Craniofacial Fibrous Dysplasia

Short Communication

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Abstract

Fibrous dysplasia (FD) is an osseous growth disorder, producing immature bone and characterized by the replacement of normal bone with fibro-osseous connective tissue. It is a bone dysplasia that has the potential to cause significant cosmetic and functional disturbances, particularly in the craniofacial skeleton. Craniofacial fibrous dysplasia is one of the three types of polyostotic fibrous dysplasia that can affect the bones of the craniofacial complex. In this article, we report a case of craniofacial FD which caused hemifacial swelling and orbital asymmetry with no significant neurological symptoms including visual disturbances.

Key words: • Polyostotic fibrous dysplasia •Craniofacial, abnormality • Dental, radiography

Case Report

The patient was a 19 year old man who referred to Mashhad university of medical sciences dental school, department of oral medicine, with a complaint of jaw expansion since one year ago. He had no pain and other neurological symptoms.

In clinical examination, we observed unilateral facial expansion and distinct orbital asymmetry (Figure1).

Intraoral examination revealed bony hard expansion in buccal side of molar, premolar, canine and lateral teeth areas of right side maxilla and mandible.

In panoramic view, right mandibular expansion including symphysis, body and ramus was noted (Figure 2). The lesion had opaque internal pattern with mottled appearance. Some lucent areas were detected which could be related to remaining radiolucent area or cystic degeneration raised in the lesion.

In periapical view, opaque lesion with ill-defined borders and ground glass appearance was observed. The lamina dura in apical region of mandibular second molar was indistinct. In occlusal view, mandibular expansion in buccal and lingual cortex with fusiform appearance was noticed (Figure 3). In posterior-anterior view, mottled opaque appearance of right frontal bone, opacification of ethmoid sinus and smaller size of right orbit in comparison to left orbit due to inferior displacement of superior orbital rim were spotted.

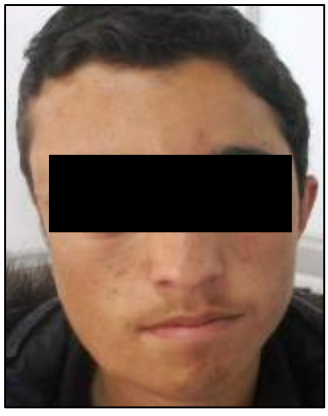

Figure1. Unilateral facial swelling and distinct orbital asymmetry are observed in clinical view.

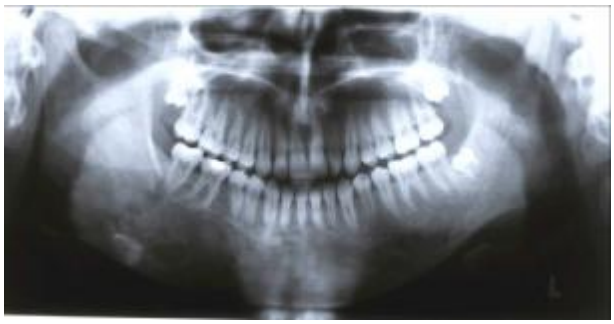

Figure 2. Mandibular expansion including symphysis, body and ramus is observed in Rt side of panoramic view. The lesion has opaque internal pattern with mottled appearance. Some lucent areas are identified.

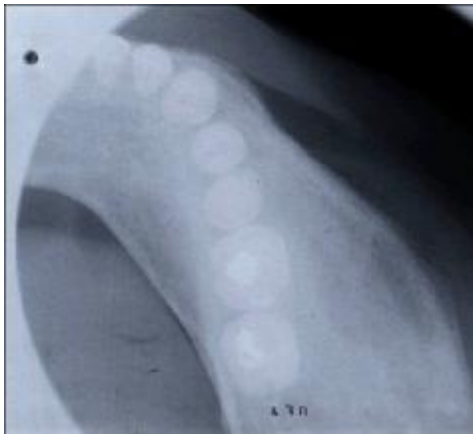

Figure 3. Fusiform expansion in buccal and lingual cortex of manible in occlusal view

Discussion

FD is one of the fibro-osseous lesions that affects the craniofacial region.[1] It is caused by primary developmental abnormality of the bone forming mesenchyme due to mutation which is characterized histologically by replacement of normal bone with fibro-osseous connective tissue exhibiting varying degrees of osseous metaplasia.[2,3] The clinical pattern of the disorder is variable depending on the stage of embryogenesis or development at which the mutation occurs, and the location of the mutation within the cell mass.[4] This includes involvement of a single bone or site (monostatic) or multiple sites (polyostotic).[5] Approximately 70% to 80% of FDs are monostotic.[6] In order, the most common sites are the ribs, femur, tibia, maxilla, and mandible.[8] The polyostotic type may be divided into 3 types: craniofacial FD, Lichtenstein-Jaffe type and Albright's syndrome. Craniofacial fibrous dysplasia is a disease that functionally and aesthetically affects the patients.[6]

Diagnosis of polyostotic FD is generally based on clinical symptoms and radiological images. This is not so with the monostotic FD, which requires bone biopsy. Bone biopsy is generally better to be avoided where the risk of pathological fracture and osteomyelitis is high.[7,8]

The most common clinical sign of craniofacial FD is swelling, and other signs and symptoms depend on the area of involvement .In this case, the chief complaint of patient was jaw expansion and no other symptoms including visual disturbances were observed. The most commonly involved bones of the craniofacial skeleton are maxilla and frontal bones[6] which were detected in this case. Radiographic features of opaque ground glass appearance with ill-defined borders, the characteristic signs of superior displacement of inferior alveolar nerve canal as well as expansion of mandible, maxilla and frontal bones confirmed the diagnosis of craniofacial fibrous dysplasia. Cystic change may develop in areas affected by FD[9], which was observed in panoramic view in this case and may correspond to cystic degenerations. Indistinct appearance of lamina dura due to replacement of normal bone may be one of the diagnostic signs of fibrous dysplasia[10] which we observed in periapical view. The patient was asymptomatic and he was not concerned about cosmetic problems so we decided to follow him 6 month later.

Conclusion

This case with its radiographic features and clinical presentation accumulates to the reports of craniofacial polyostotic FD which is an uncommon form of fibrous dysplasia. Accordingly, having possible complications and follow up would be of great importance after its diagnosis.

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