Maxillary Sinus Fibrous Dysplasia

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ABSTRACT
The authors presented a case of maxillary sinus fibrous dysplasia developed in a thirteen-year-old boy. The disease was almost asymptomatic except for mild facial asymmetry and exophthalmos. The diagnosis was made on the basis of CT scans and MR imaging, and confirmed by histological study. The condition was classified as a non-aggressive form of the disease. No recurrence of the fibro-osseous lesion was found at postoperative one year.

KEY WORDS Fibrous dysplasia · Maxillary sinus.

INTRODUCTION
The bone lesion now known as fibrous dysplasia was originally described by Von Recklinghausen in as early as 1891. McCure and Bruch¹ described the clinical entity of this disease as distinctive from other bone abnormalities. The term “fibrous dysplasia” was introduced by Lichtenstein² in 1938. The disease may have one of three forms¹ (1) monostotic fibrous dysplasia, characterized by involvement of a single bone¹ (2) polyostotic fibrous dysplasia, characterized by multiple lesions of a number of bones, and (3) McCune-Albright syndrome, characterized by manifest changes in the skin and the endocrine system as well as by bone lesions.

The monostotic form makes up about 7% of all bone tumors and is the most frequently occurring benign lesion affecting the skull. Although only 10% of lesions in the craniofacial structures have been of the monostotic form, it is the most prevalent form of fibrous dysplasia in the head and neck." The maxilla and mandible are the most commonly affected cranio-maxillo-facial bones.

The object of this paper was to present the case report of a patient with maxillary sinus fibrous dysplasia whose clinical course was almost undetectable.

CASE REPORT
A thirteen-year-old boy was referred to us because of a painless swelling of the face and mild exophthalmos. No other otolaryngologic signs were present, and none were found in the boy’s clinical history or upon physical examination. General and neurological examinations did not show any positive findings. The results of visual acuity, ocular movement, visual field and ophthalmoscopic examinations were within normal limits. Classical radiographic examination revealed severe opacity of the right maxillary sinus with apparent disruption of its walls (Fig. 1). Computed tomography (CT) demonstrated a pronounced sclerotic mass (ground glass appearance) filling the lumen of the right maxillary sinus completely and expanding to the alveolar process, the orbital floor and the ipsilateral nasal bones (Fig. 2). MR imaging of the facial region showed extensive soft-tissue changes throughout the right maxillary bone (Fig. 3). Such findings suggested the sclerotic type of fibrous dysplasia.

The child underwent trephination of the right maxillary sinus. Almost complete obliteration of the sinus lumen by bone mass was found at surgery. Following the removal of this mass, a sinus cavum was created. Histological findings of the removed sinus bone mass confirmed fibrous dysplasia of the sclerotic type. Postoperative course was uneventful at postoperative one year.

DISCUSSION
Fibrous dysplasia is a disease of slow progress and poorly manifested symptomatology. Monostotic fibrous dysplasia is considered to be the result of disordered development of the bone-forming mesenchyme, although the mode of transmission (autosomal dominant or recessive) has not been elucidated yet." According to other theories, monostotic fibrous dysplasia develops secondary to an arrest of bone maturation. The disease develops most frequently during the first two decades
of life.

According to the literature, fibrous dysplasia of the paranasal sinuses has been reported to appear in single cases,\(^5\)\(^\text{(5)}\) and is thought to be considerably rare in series of patients.\(^8\) All authors stress the silent clinical course of the disease, which means that the disease is sometimes detected only after the occurrence of complications such as visual impairment. In their study of 34 patients with aggressive maxillary sinus fibrous dysplasia, Shapeero et al.\(^8\) stressed that all the cases, before definite diagnosis, were clinically suspected to be instances of maxillo-facial sarcoma. After surgical resection, there was recurrence of the disease in every case, with the disease being histologically proved to be fibrous dysplasia.

In the diagnosis of fibrous dysplasia, conventional radiography reveals most frequently various degrees of sinus opacification, with the degree depending upon varying content of connective and bone tissue, and it is difficult to differentiate the opacification from other types of bone lesions. The CT scan is a far more valid method of diagnosis, for both primary and recurring lesions. CT findings of a “ground glass” mass with calcifications surrounded by the maxillary sinus may suggest a diagnosis of aggressive fibrous dysplasia.\(^8\) Soft-tissue lesions and their prevalence may be differentiated more precisely by MR imaging.

Our case is particularly interesting due to the extremely silent clinical course of the disease, characterized as it was by mild facial asymmetry and inconspicuous exophthalmos. Following a CT scan and MR imaging, fibrous dysplasia of the maxillary sinus was suspected, and the suspicion was subsequently confirmed histologically. No regrowth of the tumor was found at postoperative one year.

REFERENCES


![Fig. 1. X-ray radiography showing severe opacification of the right maxillary sinus with disruption of its walls.](image1)

![Fig. 2. CT scan showing a pronounced sclerotic mass obliterating the right maxillary sinus. This tumor extended into the alveolar process, the orbital floor, and the ipsilateral nasal bones.](image2)

![Fig. 3. MR imaging showing extensive soft-tissue lesions of the right maxillary bone.](image3)


