Introduction

Fibrous dysplasia (FD) is a slowly progressive, self-limiting, non-neoplastic fibro-osseous lesion. First described by Lichtenstein in 1938, it is characterized by transformation of normal bone into isomorphous fibrous tissue and poorly formed woven bone. This process may extend beyond normal bony boundaries and give rise to expansion, distortion, and structural weakness of the involved region. There are three clinical variations of FD: monostatic FD affecting a single bone and accounting for more than 70% of all cases; polyostatic FD (PFD) affecting multiple bones and accounting for 20%-30% of all cases; and McCune-Albright syndrome (MAS), in which PFD is associated with endocrine disorders and cutaneous hyperpigmentation. In addition to the above variations, there is a hereditary form of localized FD described by Jones that is called cherubism.

Paranasal sinus involvement in FD is infrequent, and the maxillary sinus is involved more than the ethmoid sinus. Involvement of the frontal sinus, sphenoid sinus, and middle turbinates is rare, with only sporadic cases reported in the literature. Nasal turbinates and especially the inferior turbinates are the least involved bones of the craniofacial region. To the best of our knowledge, only one case with McCune-Albright syndrome had FD of the inferior turbinates. Here, we report a rare case with FD of inferior and middle turbinates and review literature concerning FD of the craniofacial region.

Case report

A 14-year-old male suffering from facial asymmetry and headache for three years was admitted to the otorhinolaryngology polyclinic. Physical examination revealed mild facial deformity and a nontender bony projection of the right side of the frontal bone, displacing the eye globe slightly downwards. Axial and coronal computed tomography (CT) scans showed diffuse hyperostosis and bony expansion into the right frontal sinus. Osseous hypertrophy was seen in the right middle and inferior turbinates as well as in the anterior wall and at the base of the sella. The involved bones exhibited a ground glass appearance; however, no destruction or erosion in the cortical bone was evident (Figures 1, 2). Based on diagnostic nasal endoscopy, the middle and inferior turbinates seemed enlarged, and a whitish color change was present, attributable to hyperostosis of the bone (Figure 3).

Physical examination revealed no café au lait spots. The patient had no history of precocious puberty, and his serum growth hormone levels were in normal limits. He also had no endocrine disorders and no lesions in his long bones.

Endonasal endoscopic biopsy obtained from the right inferior turbinate showed that the turbinate...
bone was spongiotic in nature, and the histopathological diagnosis was “fibro-osseous lesion, resembling FD.” Histopathological examination revealed fibrous stroma surrounding variable-shaped bony trabeculae enclosed by osteoblastic rimming. The stroma exhibited no mitotic figures or cytological atypia. Hypocellular proliferation of plump spindle cells with collagen production accompanies FD; the spindle cells usually are not arranged in any meaningful fashion but occasionally produce a stariform arrangement. The diagnostic feature is the production of woven bone (Figures 4, 5).

Because of the lack of major facial deformity, severe pain, or functional loss, we did not plan surgery. At present, the patient is on regular follow-up. He is now 17 years old, and after 33 months of follow-up, his craniofacial lesions have not enlarged. He still has no lesions in his other bones or any disorders of his other organ systems.

Discussion

The polyostotic form of FD involving multiple bones is observed in 20%-30% of patients, with craniofacial involvement in 50%-100% of these patients. Our patient had the polyostotic form of the disease, with craniofacial bone involvement. The lesion was situated in the frontal bone and filled the right frontal sinus completely. The maxillary sinus was not involved; however, the bony part of the middle turbinate was thickened and appeared sclerotic, and the inferior turbinate had the same appearance. In the literature, there is only one reported case of FD of the inferior turbinate accompanied by MAS, FD of the ethmoid sinus and middle turbinate, café au lait spots, and precocious puberty. MAS is defined as the triad of PFD, precocious puberty, and café au lait spots. The hypophysis, thyroid, parathyroid, and adrenal glands may be affected, and hypersecretions of growth hormone, prolactin, thyroid hormones, parathormone, and cortisol may cause various endocrine abnormalities as well as long bone involvement and fractures. Our patient had no café au lait spots, endocrinopathies, or abnormalities regarding other organ systems of his body.

A review of the literature shows that craniofacial FD primarily

Figure 1
Coronal CT showing diffuse hyperostosis in the right frontal sinus and the middle and inferior turbinates.

Figure 2
Axial CT showing diffuse hyperostosis and bony expansion into the right frontal sinus.

Figure 3
Endoscopic view of the right inferior and middle turbinates.

Figure 4
Bony trabeculae are surrounded by a dense fibrous stroma (H & E, × 100).

Figure 5
High-power view of fibrous dysplasia. Bony fragments and fibrous stroma are visible (H & E, × 200).
involves the maxillary sinus, the mandible, and the fronto-orbital region; however, it is less common in the ethmoid sinus, sphenoid sinus, and middle turbinate (Table 1). Only one other case has been reported with involvement of the inferior turbinate.

In 1957, Fries classified craniofacial FD into three types on the basis of conventional radiography: pagetoid (56%), sclerotic (23%), and cyst-like (21%). In the pagetoid type, radiodense and radiolucent areas (scattered island) are seen on CT whereas the ground glass appearance is evident in the sclerotic type. Our patient’s lesion fit the latter criterion.

The differential diagnosis of FD from ossifying fibroma is important because FD is a self-limiting disorder, while ossifying fibroma grows progressively and requires surgical resection. The differential diagnosis rests on radiological criteria after histopathologic analysis identifies a fibro-osseous lesion. The most significant feature distinguishing the ossifying fibroma from FD is the circumscribed nature of the ossifying fibroma. The histopathology in our case was reported as a “fibro-osseous lesion, resembling FD.” Because the CT revealed a ground glass appearance, the patient was diagnosed as having FD.

Treatment depends on the size and localization of the lesion and the symptoms. Small, non-progressive, asymptomatic lesions require no treatment. Indications for surgical intervention include severe deformity, pain, or functional interference, and surgery may consist of resection or curettage. Radiotherapy is not recommended because of its limited therapeutic effect and the risk of subsequent malignant transformation. Patients must be followed up regularly because of the risk of malignant transformation even in the absence of previous irradiation. We have not performed surgery in our case because of a lack of severe deformity, pain, or functional loss.

**Conclusion**

Paranasal sinus and nasal turbinate involvement is extremely rare in FD, and to our knowledge, our case with involvement of the inferior turbinate is only the second reported in the English-language literature.

**References**

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