Case Report

Fibrous Dysplasia of Paranasal Sinuses – An Atypical Presentation

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Abstract
Fibrous Dysplasia is a benign bone disease of unknown etiology. The involvement of the craniofacial skeleton is not uncommon but rarely involves the paranasal sinus. We report a case of fibrous dysplasia of paranasal sinuses, describing its unusual clinical presentation, radiological features, histopathological appearance and surgical management.

Introduction

Fibrous dysplasia is a fibro-osseous lesion where normal bone and marrow is replaced with fibrous tissue, resulting in formation of bone that is weak and prone to expansion. It represents 2% of the osseous tumours. Individual bone lesions typically manifest during the first few years of life and expand during childhood. The vast majority of clinically significant bone lesions are detectable by age of 10 years, with few new and almost no clinically significant bone lesions appearing after age 15 years and has a major preference for the white race and involves one-fourth of head and neck and is more common in females. Craniofacial presentation of fibrous dysplasia is usually cosmetic or due to mass effect on cranial structures resulting in following features - cranial asymmetry, facial deformity, nasal stuffiness, proptosis, visual impairment/unilateral blindness but in this case the presentation was different. The morbidity here is secondary to the location of the FD and its impingement on contiguous structures. The craniofacial involvement is difficult to treat due to the location of the lesion.

Case Report

A 61-year-old male patient came with chief complaints of frontal headache for 3 months duration and pain and redness in right eye for 1 week. He denied deficit of visual accuracy, watering and discharge from eyes, neither any complaints of otalgia, nasal obstruction, nasal discharge and toothache. The patient was a known case of diabetes and had been taking medications for that and his blood sugar level was controlled. There was mild proptosis of right eye. There was no facial asymmetry, the glasses worn by patient was according to his visual accuracy, the intraocular pressure was within normal limits. No other abnormality was detected on ophthalmoscopy. Systemic examination was clinically normal, and routine blood examinations were within normal limits. He was managed on OPD basis for iritis of right eye and for identifying the etiology behind mild proptosis of right eye, CECT paranasal sinuses was done. It showed ground glass opacity predominantly involving the right side, bilateral maxillary sinus, and obliterated right fronto-ethmoidal and bilateral sphenoid recess (Figure 1). He underwent nasal endoscopy and biopsy. Histopathology of excised tissue showed a benign fibro-osseous lesion formed by a hypercellular fibroblastic stroma consisting of spindle cells arranged in whorls with storiform pattern and also in loose sheets with intervening collagen interspersed amongst these was immature woven bone with Chinese letter configuration. These features were consistent with fibrous dysplasia. But since fibrous dysplasia is a benign bone disease and the patient’s age was 61 years, there was no surgical treatment required. He was continued with the treatment of iritis. Progress after treatment was on expected lines and recovery was complete by the time of discharge.

Discussion

The first author to describe the characteristic osseous lesion, also known as fibrous dysplasia, was VON RECKLINGHAUSEN in 1891, but it was LICHTENSTEIN, in 1938 who introduced the term Fibrous Osseous Dysplasia into the worldwide literature. There are two primary categories of the disease: monostotic fibrous dysplasia, that involves only one bone and represents 70% of the cases; and polyostotic fibrous dysplasia, that presents the involvement of several bones.

It affects the bones of the cranium and face in 3 ways:

1. As a monostotic lesion.
2. As one or more lesions of polyostotic disease.
3. As one or more lesions of Albright – McCune’s syndrome, in which the polyostotic lesions are disseminated and associated with extra-skeletal manifestations such as skin pigmentation and various endocrine manifestations.

The nature and etiology of fibrous dysplasia is obscure and its lesions tend to become inactive or stabilized as skeletal maturity is reached. Monostotic fibrous dysplasia is more common than the polyostotic variety.¹ It occurs in 70-80% of all cases of fibrous dysplasia. Although the maxilla and mandible are most commonly involved, maxillary sinus involvement is rare. The

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Received: 15.08.2016; Accepted: 25.04.2019
Craniofacial bones are more affected in the polyostotic form (50-100%) than in the monostotic form (20%). The disease is initially asymptomatic. The anterior craniofacial bones are more frequently involved than more lateral or posterior portions; sphenoid, frontal, maxillary, ethmoid bones > occipital, temporal bones. Extracranial involvement is rare. CT scan will demonstrate classic heterogeneous ground glass appearance with calcifications. The FD signals and symptoms depend on the location of the lesion(s) and the compressive effect in the adjacent structure as the tumor progresses slowly: facial asymmetry and deformity; pathological fractures; obstruction of the paranasal sinuses which generate recurrence infections, cysts and mucoceles, anosmia, headache, loss of visual accuracy for compression of the optic nerve; alteration of the ocular movements, descent, exophthalmia, squint, conductive hearing loss. CT scan is the investigation of choice for the study of the lesion(s), analysis of their extension and surgical preparation. Basically, three radiographic standards in the cranium fibrous dysplasia and facial bones are described: pategoid: that alternates the radiodense and radiolucent areas, sclerotic: homogeneously dense and cystic standard, with spherical or ovoid radiolucent area surrounded by dense limits. In the case reported, the lesion tomographic images presented a hyperdense standard intermixed by imprecise limits hypodense areas, which resulted in the classical aspect of “opaque glass”. Areas of low enhancement and cyst formation can be seen, which aids in differentiating the lesion from malignancy. Histopathologically, it presents woven-type bone embedded in a cellular fibrous stroma without osteoblastic rimming. The FD definitive diagnosis is made by the correlation of clinical, radiological and histopathological findings.

The FD differential diagnosis includes malignant (sarcoma, metastatic osteoblastic lesions) and benign lesions (ossifying fibroma, Paget’s disease, aneurismatic osseous cyst, cystic Cristeller Syndrome, ameloblastoma, osteochondroma, hyperthyroidism etc.)

The main factors that guide the FD approach are the- presence and the intensity of the symptoms, the tumor location and the patient’s age. The simple presence of the lesion does not justify surgical intervention. The main indications for surgical treatment of FD are the presence of significant clinical symptoms and the control of large aesthetic deformities. Because of the benign nature of lesion, the treatment must be relatively conservative with the main objective of preserving the function. In this case, we chose a conservative treatment. Radiotherapy is contraindicated not only because the tumor is radioresistant but also because of the probable increase of the capacity for the dysplasia into sarcomatous transformation. The patient reported in this case was 61 years old with a lesion that was reported to be non-progressive, which is unlike the natural history of the disease, where the growth corresponds to the period of skeletal growth. Besides, the involvement of the paranasal sinuses is very rare.

**Conclusion**

Fibrous dysplasia is a benign lesion, which rarely affects the head and neck region. Moreover, the involvement of the paranasal sinuses is rare. Fibrous Dysplasia is significant because it may affect facial and cranial bones and may cause deformities and dysfunctions. In spite of its benign nature, the signs and symptoms resulting from the compression of noble structures of the cranial base and orbit may generate diagnostic doubts as for the possibility of a malignant lesion.

**References**