Isolated fibrous dysplasia of the ethmoid sinus

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Abstract
Fibrous dysplasia is a benign fibro-osseous lesion progressing with one or more bone involvements in the skeleton. Even though it is a benign tumor, it may potentially transform into a malignant one. While the most frequently involved zones in the head–neck zone include the maxilla, mandible, parietal, occipital, and temporal zones, the involvement of the ethmoid bone is rather rare. This article presents the case of a female patient who was diagnosed with fibrous dysplasia in the right ethmoid sinus based on the paranasal computerized tomography scan that was taken due to the symptoms of pain around the right eye, pressure, and a feeling of being pushed.

Keywords
Fibrous dysplasia, paranasal sinuses, ethmoid bone

Introduction
Fibrous dysplasia (FD) is a benign disease that progresses slowly, with a partially known etiology in which the normal medullar bone is replaced by the abnormal fibro-osseous tissue.1 It was originally described in 1938 by Lichtenstein.2 Monostotic and polyostotic forms are defined, depending on whether one or more bones are affected, which are noted in 30% and 70% of patients, respectively. Malignant transformation is rare (0.5%) and is usually seen only in polyostotic cases.3 While the maxilla and mandible are most frequently involved in the head–neck zone, the ethmoid bone is rarely involved.3 The disease develops in adolescents and young adults. With FD, the bone grows in the form of a pain-free swelling.4 In computerized tomography (CT) scan, radiolucent and sclerotic zones are observed depending on the intensity of the fibrous and osseous tissues.5 For the treatment of FD, a benign lesion, total excision is a method that is preferred if the FD lesions located in the paranasal sinus have become symptomatic. An endoscopic sinus surgery is the first choice of treatment in such limited lesion to avoid a wound of external ethmoidectomy in the face.3

This article presents an 18-year-old female patient who was diagnosed with FD in the right ethmoid sinus based on the CT scan taken due to the symptoms of pain around the right eye and pressure.

Case report
This is a case report. Written informed consent was obtained from patient for publication in an international medical journal. Our institution does not require ethics approval for reporting individual cases.

An 18-year-old female patient presented to our clinic due to pain and pressure around her right eye, and her written informed consents were received. There were no findings after the anterior rhinoscopic examination performed on the patient. During the endoscopic nasal examination, a non-tender mass with a hard consistence and a smooth surface medializing the right middle concha medially and extending to the lamina papyracea laterally were found. On the paranasal CT scan, a lobular mass sized approximately 2.5 × 3.6 cm with the same density as the bone tissue was observed which completely covered the right ethmoid cells, displaced the nasal septum to the left, extended medially to the septum and laterally to the lamina papyracea, and was characterized by significantly hyper-dense and sporadically hypo-dense areas

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The patient underwent right endoscopic sinus surgery under general anesthesia. The mass was removed from the adjacent tissues with the aid of curettage and drilling; after that, it was removed in two pieces from the nostrils (Figure 3). Following the surgery, no complications developed in the follow-up visit a week later. In the post-operative period on 1-year follow-up, the patient received intermittent nasal endoscopic examinations. The patient, who is clinically asymptomatic, is under follow-up every 6 months.

During the histopathological examination of the material, it was observed that an irregular osseous trabeculae, which had no osteoblastic activities, dispersed inside the fibrous stroma. The diagnosis of monostotic FD was obtained in the light of the current clinical, radiological, and histopathological findings (Figure 4).

**Discussion**

FD is a benign disease that progresses slowly. The disease is physiopathologically underpinned by the replacement of the normal bone by fibrous tissue. The lesion starting at the medullary bone expands and involved the adjacent cortex. The bone trabeculae become irregularly placed, and a bone tissue with no internal lamellar structures develops. It is believed that a gene mutation (especially GNAS1) is responsible in its etiology. The disease onset is generally in the childhood age, the frequency gradually increases during puberty and adolescence, while the incidence is decreased in the early adulthood. FD accounts for 2.5% of bone tumors, whereas it accounts for 7.5% of the benign bone tumors. The head–neck zone is involved in 25% of the patients. In the head–neck zone, the most frequently involved areas are the maxilla and mandible.

CT scan generally is recommended as first choice in the FD. At the same time, magnetic resonance imaging (MRI) is beneficial for the assessment of soft tissue components. T2-weighted images in patients with FD reveal low signal from the involved bones, while ossifying fibromas give high signal. While the maxilla and mandible are involved most frequently in the head–neck zone, the ethmoid bone is rarely involved. However, Lustig et al. reported in a series of 15 years that the ethmoid bone involvement was the most frequent one (71%) among 21 patients in total. This article presented an 18-year-old monostotic FD case. The most
frequently observed clinical findings include atypical pain in the head and face as well as sinusitis-related symptoms. In advanced cases, changes due to the pressure of vital structures such as proptosis, diplopia, and visual changes may be observed depending on the location and orientation of the lesion.\textsuperscript{4} In our case, no proptosis, diplopia, and visual changes were observed. There was only a complaint about a feeling of pressure and pain in the right eye.

It is characteristic of FD to have a ground glass appearance in the CT scan and to have no definitive demarcation lines around the lesion. In our case, the paranasal CT scan showed a lobulated mass containing amorph-looking necrotic areas, which extended up to lamina papyracea in the right ethmoid cells, displaced the nasal septum toward the left, and extended up to the skull base. The definitive diagnosis of FD is made after a histopathological examination. In the histopathological examination, irregular trabeculae of the spongious bone as well as fibrous connective tissue, that is, stroma, are observed.\textsuperscript{5}

The external approaches most frequently used to treat paranasal sinus FDs include the Caldwell Luc technique, lateral rhinotomy, external ethmoidectomy, and craniofacial resection. Recently, more conservative trans-nasal endoscopic approaches have also been implemented for the masses limited to the ethmoid.\textsuperscript{3} In cases where the mass cannot be entirely excised, the generally preferred approach is to remove the lesion in the zones where pressures are present.\textsuperscript{3} In our patient, a surgical excision with an endoscopic approach was performed since the adjacent bone tissues caused pressure and the patient had pain, feeling of being pushed, and pressure around the right eye. As seen in the tomography scan, the lesion had the shape of a structure that was very hard and extended to the skull base. The patient underwent right endoscopic sinus surgery under general anesthesia. The mass was removed in two parts using the drill. Following the operation, no deformities developed and no recurrences were observed in the follow-up.

FD is a fibro-osseous lesion that is benign in nature, and the involvement of ethmoid bone is rarely reported. This situation is due to the fact that the ethmoid sinus is a zone that may be overlooked during the normal examination and conventional radiological procedures, unless the disease is complicated. The literature data and clinical experience emphasize that surgical treatment is required if the FD lesions located in the paranasal sinus have become symptomatic and caused loss of function and cosmetic deformities. The widely accepted protocol for treatment is to avoid radical surgery, which may cause larger defects and losses of function, and to adopt a conservative approach given that the disease has a benign progress. In cases where the mass cannot be excised en bloc, the recommended approach is to excise the parts that cause pressure in small parts to the possible extent.

Declaration of conflicting interests
The authors declare that there is no conflict of interest.

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