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Nasal Obstruction due to Fibrous Dysplasia Invading Inferior and Middle Turbinates: A Case Report and Literature Review

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중, 하비갑개를 침범하는 섬유 이형성증에 의해 발생한 코 막힘

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Fibrous dysplasia (FD) is a rare type of fibro-osseous lesion characterized by progressive replacement of normal bone with immature tissue. The involvement of craniofacial bones is reported in 10% of FD cases, while the involvement of sinonasal cavity is extremely rare. We report a unique case of FD in which the primary complaint was nasal obstruction. As FD cases involving the turbinate bones are very rare, we also reviewed all reported cases of FD involving the inferior or middle turbinates. Based on our experience and a review of the relevant literature, we conclude that inferior and/or middle turbinectomy via endoscopic approach and septoplasty can improve nasal symptoms.

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Key Words Endoscopic sinus surgery · Fibrous dysplasia · Sinonasal neoplasm.

Introduction

Fibrous dysplasia (FD) is a rare type of fibro-osseous lesion characterized by fibroblastic proliferation and progressive substitution of the normal bone with fibrotic disorganized bony trabeculae.[1)] Although several etiological theories, such as neoplastic degeneration and genetic or embryologic modification, have been proposed, the exact pathogenesis of FD is still unknown.[2,3)] 70% of all FD cases are of the monostotic form, and only 10% of these involve the craniofacial bones. 30% of all FD cases are of the polyostotic form, and more than 50% of these involve the craniofacial bones.[4,5)] The polyostotic form may be associated with McCune-Albright syndrome, which is the most severe form of polyostotic FD. As FD has a tendency to affect membranous bones such as the femur or tibia, involvement of the sinonasal cavity is rare, and involvement of the inferior and/or middle turbinate bones is extremely rare. In this report, we describe a case of severe FD involving the right inferior and middle turbinates and the almost-complete obstruction of the right nasal cavity. We also review previously-reported cases of FD-induced nasal obstruction symptoms with inferior and/or middle turbinate involvement.

Case

A 42-year-old woman presented with nasal obstruction that had persisted for at least five years. The patient had no additional complaints of nasal symptoms, including rhinorrhea, sneezing, itching, or hyposmia. She reported no history of concurrent medical disease, except for fibroadenoma of the right breast. Endoscopic examination revealed hypertrophy

of the inferior and middle turbinates of the right nasal cavity; severe septal deviation to the left nasal cavity was also noted. A computed tomography (CT) scan showed that fibrous dysplasia, involving the right frontal bone, sphenoid bone, and nasal cavity structures, had obstructed nearly the entire right nasal cavity (Fig. 1A and B). Laboratory test results, including serum calcium and alkaline phosphatase levels, were normal. As the right side, nasal cavity was nearly obstructed and the patient's symptoms were very severe; we partially removed the right inferior middle turbinate bone using a drill and bone rongeur via a navigation-assisted endoscopic approach. Conservative septoplasty to correct the lower septal area was also performed. The overlying mucosa was saved. Post-operative CT and endoscopic imaging showed a widened right nasal cavity (Fig. 1C and D). The patient's subjective symptoms were also improved, and remnant mucosa was well preserved. There were no additional complications or deterioration of symptoms over a one-year follow-up period.

Discussion

Involvement of the craniofacial and paranasal sinuses in FD is rare,[6,7] and is associated with symptoms atypical for FD, including headache, epistaxis, nasal obstruction, and sinus obstructions leading to recurrent infectious conditions. We investigated all FD cases reported to involve the inferior or middle turbinate bones, and found a total of five FD cases involving the middle turbinate in addition to four cases of FD involving the inferior turbinate. Among the five cases of FD involving the middle turbinate, we could access the full English text of four reports.[3,8-10] Based on a review of those cases, FD involving the middle turbinate was predominantly observed in female patients, whose ages ranged from 14 to 64 years. Although it has been reported that FD usually presents in the first two decades,[11] our case and the review of other case reports suggest that age is not a reliable factor for the diagnosis of FD. One male patient presented with bronchial asthma, aspirin intolerance, and allergy to grass pollen, a Widal syndrome. Another patient showed a septal deviation to the contralateral side, similarly to our case. Three patients underwent resection of the middle turbinate via an endoscopic approach to reduce their nasal symptoms. A patient with McCune-Albright syndrome in the form of a unique polyostotic type of FD involving the middle turbinate was conservatively

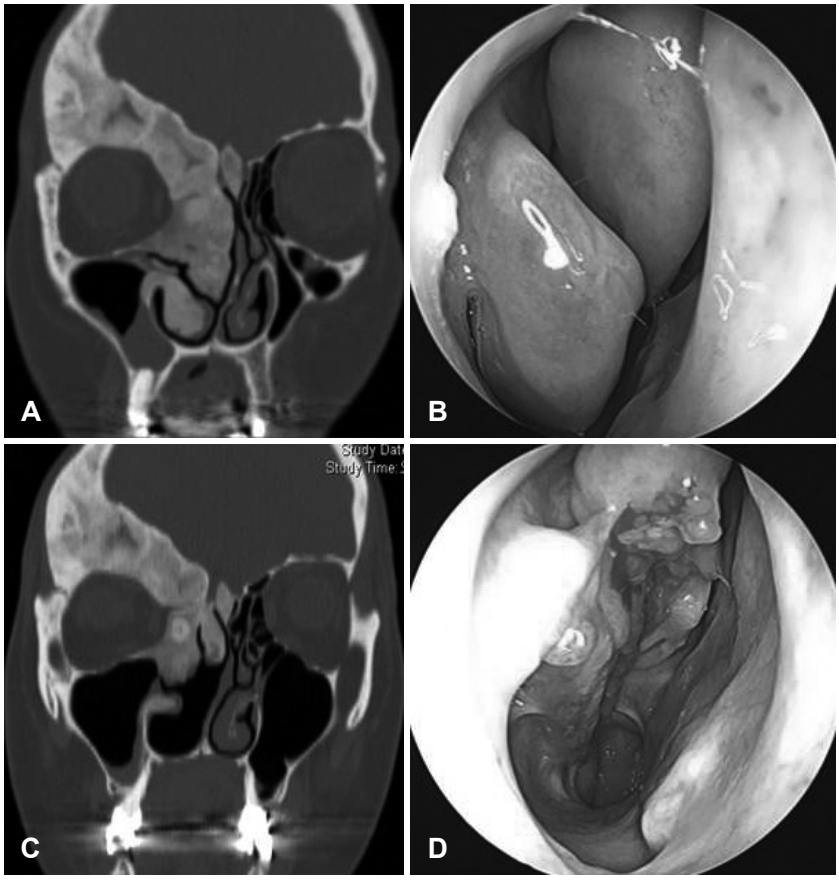

Fig. 1. Pre-operative CT image of the patient. Fibrous dysplasia invading inferior, middle turbinates and frontal bone was noted (A). Pre-operative endoscopic findings in the patient. The enlarged inferior middle turbinate was nearly obstructed in the right nasal cavity, and the septum was deviated to the contralateral side (B). Post-operative CT image of the patient. Partial resection of the right inferior middle turbinates and conservative septoplasty were performed (C). Post-operative endoscopic findings in the patient. The right nasal cavity was widened, and the overlying mucosa was well preserved (D).

managed without surgery. In all cases, there was no reported aggravation of the disease throughout the duration of the follow-up period (Table 1). Although we did not include it in Table 1, we found an interesting case report of a patient who was diagnosed with FD of the middle turbinate pre-operatively, with the diagnosis changed to ossifying fibroma after pathologic review. This suggests that the accurate diagnosis of fibro-osseous lesions is complicated in some cases, and that radiologic, pathologic, and endocrinological evaluations should be considered.

Among the four cases of FD involving the inferior turbinate, we could access the full English text of three reports.[1,12,13] Only one patient was male, and was under treatment for hematologic malignancies. This patient also suffered from pansinusitis, as revealed by CT examination. The management of sinusitis or FD was not clearly described for this case. The other two patients were females aged 29 and 68 years. Two female patients were surgically managed via endoscopic resection of the inferior turbinate. Septoplasty was performed in one patient. In all cases, there was no reported aggravation of the disease throughout the duration of the follow-up period (Table 2).

Based on our case and review of the literature, we suggest that patients whose symptoms were not specific, and maintained normal nasal mucosa should be suspected for underlying diseases such as FD. And conservative surgical management should be considered, because it was very effective for the improvement of subjective symptoms. We found that in patients with suspected FD, imaging studies should be performed to rule out the possibility of polyostotic FD. Endocrinological studies should be performed to rule out concurrent medical conditions, and physical examinations for diagnostic indicators such as café au lait spots should not be ignored. Furthermore, pathologic findings should be carefully reviewed, because different types of fibro-osseous lesions may share clinical and radiologic features.[14] In all cases, the nasal mucosa overlying FD-affected tissue were in normal condition, and the initial clinical impression was of inferior or middle turbinate hypertrophy. Therefore, in the case of severe asymmetric turbinate hypertrophy, CT scans should be performed to rule out the possibility of monostotic FD affecting the nasal cavity. In all patients, endoscopic surgical removal of affected turbinate bones relieved patient symptoms and did

Table 1. Review of the literature related to FD with involvement of the M.T

Researcher	Age/ sex	Chief complaint	Concurrent disease	Physical findings	Treatment	F/u (year)	Aggravation of symptoms
Saetti, et al.[3]	28/M	Nasal obstruction	Widal syndrome	M.T hypertrophy (L) Polyposis (R)	Endoscopic resection of M.T	2	No
Uzun, et al.[10]	14/F	Nasal obstruction	McCune-Albright syndrome	M.T hypertrophy (R) Septal deviation	Observation	5	No
Stoeckli, et al.[9]	50/F	Diplopia (R)	Subperiosteal abscess of medial orbital wall (R)	M.T hypertrophy (R)	Endoscopic resection of M.T (Secondary approach after management of subperiosteal abscess)	1	No
LaBagnara, et al.[8]	64/F	None	Hypercalcemia	M.T hypertrophy (L)	Endoscopic resection of M.T	Not identified	Not identified

F/u: follow-up, M.T: middle turbinate, R: right, L: left, FD: fibrous dysplasia

Table 2. Review of the literature related to FD with involvement of the I.T

Researcher	Age/ sex	Chief complaint	Concurrent disease	Physical findings	Treatment	F/u (year)	Aggravation of symptoms
Alvarez Vázquez, et al.[12]	11/M	Nasal obstruction	Acute lymphoblastic leukemia	I.T hypertrophy (R)	Not identified	Not identified	Not identified
Park, et al.[1]	29/F	Nasal obstruction	None	I.T hypertrophy (R) Septal deviation	Endoscopic resection of I.T Septoplasty	2	No
Karligkiotis, et al.[13]	68/F	Nasal obstruction	None	I.T hypertrophy (R) Septal deviation	Endoscopic resection of I.T	1	No

F/u: follow-up, I.T: inferior turbinate, R: right, FD: fibrous dysplasia

not induce any complications. Interestingly, one study reported a case of FD of the middle turbinate causing periorbital abscesses that obstructed the middle meatus. Therefore, when a patient with FD experiences nasal symptoms, it is appropriate and effective to surgically resect the affected turbinate bone. Surgical access is via an endoscopic approach, and saving the overlying mucosa above the affected bone is important for minimization of post-operative nasal symptoms.

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