate, spindle-shaped cells in a loose myxoid extracellular matrix. The fibromyxoid lesions may be distinguished from typical myxoma with higher amount of collagen fibers. The presence of cells positive for actin fibers put up the argument that myofibroblasts may play an absolute role in cell proliferation in cooperation with the islands of odontogenic epithelium and mast cells.[11-13] In our case; the tumor was composed of a loosely cellular proliferation of stellate or spindle-shaped cells within an abundant myxoid matrix. Immunohistochemical analysis showed positivity of tumor cells for anti-vimentin antibody and SMA. But the cells remained negative for S-100, desmin and anti-GFAP (glial fibrillary acid protein).

The radiological, histological and histochemical investigations help diagnosis. The radiological investigation reveals homogeneous radiolucencies or sclerotic trabeculations with different appearances, like "honeycomb", "soap bubble" and "tennis racket".[14] Adekeye at al.[15] in their cases of myxomas showed two different types radiologic

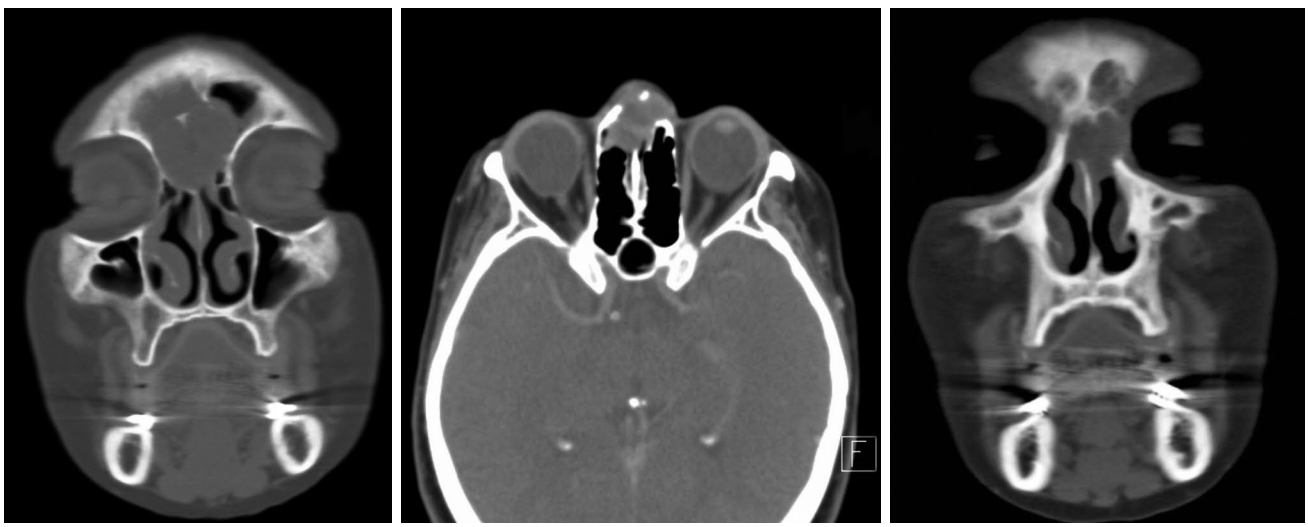

Figure 1. *Computed tomography reveals expansion of the walls of the right frontal sinus, obstruction with low density tissue of the whole cavity, and local erosion of the nasal bone.*

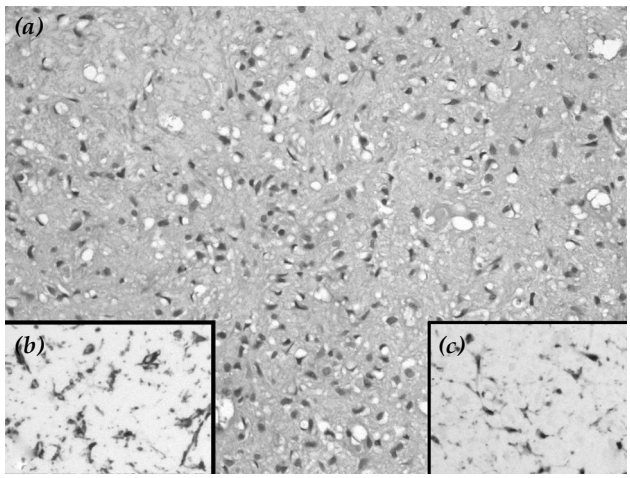

Figure 2. *The tumor was composed of a loosely cellular proliferation of stellate or spindle-shaped cells within an abundant myxoid matrix. Immunohistochemical analysis showed positivity of tumor cells for anti-vimentin antibody and smooth muscle antigen.* ***(a)*** *(H-E x 400),* ***(b)*** *vimentin,* ***(c)*** *Smooth muscle actin.*

appearance: one type showed a radiolucent image with diffuse margins and "soap bubble" or "honeycomb" appearance, the second radiologic type showed a well-defined radiolucency with a faint sclerotic margins. Cohen et al.[16] reported a case with computed tomography findings that revealed a large, honeycombed expanding lesion involving the entire maxillary sinus.

In our case the lesion appeared as a large low-density lesion which destroyed the anterior frontal sinus wall and upper nasal bones. On magnetic resonance imaging (MRI) the myxoma shows low-signal intensity in the T_1 and high-signal intensity in T_2.[17] Radiological examination plays a crucial role for the differential diagnosis of myxomas/fibromyxomas and also between benign myxomas and malignant neoplasms with myxomatous tissue. Ameloblastoma, metastatic lesions, mucocele, fibrous displasia, odontogenic cycts, well-defined liposarcoma and aneurysmal cysts should be kept in mind in the differential diagnosis.[17]

The treatment of the fibromyxoma is surgical and involves enucleation and curettage. Recurrence is strongly related to incomplete resection of the lesion.[17] Its local aggressiveness and gelatinous consistency contribute to difficulties in removing it completely.[18]

Myxomas and fibromyxomas show a recurrence rate between 25 and 42%.[1,2] Because of the high recurrence rate, the patient should be follow-up at least two years after surgery. One year postoperatively, our patient showed no signs of recurrence.

In conclusion, the frontal sinus is a rare location for a fibromyxoma. The radiological examination by means of CT and MRI play an important role in the diagnosis of a fibromyxoma and in the differential diagnosis from other pathological entities such as sinus malignancies and mucoceles. In destructive sinus lesions, fibromyxoma should be kept in mind by otolaryngologists.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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