



FIBROUS DYSPLASIA: MISTAKEN AS RADICULAR CYST - A CASE REPORT

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<p>Article Info Received 17/08/2014 Revised 02/09/2014 Accepted 12/09/2014</p> <p>Key words: Radicular cyst, Monostotic fibrous Dysplasia.</p>	<p>ABSTRACT A patient reported with firm swelling in maxillary anterior region. Patient also had history of trauma in same region around 10-15 years back. Radiographs revealed well defined periapical radiolucency at the apical region of maxillary central incisors and were diagnosed as radicular cyst secondary to trauma; but histopathology reports revealed it to be monostotic fibrous dysplasia.</p>
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INTRODUCTION

Fibrous dysplasia (FD) is a benign disease of bone that was originally described by Lichtenstein more than sixty years ago. FD is a developmental dysplastic disorder of bone in which the normal bone matrix is replaced by fibroblastic proliferation.

Lesions contain irregular trabeculae of partially calcified osteoid. Some believe that the immature woven bone is formed directly from abnormal fibrous connective tissue that is unable to form mature lamellar bone, hence the term dysplasia.¹ Its etiology has been linked with a mutation in the Gs α gene that is located at chromosome 20q13.2-13.3. The mutation was first identified in patients with McCuneAlbright syndrome but was later demonstrated in the lesions of patients suffering from either monostotic or polyostotic FD. All cells that arise from the mutated cells manifest the dysplastic features [1].

CASE REPORT

A patient reported to the outpatient department with the chief complaint of pain & swelling in the upper front region of her face since the past 2months (Figure 1).

Patient first had history of trauma in childhood but did not have any fracture or avulsion of tooth at that time. Patient now mentions that her teeth have shifted close together since 15 days. Extra oral examination revealed a slight swelling in the anterior upper jaw region: lips were found to be competent.

On intra oral examination, a swelling seen extending in maxillary anterior labial vestibule extending mesiodistally from canine on left side to canine on right side and superoinferiorly from marginal gingiva through the entire labial vestibule.

The overlying mucosa appears slightly stretched. The mucosa shows no erythema, ulceration or sinus tract associated with the area of chief complaint. (Figure 2) On palpation, inspeactory findings were confirmed. The swelling was approximately 3cmX4cm, roughly oval in shape. The swelling was firm to hard in consistency. The surface of the swelling was smooth with no fluctuation noted (Figure 2). On basis of history recording and clinical findings a provisional diagnosis of Radicular cyst secondary to trauma with 11, 21 were given.



Radiographic study included intra oral periapical radiograph with 11, 21 and maxillary occlusal radiography and a OPG was taken. (Figure 3, 4, 5). An ill-defined radiolucency at the apical third region of 11 and 21 extending into the floor of nasal cavity was seen. It was associated with displacement of roots of 11 and 21.

Based on clinical and radiographic findings a diagnosis of radicular cyst secondary to trauma was given. After surgical excision of the lesion, specimen was obtained which was sent for histopathologic evaluation. The specimen was round to oval in shape, whitish in color measuring about 3 by 2.5 by 2 cm. It was firm in consistency .The surface was smooth with irregularity at one end, and the cut surface was smooth (Figure 6).

Low power view shows fibro-cellular connective tissue stroma with areas of bone deposition & focal distribution of giant cells can be appreciated throughout the stroma (Figure 7).

High power view of bone deposition. Showing woven type of bone with large osteocytic spaces, Scattered, irregularly shaped trabeculae of woven bone are seen throughout the stroma. The trabeculae tend to be delicate and curvilinear and have been likened to Chinese script-writing. Osteoblastic rimming is seen along the borders of bone formation (Figure 8).

Based on histopathology findings, final diagnosis of fibrous dysplasia of monostotic type was made.

Figure 1. Swelling seen in relation to front of upper jaw



Figure 2. Intra oral picture showing swelling in anterior region of upper jaw



Figure 3. IOPA showing radiolucency

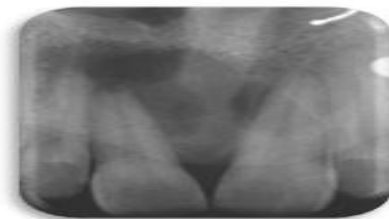


Figure 4. Displacement of roots seen

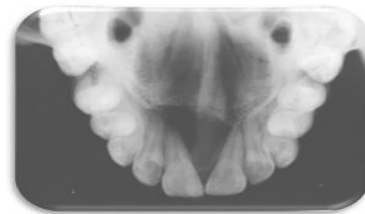


Figure 5. Radiolucency extending till the floor of the nasal cavity



Figure 6. Gross specimen showing smooth surface



Figure 7. Connective tissue stroma showing areas of bone distribution

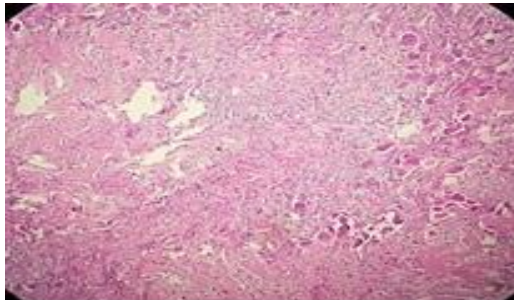
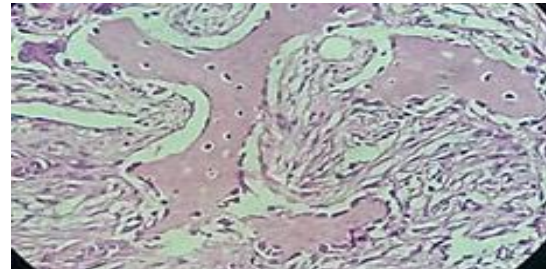


Figure 8. Scattered, irregularly shaped trabeculae of woven bone seen



DISCUSSION

Fibrous dysplasia is a perplexing disease of bone, of unknown etiology, uncertain pathogenesis and diverse histology. It is often described as a hamartomatous malformation characterized by idiopathic arrest in the normal maturation of bone in the woven bone stage, interspersed with abnormal proliferation of fibrous connective tissue containing varying amounts of osteoid and osseous structures of presumably metaplastic nature [2].

There are two primary categories of the disease: monostotic fibrous dysplasia that involves only one bone and polyostotic fibrous dysplasia, which involves several bones. Jaffe–Lichtenstein syndrome is a variant of polyostotic fibrous dysplasia with café-au-lait pigmentation of the skin. A more severe form of the polyostotic form of fibrous dysplasia accompanied by endocrine disturbances of varying types in addition to skin pigmentation is called the McCune–Albright syndrome.

Monostotic fibrous dysplasia accounts to 80-85% of all fibrous dysplasias and jaws are most commonly affected sites. Monostotic fibrous dysplasia may occur at any age, but are most common in children and young adults [3].

FD usually presents with bone pains, deformities, recurrent pathological fractures of the affected site and sometimes associated with endocrine hyperfunction. There is no gender predilection for FD but it is more common in children and adolescents as compared to adults and older patients [4]. It has now become evident that fibrous dysplasia is a genetic disease caused by somatic activating mutation of the G α subunit of G protein-coupled receptor resulting in upregulation of cAMP. This leads to defects in differentiation of osteoblasts with subsequent production of abnormal bone in an abundant fibrous stroma. In addition there is an increased production of IL-6 by mutated stromal fibrous dysplastic cells that induce osteoclastic bone resorption [5].

Most common clinical presentation is a painless swelling of the affected area. The maxilla is more commonly involved than mandible. Teeth involved within the lesion may be firm but may be displaced by the bony mass. Involvement of mandible may result in expansion of buccal and lingual cortical plates and displacement of inferior alveolar canal is also seen. When maxilla is involved displacement of sinus floor is seen. Imaging modalities show increase density of base of the skull and is

one of the characteristic feature of fibrous dysplasia of the skull [6].

According to recent WHO classification of FOLs, OF is a well demarcated lesion composed of fibrocellular tissue and mineralized material of varying appearances [7].

Reed has defined the condition as an “arrest of bone maturation in woven bone with ossification resulting from metaplasia of a nonspecific fibro-osseous type”. Schlumberger, first reported single bone involvement by the disease process and described it as “monostotic fibrous dysplasia”. Presently, the terms “monostotic” (single bone) and “polyostotic” (multiple bones) are used to describe the extent of this condition in terms of number of bones involved. Monostotic fibrous dysplasia (MFD) constitutes about 70-80% of FD patients and is mostly seen in the second and third decade. Diagnosis of polyostotic FD is generally based on clinical symptoms and radiological images. In contrast, the monostotic FD requires bone biopsy [8].

Radiographically, the appearance of fibrous dysplasia will vary depending on the stage of development and quantity of bony matrix within the lesion. Thus the lesion is more radiolucent and well defined initially and gradually changes to a mottled, ill-defined radiopacity in the later stages. The radiological picture in fibrous dysplasia is very distinctive showing a thin bony cortex with well-defined borders and ground glass appearance. Three distinct patterns have been described by Panda et al. The pagetoid appearance on CT imaging is characterized by bone expansion and scattered islands of bone formation in a low-attenuation field. The sclerotic type has a homogeneous appearance with a ground-glass appearance. The cystic type appears as a well-defined low-attenuation lesion with a sclerotic margin. The differential diagnosis of fibrous dysplasias includes lesions like ossifying fibroma, Paget's disease; diffuse sclerosing osteomyelitis etc [9].

The treatment aims at preventing fractures, reducing pain and at re-establishing the bone facial contours. In most situations, it involves a surgical intervention, even if attempts of drug therapy, generally with bisphosphonates, have been made. Nevertheless, the surgical treatment remains essential, involving modelling resections especially in the growth period, the disadvantage being that they permit recessions (up to 20%). In cases of progression towards the skull base, when the risk of compression on the ocular foramen may appear, more extended ablations are recommended [10].

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