Monostotic Fibrous Dysplasia Involving the Maxilla

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Abstract
Fibrous dysplasia is a skeletal developmental anomaly of bone-forming mesenchyme that manifests as a defect in osteoblastic differentiation and maturation. It is one of the most perplexing diseases of osseous tissue because it is a nonhereditary disorder of unknown cause. Here we present a case of monostotic fibrous dysplasia in the maxilla of a 12-year-old girl.

Introduction
The term ‘fibrous dysplasia’ was coined by Lichtenstein in 1938 (1) to describe a condition to which attention had been drawn by Hunter and Turnbell in 1931 (2). Fibrous dysplasia is defined as an asymptomatic regional alteration of bone in which the normal architecture is replaced by fibrous tissue and nonfunctional trabecular osseous structures; lesions may be monostotic or polyostotic, with or without associated disturbances (3).

In 1942, Lichtenstein and Jaffe described the various possible clinical manifestations of fibrous dysplasia from a study of 86 patients (4). Fibrous dysplasia has four different disease patterns: monostotic form, polyostotic form, craniofacial form, and cherubism.1 The monostotic form is seven times more common than the polyostotic variant (5). It is caused by a mutation or deletion of a gene that encodes for an intra-cytoplasmic transducer protein required for bone maturation (6).

The severity varies according to the time when this genetic defect occurs during embryogenesis. If it occurs before the sixth week of intra-uterine life, the condition will be more severe, involving multiple bones along with skin and endocrine glands. If the same genetic defect occurs slightly later during intra-uterine life, the condition will be more localized, affecting a single bone (6).

Case report
A 12-year-old female patient reported to the Department of Oral Medicine & Radiology with swelling on the right side of her face since birth. Initially, the swelling was small and gradually increased to attain its present size.

An extra-oral examination found a solitary swelling, present in the right middle third of the face, approximately 4×6 cm in size and oval in shape with diffused margins, extending superiorly and inferiorly from the right infraorbital rim to the right corner of the mouth and medio-laterally from the right lateral nasal fold to the outer canthus of eye (Fig. 1). The overlying skin appeared normal. The right nasolabial fold was obliterated. Infraorbital edema was present with no visible pulsation. On palpation, the swelling was stony hard in consistency, nontender, and immobile. Right and left submandibular lymph nodes were palpable, single, mobile, and firm, measuring about 1×1 cm and nontender.

On intraoral examination, the right labial and buccal maxillary vestibules were found to be obliterated, extending from the distal aspect of the lateral incisor to the maxillary tuberosity region (Fig. 2). Expansion of the buccal and palatal cortical plates was noticed. There was generalized gingival inflammation with bleeding on probing. A retained grade II mobile deciduous maxillary right second molar and a palatally erupted second premolar were present.

On radiographic investigation, the maxillary lateral topographic occlusal view revealed diffused radiopacity in
the periapical region of 16 and 17. The borders of the radiopacity were diffuse and extending to the mesial aspect of 55 (Fig. 3). Intraoral periapical radiographs revealed that the trabecular bone pattern was altered and 18 was found to be impacted (Fig. 4). An orthopantogram showed a dense trabecular pattern in the right maxillary posterior region with diffuse borders (Fig. 5). The paranasal sinus view showed right maxillary sinus obliteration with slight expansion of the maxilla on the right side (Fig. 6).

To clearly delineate the lesion, the patient underwent a computed tomography scan in which the coronal section showed the right maxillary involvement with maxillary sinus obliteration, the lesion extending superiorly and inferiorly from the right infraorbital rim to the alveolar process, and that the lesion was approx. 4x6 cm in size (Fig. 7). The axial section of the computed tomography scan showed right maxillary expansion extending from the lateral incisor to the third molar region (Fig. 8).

From a laboratory analysis of the patient’s blood, her serum calcium, phosphorus, and alkaline phosphatase levels were found to be normal.

A bone biopsy was performed and a hard tissue specimen
that was yellowish white in color was removed. The specimen was decalcified for 3 weeks, routinely processed, and stained with hematoxylin and eosin. The sections revealed irregularly shaped trabeculae of immature bone dispersed in a cellular, loosely arranged fibrous stroma. The bone trabeculae were not connected to each other (Figs. 9 and 10). Few of them assumed curvilinear shapes; the trabeculae of bone were not lined by osteoblasts. The fibrous tissue showed numerous fibroblasts. The bone trabeculae had been replaced by fibrous stroma.

The diagnosis of monostotic fibrous dysplasia was determined and surgical contouring for the esthetic correction was planned. The first surgery was performed under general anesthesia. The patient did not return for further follow-up.

Discussion

Fibrous dysplasia of bone is characterized by the

Fig. 6 PNS view showing the right maxillary sinus obliteration.

Fig. 7 Coronal section of CT scan showing the right maxillary involvement with maxillary sinus obliteration.

Fig. 8 Axial section of CT scan showing right maxillary expansion.

Fig. 9 Photomicrograph showing bone trabeculae dispersed in a loosely arranged fibrous stroma. (H&E; 5x)
replacement of normal bone and marrow by fibrous tissue, within which irregular trabeculae of woven bone are haphazardly distributed (7). Fibrous dysplasia is a developmental or possibly hamartomatous condition of unknown etiology. In over 80% of cases, it affects only one bone (monostotic); the jaws and skull are often individually affected by monostotic fibrous dysplasia (8). However, fibrous dysplasia may also be polyostotic and affect multiple bones. The polyostotic type may also be associated with ‘cafe-au-lait’ skin pigmentation and precocious puberty as in McCune-Albright syndrome, which almost exclusively affects young females. Monostotic fibrous dysplasia, although less serious than polyostotic fibrous dysplasia (9), may be of greater concern due to increased involvement of jaw bones, resulting in esthetic problems (4). Monostotic fibrous dysplasia occurs equally in males and females (in contrast to the 3:1 female to male ratio in polyostotic fibrous dysplasia) (9).

Monostotic fibrous dysplasia is more commonly newly diagnosed in children and young adults than in older persons (1), but in the cases reported by Zimmerman and associates, the mean age was found to be 27 years and according to Gardner and Halpert, the mean age was 34 years, while the patient’s age in the present case was 12 years.

After a combined statistical study of nine different series involving 104 total cases of fibrous dysplasia of the jaws in various populations, the mean age was found to be 25 years, with a male to female ratio of 9:11 and a maxilla to mandible involvement ratio of about 2:1 (10).

Involvement of the maxilla in monostotic fibrous dysplasia is relatively rare compared to other bones. In 1946, Schlumberger, in his study series of 67 cases of monostotic fibrous dysplasia, found that it most frequently occurs in the rib (43%), followed by femur (13%), tibia (11%), maxilla (10%), calvarium (7.46%), and mandible (2.98%) (1).

The first clinical sign of the disease is a painless swelling or bulging, similar to the present case in which the patient appeared with painless swelling that had present since birth with obliteration of the upper right buccal vestibule.

The radiographic appearance of fibrous dysplasia of the jaws is extremely variable. It has been categorized as either pagetoid, sclerotic, or cystic (9). Variations in the relative amount of bone to fibrous tissue contents influence the radiographic appearance. Regardless of their type, the lesions almost always have ill-defined borders that blend imperceptibly into the adjacent normal bone (9). A similar radiographic appearance was found in the present case; diffuse radiopacity appeared to blend with normal bone without clear borders of the lesion.

There is considerable variation in the histopathological features of monostotic fibrous dysplasia of the jaws. Irregular trabeculae of bone are scattered randomly throughout the lesion. Characteristically, some of these trabeculae are C-shaped or Chinese character-shaped (11, 12) as seen in the present case. These trabeculae are usually coarse woven bone but may also be lamellar (1).

The trabeculae in bone typically lack osteoblastic rimming similar to our finding, but often contain numerous osteocytes. The osseous components thus appear to arise directly from the fibrous stroma (6). Biochemical abnormalities are rarely seen in cases of monostotic fibrous dysplasia. Elevation of alkaline phosphatase is encountered in 70% of polyostotic fibrous dysplasia cases (9).

Treatment of fibrous dysplasia is exclusively surgical but is not always indicated. In the past, surgical intervention was delayed as long as possible until after puberty (11, 12) with the hope that the disease would become quiescent. Now it is recommended that surgery be performed as soon as the lesion becomes marked with progressive deformity, pain, or interference with function.

Simple contouring of facial or skull bones back to normal dimensions has proved to be quite effective. About 25% of the patients treated in this way will require several operations for recurrence of the bony enlargement (9).

Malignant degeneration has been reported in 0.5% of cases left untreated. This rate increases 400-fold in patients...
who have received radiotherapy. For the same reason, radiotherapy is definitely contraindicated in patients with fibrous dysplasia and long-term follow-up is mandatory (9).

Conclusion

Fibrous dysplasia may manifest as monostotic or polyostotic forms. Diagnosis of the polyostotic form may be easier due to extra-skeletal involvement, while the monostotic form is usually free of such features. The monostotic form is common in jaws, so an oral diagnostician must evaluate other skeletal involvement and features and perform tests for an accurate diagnosis.

References