

CASE REPORT

## A case of monostotic fibrous dysplasia of the maxillary sinus

### Maksiller sinüste monostotik fibröz displazi: Olgu sunumu

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Fibrous dysplasia is an uncommon benign disease of the bone, with slow progression. Monostotic involvement of the paranasal sinuses is rare. We report a 54-year-old woman who had complaints of facial asymmetry, chronic sinusitis, recurrent headaches, and nasal obstruction for two years. Conventional radiography showed opacification and expansion of the maxillary sinus. Axial and coronal computed tomography scans showed a heterogeneous mass that expanded the right maxillary sinus, leading to nasal obstruction and cortical thickening of the maxilla. No signs of destruction or erosion in the cortical bone were identified. An endonasal endoscopic biopsy was performed and the diagnosis of fibrous dysplasia was confirmed histologically.

**Key Words:** Fibrous dysplasia, monostotic/pathology/radiography; maxillary sinus/pathology/radiography; paranasal sinus diseases/pathology/radiography; tomography, X-ray computed.

Fibröz displazi, yavaş ilerleme gösteren, benign karakterli nadir bir kemik hastalığıdır. Paranasal sinüslerin monostotik tutulumu nadirdir. İki yılı aşkın bir süredir yüz asimetrisi, kronik sinüzit, tekrarlayıcı baş ağrısı ve nefes darlığı şikayetleri olan 54 yaşındaki kadın hastanın çekilen düz grafilerinde opasifikasyon ve maksiller sinüste genişleme gözlemlendi. Aksiyel ve koronal bilgisayarlı tomografi görüntülerinde maksiller sinüste genişlemeye, burun tıkanıklığına ve maksillada kortikal kalınlaşmaya yol açan heterojen kitle belirlendi. Kortikal kemikte herhangi bir erozyon ya da bozulma belirtileri yoktu. Hastaya endonazal endoskopik biyopsi yapıldı ve fibröz displazi tanısı histolojik olarak doğrulandı.

**Anahtar Sözcükler:** Fibröz displazi, monostotik/patoloji/radyografi; maksiller sinüs/patoloji/radyografi; paranasal sinüs hastalıkları/patoloji/radyografi; bilgisayarlı tomografi.

Fibrous dysplasia (FD) involving the paranasal sinuses is a subset of craniofacial FD. It is characterized by physiologic resorption and replacement of medullary bone with distorted, structurally unsound fibrous tissue. This process may extend beyond normal bony boundaries, and when the paranasal sinuses are involved, obstruction may occur, leading to recurrent infections, mucocele formation, and nasal airway obstruction.<sup>[1,2]</sup>

Although craniofacial involvement in FD is well described, isolated involvement of the paranasal sinuses is rare.<sup>[3]</sup> We here report a case of FD involving the right maxillary sinus, together with its clinical symptoms and radiological characteristics.

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A fifty-four-year-old woman was suffering from facial asymmetry, recurrent headaches, and unilateral

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nasal airway obstruction for two years despite medical treatment. She underwent diagnostic endoscopy for chronic sinusitis three years ago. Physical examination revealed facial deformity and a non-tender bony overhang above the right maxilla, displacing the globe and causing a downward limitation of eye movements. Conventional radiography showed opacification and expansion of the maxillary sinus (Fig. 1). Axial and coronal computed tomography (CT) scans showed a heterogeneous mass that expanded the right maxillary sinus with focal punctate calcifications in the matrix, and developed nasal obstruction and cortical thickening of the maxilla. No signs of destruction or erosion in the cortical bone were identified (Fig. 2a, b). The mass was located in the anterior wall of the sinus with a wide base extending superiorly to the orbit. There were tissue defects and air-dense areas within the lesion and in the anterior wall of the sinus associated with the former biopsy. A diagnostic endonasal endoscopic biopsy of the mass was performed and an histologic diagnosis was made as fibrous dysplasia.

#### DISCUSSION

Fibrous dysplasia is usually a slowly progressive benign disease that develops over several years,



Fig. 1 - Water's view of the facial bones showing opacification and expansion of the right maxillary sinus.

leading to deformity or mild symptomatology.<sup>[3]</sup> It is a developmental disorder of a growing bone, the etiology of which is unknown.<sup>[4]</sup> Trauma has been suggested as a responsible factor for the onset of FD.<sup>[5]</sup>

Fibrous dysplasia may involve single bones as monostotic or multiple bones as polyostotic forms.

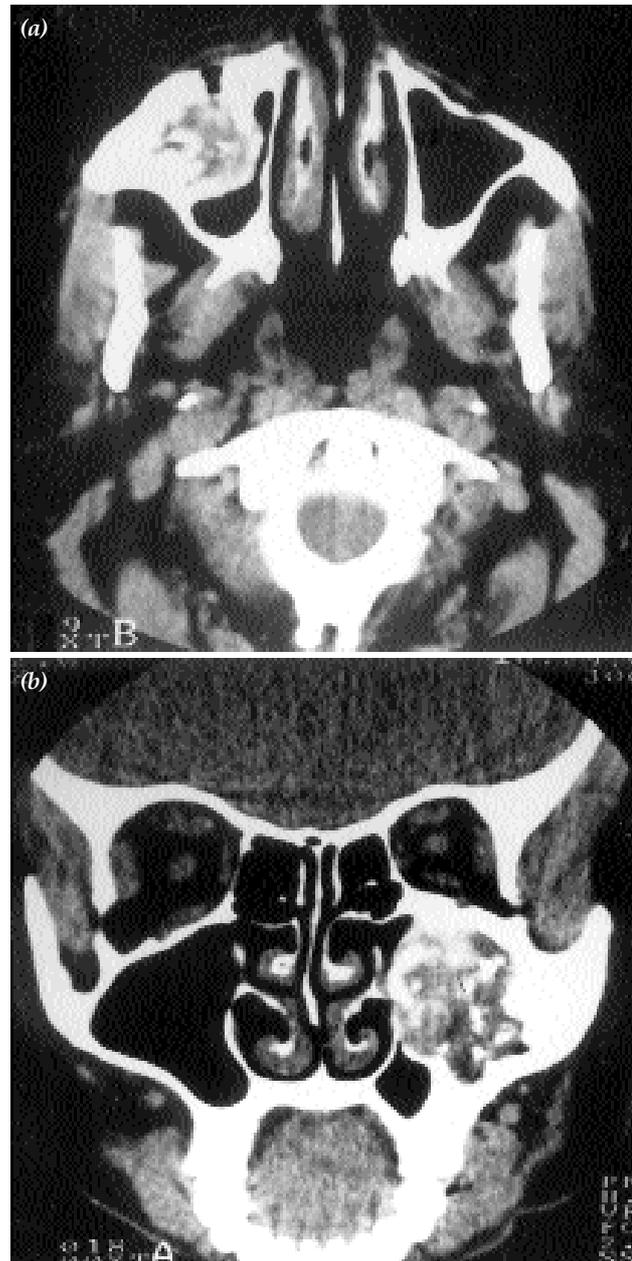


Fig. 2 - (a) Axial computed tomography scan showing a heterogeneous mass leading to expansion of the right maxillary sinus and thickening of the maxillary bone. (b) A coronal view showed both central and peripheral calcifications in the matrix.

Monostotic FD is the most common form.<sup>[1]</sup> Its frequency in the head and neck region is in the following order: the maxilla, frontal bone, mandible, and parietal and occipital bones.<sup>[6]</sup> The polyostotic form occurs less frequently, half or more of the cases having involvement of the craniofacial bones.<sup>[7]</sup> Though FD of the craniofacial bones are frequently reported, reports on FD involving the paranasal sinuses are rare.<sup>[2,3,8,9]</sup> As FD is usually secondary to extension of the disease from adjacent bones, it is rarely confined to the sinuses. The incidence of exclusive sinus involvement is not known due to its rarity and often asymptomatic nature.<sup>[1]</sup>

The diagnosis of FD involves a combination of clinical examination, radiographic studies, and histologic examination of the biopsy.<sup>[3]</sup>

In contrast to its benign and slowly growing course, FD of the maxillary sinus may become voluminous and invasive, causing considerable disfigurement, deformity, and dysfunction. Patients most commonly present to physicians with complaints of slowly progressive facial deformity or chronic symptoms of sinusitis, headache, nasal obstruction, epistaxis, proptosis, or visual impairment.<sup>[3,8,10,11]</sup> In the present case, radiological imaging showed that FD was confined to the maxillary sinus, almost filling the entire sinus cavity. It might have originated from the inner maxillary bone adjacent to the sinus.

Imaging studies show characteristic patterns in FD. Three main radiographic patterns have been observed in the craniofacial region: sclerotic, cystic, and pagetoid. The sclerotic form, with a homogeneously dense area or a classic ground-glass appearance, is easily recognized. The density of the lesion varies depending on the extent of osseous metaplasia. The cystic pattern usually represents a single, well-defined, and radiolucent lesion which is round or oval in shape, and frequently has a thin, sclerotic margin. Bony erosions and periosteal reactions are not usually associated with FD.<sup>[12]</sup> We observed a sclerotic pattern in our case and no bony erosions or periosteal reactions were detected.

Though conventional radiographic techniques enable a diagnosis of FD, CT scan is essential to establish the diagnosis, to plan an appropriate surgical procedure, to define the extent of the lesion, and to measure the growth rate.<sup>[8,13]</sup> Characteristics of FD on CT scans include an expansion of the

involved bone with a heterogeneous pattern of CT density and scattered or confluent islands of bone formation.<sup>[13]</sup> Most of the CT scans of the paranasal sinuses were found to exhibit a pagetoid or sclerotic appearance, both of which are easily recognized by the thickening of the bone.<sup>[8]</sup> Computed tomography may also provide precise information on the degree of ossification.<sup>[12]</sup>

Observation of a calcified maxillary sinus wall on CT scans, even when incomplete or expanded, may suggest a benign process including FD. Rarely, some benign tumors such as chondroblastoma and chondroma may present as partially calcified soft tissue masses, with calcific rims in the maxillary sinus. Thus, FD may be confused with osteosarcoma or chondrosarcoma unless the wall is evident on CT,<sup>[3,14]</sup> requiring histopathologic diagnosis to differentiate FD from malignancy. Histopathologic confirmation was made in our patient though clinical and radiologic findings did not suggest any pathologies other than FD.

With the help of this case illustration, it is recommended that FD originating from the maxillary sinus should be kept in mind in patients with long-term complaints of chronic sinusitis, nasal obstruction, or headaches, and that a detailed radiologic examination must be made before surgical planning.

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