

## CASE REPORT

# Fibrous Dysplasia of Ethmoid

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## ABSTRACT

Fibrous dysplasia (FD) is a benign skeletal disorder in which abnormally overgrowing bony lesion replaces the normal bone. It can affect one bone (monostotic form) or multiple bones (polyostotic form). The craniofacial bones are involved in about 10% of subjects with monostotic FD. Among FD of the head and neck, the maxilla and mandible are the most frequent sites to be involved. However, its occurrence in the sinonasal tract is very rare. We report a case of a large monostotic FD of the ethmoid bone in a 29-year-old woman. The lesion was removed by lateral rhinotomy approach and pathology proved to be FD.

**Keywords:** Ethmoid bone, Fibrous dysplasia, Monostotic.

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## INTRODUCTION

Fibrous dysplasia (FD) of ethmoid is a rare entity and often presents a diagnostic challenge.<sup>1</sup> It 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.<sup>2</sup> It was originally described in 1938 by Lichtenstein.<sup>3</sup> Fibrous dysplasia accounts for 2.5% of bone tumors, whereas it accounts for 7.5% of benign bone tumors.<sup>4</sup> Monostotic and polyostotic forms are defined, depending on whether one or more bones are affected, which are noted in 30 to 70% of patients respectively. Few cases of FD of the ethmoid have been reported in the literature.<sup>5</sup>

We report a case of FD of ethmoid in a 29-year-old female who presented with large right-sided nasal mass, eroding the septum and reaching up to the cribriform area and fovea ethmoidalis.

## CASE REPORT

A 29-year-old female reported to the ear, nose, and throat outpatient department with complaints of gradually increasing right-sided nasal obstruction and swelling of

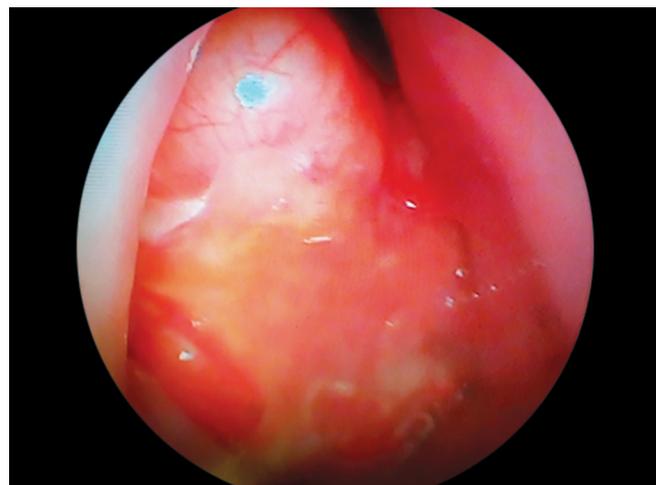
the right nasal dorsum for 2.5 years. The patient also had a history of hyposmia and change of voice. History of bleeding from nose, double vision, protrusion of eyeball, and orbital pain were not present. She denied any other significant past or family medical history.

Physical examination revealed external nasal deformity on the right side of the nasal dorsum. On anterior rhinoscopy, a smooth, pale, globular mass was seen filling the right nasal cavity. On probing, it was insensitive to touch, nonfriable, bony hard, and did not bleed on touch. Probe could be passed inferiorly and laterally. The septum was deviated to the opposite side.

On nasal endoscopic examination with 0° endoscope, a pale smooth mass was seen filling the right nasal cavity and pushing the septum toward the left side. Routine hematological investigations were normal (Fig. 1).

Computed tomography (CT) scan of the nose and paranasal sinus (PNS) showed a large well-defined soft tissue density mass of 3.7×3.7 cm in the right nasal cavity, eroding bony ethmoidal septae and causing displacement of nasal septum toward the left side (Figs 2 and 3). Multiple hyperdense foci suggestive of calcifications were seen within the mass. Superiorly, the mass reached up to the cribriform plate, and inferiorly, it was lying above the palatine process of the maxilla.

A biopsy was taken from the mass, which revealed features suggestive of FD. Under general anesthesia, right lateral rhinotomy was done, and the entire mass was removed *in toto* (Fig. 4). The nasal mass was sent for histopathological examination, which showed the presence of narrow curved bony trabeculae interspersed with fibrous tissue of moderate cellularity, without osteoblastic

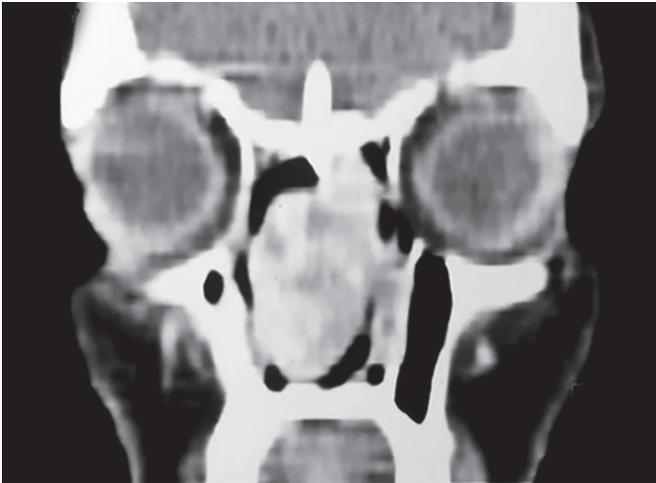


**Fig. 1:** Endoscopic view of the right nasal cavity showing the mass

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**Fig. 2:** Coronal CT scan of the nose and PNS showing a large well-defined soft tissue density mass in the right nasal cavity



**Fig. 3:** Axial CT scan of the nose and PNS showing a large well-defined soft tissue density mass in the right nasal cavity



**Fig. 4:** Macroscopic view of the specimen following excision by the lateral rhinotomy approach

rimming, confirmatory of benign fibro-osseous lesion, and FD (monostotic variant).

The patient exhibited an excellent postoperative recovery without any complications.

## DISCUSSION

Fibrous dysplasia is a benign pathologic condition of the bone in which fibrous tissues gradually expand and replace the normal bone.<sup>6</sup> The bone trabeculae become irregularly placed, and a bone tissue with no internal lamellar structures develops. The disease develops in adolescents and young adults, whereas the incidence is decreased in early adulthood.<sup>4</sup>

The most frequently observed clinical findings include atypical pain in the head and face as well as sinusitis-related symptoms.<sup>7</sup> 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.<sup>8</sup> In our case, no

visual complaints were observed, because the mass was limited only to the right nasal cavity.

The “ground-glass” bone appearance on CT scan with bone window is the most useful radiographic sign for the diagnosis of FD.<sup>2</sup> Radiolucent and sclerotic zones are observed depending on the intensity of the fibrous and osseous tissues.<sup>9</sup> At the same time, magnetic resonance imaging is beneficial for the assessment of soft tissue components. T2-weighted images in patients with FD reveal low signal from the involved bones, whereas ossifying fibromas give high signal.<sup>7</sup>

For the treatment of FD, a benign lesion, total excision is a method, i.e., preferred if the FD lesions located in the PNS have become symptomatic. The definitive diagnosis of FD is made after a histopathological examination. In the histopathological examination, irregular trabeculae of the spongy bone as well as fibrous connective tissue, that is, stroma, are observed.<sup>9</sup> There is no osteoblastic rimming around the woven bone.<sup>1</sup>

The external approaches most frequently used to treat FDs of PNS include the Caldwell-Luc technique, lateral rhinotomy, external ethmoidectomy, and craniofacial resection. Recently, more conservative transnasal endoscopic approaches have also been implemented for the masses limited to the ethmoid.<sup>10</sup> In our patient, an external approach was preferred due to large size of the mass, which would have been difficult to remove by the endoscopic approach. Surgical excision with a right lateral rhinotomy approach under general anesthesia was performed.

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