

Case Report

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Fibrous Dysplasia of the Ethmoid Sinus

Although craniofacial bone is the second common site of fibrous dysplasia involvement, it is rarely found in the paranasal sinus. Among fibrous dysplasia of the head and neck, the maxilla and mandible are the most frequent sites to be involved. Fibrous dysplasia becomes dormant in adolescence and early adult life and is more common in female. It is one of the fibrous osseous lesions and should be differentiated from osteoma and ossifying fibroma. Radiographically, fibrous dysplasia showed "groundglass" bone appearance on CT scans with bone window. Histopathologically, it presents woven-type bone embedded in a cellular fibrous stroma without osteoblastic rimming. We presented a case of 25-year-old female with fibrous dysplasia in her right side ethmoid sinus. She visited to us with the chief complaint of right side headache since adolescence. The lesion was removed by endoscopic sinus surgery and pathology proved fibrous dysplasia. The patient was free of headache after operation. The advance of endoscopic sinus surgical technique, makes it an optimal method for the pathological diagnosis and treatment to avoid the cosmetic problems caused by external approach in limited paranasal sinus fibrous osseous lesions.

Fibrous dysplasia is one of fibrous osseous lesions of bone. It most occurs in the long bone, and craniofacial bone is the second common site of involvement.¹ However, fibrous dysplasia of paranasal sinuses is rare.² There are only 6 reported cases confined to ethmoid sinus in the English literature.³⁻⁷ We present a case of fibrous dysplasia in right ethmoid sinus treated by endoscopic sinus surgery. The differentiations from other fibrous osseous lesions were discussed.

CASE REPORT

A 25-year-old female presented to the neurologic institute with the chief complaint of right headache since she was 19 years old. The headache was not associated with aura phenomenon. No nausea or vomiting occurred during the attack and the menstruation did not worsen the symptom. However, the headache was provoked when she was in a high altitude place. Computed tomographic scans of the brain, without contrast, revealed a lesion in the right-side ethmoid sinus. She was then referred to our

clinic for sinonasal evaluation. The physical examinations found neither facial deformity, nor eye movement limitation. She denied any history of purulent rhinorrhea, foul smelling in nose or posterior nasal dripping. A rigid sinonasal endoscopic examination did not demonstrate any mucopus or abnormal secretion in the nasal cavity. There was no deformity or displacement of anatomic structure in the right nasal cavity. Computed tomographic scans of the paranasal sinuses revealed a heterogeneous low-density lesion with some calcification spots in the right ethmoid sinus. The bony density surrounding the lesion was increased (Fig. 1). There was no lesion in other sinonasal cavities. The lamina papyracea of the right ethmoid sinus was intact. Fungal sinusitis was suspected initially. Under general anesthesia, an endoscopic sinus surgery was performed. The bony wall of the lesion was removed using a microdrill and the soft tissue content was then removed totally.

Under hematoxylin-eosin stain and light microscopy, the specimen showed haphazard woven bones embedded in fibrous tissue without osteoblastic rimming around the woven bone. It was a picture of fibrous

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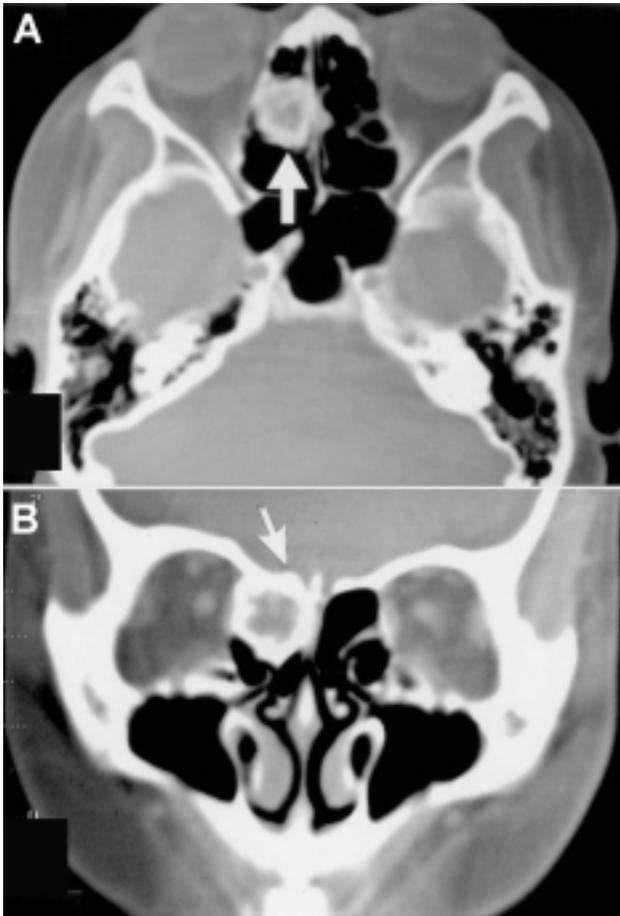


Fig. 1. Computed tomographic scans of the paranasal sinuses revealed a heterogeneous low-density lesion with some calcification spots in the right ethmoid sinus. (**A**: axial section, **B**: coronal section. Arrow).

dysplasia (Fig. 2). The patient exhibited an excellent postoperative recovery without post-operative bleeding or other complications, and was discharged 3 days later. Eight months subsequent to surgery, the patient has progressed well and is free of headache clinically with no evidence of tumor recurrence at our follow-up.

DISCUSSION

Fibrous dysplasia is a benign pathologic condition of the bone in which fibrous tissues gradually expand and replace the normal bone.⁸ The disease usually begins in childhood and progresses throughout puberty and adolescence, then becomes dormant in early adult life.

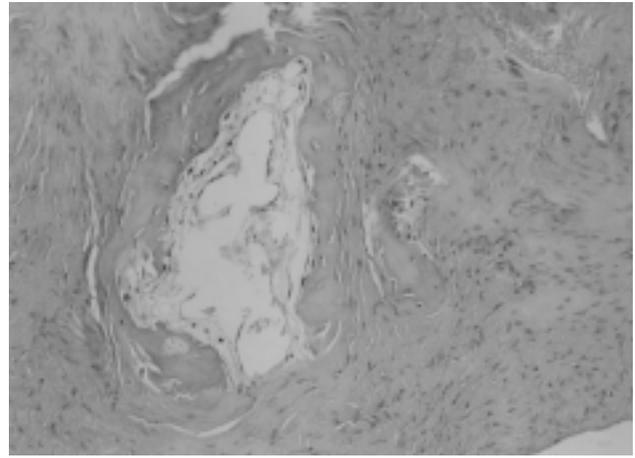


Fig. 2. The specimen showed haphazard woven bones embedded in fibrous tissue without osteoblastic rimming around the woven bone. (H&E stain, 200).

About 75% of the cases are found under the age of 30 years.⁹ 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.¹⁰

Following the ribs and long bones, craniofacial bones are the second most common site of involvement and comprise 25% of the cases.¹¹ Among fibrous dysplasia of the head and neck, the maxilla and mandible are the most frequent sites to be involved, followed by the frontal, parietal and occipital bones.¹⁰ However, fibrous dysplasia of the paranasal sinuses is very rare.^{2,3} It is usually secondary to the extension of the disease from adjacent bones.² Most commonly, fibrous dysplasia is asymptomatic until there is encroachment upon adjacent vital structures. Facial asymmetry is the most common sign of fibrous dysplasia in the head and neck, followed by pain, ocular proptosis and neurological changes.¹² In cases reported to confine to ethmoid sinus,³⁻⁷ 4 showed ocular symptoms and 2 had headache.

The “groundglass” bone appearance on CT scans with bone window is the most useful radiographic sign for the diagnosis of fibrous dysplasia.¹ The histopathological findings present irregular spicules of a woven-type bone embedded in a cellular fibrous stroma. There is no osteoblastic rimming around the woven bone.¹³ Treatment is aimed at correcting or preventing functional

problems and achieving normal facial aesthetics.

Osteomas, ossifying fibromas and fibrous dysplasia comprise a set of benign neoplasms as fibrous osseous lesions.¹⁴ Osteomas are the most common benign tumors occurring in the paranasal sinuses. The frontoethmoid region is the most common site of origin. There are three histological types of osteomas: an ivory type composed of dense cortical bone, a spongy form composed of cancellous bone, and the mixed type. Fibrous dysplasia and ossifying fibromas are less common lesions. Radiographically, ossifying fibroma is a monostotic, sharply circumscribed lesion and shows an eggshell rim on X-ray radiolucency. Fibrous dysplasia is not well circumscribed, and its borders are difficult to define. Histopathologically, ossifying fibroma has islands of osteoid rimmed by osteoblasts to form lamellar bone, and the cellular fibrous stroma shows a parallel and whorl arrangement of the collagen and fibroblasts. Those characteristics are absent in fibrous dysplasia.¹³

Fibrous dysplasia of the ethmoid sinus is a rare disease. However, the "groundglass" appearance on CT scans of the paranasal sinuses on bone window and the symptom developing history could give us some clues to include this differential diagnosis. In limited lesion, an endoscopic sinus surgery could serve as an optimal method for the pathological diagnosis and treatment to avoid the cosmetic problems caused by external approach.

In the case reported herein, fungal sinusitis was impressed initially because the computed tomographic scans of paranasal sinuses demonstrated bony thickening of the lesion wall with calcification density within the lesion. However, since her headache symptom started when she was an adolescent, fibrous dysplasia should be considered. Otherwise, mucocoeles, and other fibrous osseous disorders should be included in the differential diagnosis. The probable causes of headache in this case were the compression of frontoethmoid complex by the tumor, or the expansion of tumor itself. An endoscopic sinus surgery is the first choice of treatment in such limited lesion to avoid a wound of external ethmoidectomy

in the face. The headache condition improved dramatically after our operation.

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