Fibrous dysplasia mimicking cemento-ossifying fibroma histopathologically - A case analysis.

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ABSTRACT
Cemento-ossifying fibroma is a fibro-osseous lesion which belongs to the same category as fibrous dysplasia and cement-ossifying dysplasia. In a recent WHO classification (2005), the term ‘cemento-ossifying fibroma’ was replaced with ‘ossifying fibroma’. These are slow growing, painless lesions which are seen more commonly in women between the third and fourth decades of life. We report an unusual case of a cemento ossifying fibroma of the left mandible diagnosed histopathologically in a 52 year old female patient who presented with a painless swelling on the left side of the face. The lesion was treated with surgical resection and reconstruction.

KEYWORDS: Fibrous dysplasia, Cemento-ossifying fibroma, Jaw, Fibro-osseous lesion, Maxillofacial imaging.

INTRODUCTION
Fibro-osseous lesions are a heterogeneous group of benign lesions of unknown etiology affecting the jaws and other craniofacial bones, which are characterized by the replacement of normal bone by fibrous tissue, which contains a newly formed, mineralized product. Over the last several decades, fibro-osseous lesions have evolved which includes two major entities: fibrous dysplasia and ossifying fibroma, as well as the other less common lesions such as florid osseous dysplasia, periapical dysplasia and focal sclerosing-osteomyelitis. It may be divided into three categories; monostotic (74%), polyostotic (13%) and craniofacial (13%). The last category, identified by Davis and Yardley, appears to be confined to the face and jaws involving two or more bones. Cement-ossifying fibroma is considered by most as relatively rare, benign, nonodontogenic neoplasm of the jaw bone with female predilection.
over men (5:1), it is most frequently seen in third and fourth decades of life. The most common location is in premolar-molar region of the mandible, with 62% to 89% of patients, 77% occurring in the premolar and 75 to 89% in molar region and in maxilla, 11-26% of cases.[3] Clinically, these tumours manifest as a slow-growing intra-bony asymptomatic mass, over a period of time lesion may become large enough to cause facial asymmetry. Radiologically, cemento-ossifying fibroma shows a number of patterns depending on the degree of maturation of the lesion. Histologically, these tumours are composed of well vascularized fibro cellular tissue with the capacity to form immature bone trabeculae and cementoid formations, though these findings are not specific of the lesion and can also be seen in fibrous dysplasia. FD is an hamartomatous condition related to mutations in the gene (GNAS I) encoding the α-subunit of stimulatory G-protein (Gs-α) that result in the increased production of cyclic adenosine monophosphate (cAMP), affecting the proliferation and differentiation of preosteoblasts. This paper reports a case of FD affecting mandible that was initially characterized by osseous expansion on the left side of the mandible and slight facial asymmetry. A definitive diagnosis therefore requires correlation of the clinical, radiological and histological findings.[4]

CASE REPORT
A 52 year old female patient reported to dental OPD with chief complaint of swelling in lower left posterior region since last 4 years. Patient was apparently all right 4 years back, and then since she observed a swelling which was spontaneous and gradual in onset on left lower back teeth region. No history of pain was present with the swelling. The medical and family history was not significant. Dental history included history of extraction in left posterior 36 region since 1 year back. She gave history of gradual increased in size of swelling. No history of difficulty in speech or chewing. No history of paresthesia in the region. No history of swelling on any other body part. No history of hearing disturbances. No history of any pigmentation anywhere on skin. Pt was hypertensive since last 6 months but not under any medication. History of hystrectomy present 30 years back. No history of allergies to any known drug. Extra oral examination [Fig 1] showed mild thickening of mandible on left side, antegonial notch appears to be obliterated. Lymphnodes were not palpable. Intraoral examination [Fig 2] showed single diffuse bulging swelling seen in left mandibular alveolus., extending anterioposteriorly from 36 region to retromolar region of size 3x2 cm. Cortical expansion seen on both buccal and lingual cortex. Vertical height of mandible appears to be increased in 36, 37 and retromolar region. Overlying and surrounding
skin appeared normal. Antagonist teeth were absent. No swelling on contra lateral side. On palpation swelling was firm to bony hard in consistency, non-tender, non-compressible and does not bleed on manipulation. On hard tissue examination 34,35,37,38 were non-tender and they gave vital response on vitality testing. Based on history and clinical examination patient was provisionally diagnosed as benign fibro osseous lesion highly suspected of mono ostotic fibrous dysplasia of mandible. Differential diagnosis of- Ossifying fibroma, exostosis, Osteogenic sarcoma, Paget’s disease were given. Routine blood investigation-CBC, HBsAg, HIV screening and Specialized- Serum Alkaline phosphatase levels were performed and were within normal range.

Radiographically Panoramic radiograph [Fig 5] shows as single uniformly dense radiopaque lesion on left side of mandible involving body and ascending ramus, body appears enlarged periphery of lesion is illdefined and irregular and gradually blending with the normal bone. Pattern of radio opacity was suggestive of ground glass appearance which was confirmed on periapical and occlusal radiographs [Fig 3 & 4].

Computed tomography scan [Fig 6] was done and reports in bone window that there is evidence of expansile bone thickening (hyperdense area in bone window) with ground glass matrix involving the body, angle, ramus, coronoid process and alveolar process of mandible on left side. The rest of the mandible appears normal. The CT findings are suggestive of fibrous dysplasia involving the body, angle, ramus, coronoid process and alveolar process of mandible on left side. Panoramic view in CT scan showing diffuse radiopacities in left side of posterior mandibular region [Fig 7] Incisional biopsy was done in the patient which reported the clinicopathplogic correlation suggestive of fibrous dysplasia. Patient was referred to department of surgery and advised cosmetic recontouring of bone and surgical specimen was sent for histopathology which reported that feature suggestive of central ossifying fibroma. [Fig 8].

**FIGURE LEGEND**

**Figure 2:** Intra oral view showing swelling left side lower posterior jaw

**Figure 3:** Intra oral periapical radiograph shows finger print /ground glass appearance.

**Figure 4:** Occlusal radiograph of left mandibular lesion showing expansion of medial cortical plate with displaced posterior tooth.

**Figure 5:** Portion of Panoramic radiograph of left side of posterior jaw showing increase in jaw size and density, lifting of molars with displaced the inferior alveolar nerve canal.
**Figure 6:** Axial CT scan showing expansile bone thickening (hyper dense area) with ground glass matrix involving left side of the body of the mandible.

**Figure 7:** Panoramic view in CT scan showing diffuse radio-opacities in left side of posterior mandibular region.

**Figure 8:** Basophilic globules of calcified mass along with osteoid tissue, round to ovoid basophilic cementum-like calcifications.

**FIGURES**
DISCUSSION

Fibrous dysplasia (FD) and cemento-ossifying fibroma (COF) are benign fibro-osseous lesions that are generally considered to be separate entities, distinguished by histologic and radiographic features. In our experience, some lesions lack the classic clinical, radiographic, or pathologic features of FD or COF and rather have overlapping features of both entities. FD is one of several fibro-osseous lesions that affect the maxillofacial region. It does not occur exclusively in the jaws, such as cherubism or focal cemento-osseous dysplasia, but it can occur anywhere in the skull. Most clinicians are trained to associate a ground glass radiographic pattern with FD when reading a panoramic radiograph. This is inaccurate. When confronted with a lesion of variable radiographic appearance, a diagnosis of FD should be considered in the differential. When a lesion is observed in a panoramic radiograph, it is important for the clinician to be aware of the radiographic patterns possible in FD and not exclude FD in the absence of typical ground glass radiographic pattern.\(^8,9\)

Several textbooks indicate that the appearance of maxillofacial FD in panoramic radiographs may vary. In this case, images were sufficient for diagnosis of this condition. The radiographic presentation was unusual in the left mandible. The hypoplastic left mandible was most unusual. A biopsy performed in this site confirmed FD sites of the FD lesions presented in this case were radiopaque and presented with a characteristic ground glass appearance. CT was very helpful in the diagnosis. It provided clear visualization of the classical ground glass pattern. FD is a condition that results from a postzygotic mutation in the GNAS 1. The clinical severity of the condition presumably depends on the point in time during fetal or postnatal life that the mutation of GNAS 1 occurs.\(^5\) The mutation occurred during later period in this case where the progeny of the mutated cell dispersed and

In fibrous dysplasia, an intimate continuity between the lesion and normal bone is generally found. The tumor expands throughout its length and the margins are diffuse and radiographically poorly defined.[6] Its radiological structure is more homogenous than that of ossifying fibroma or cemento-ossifying fibroma, both of which are filled with radiopaque foci.[5,6,7]

Cemento-ossifying fibroma is a well-circumscribed tumor that grows expansively and has clearly defined margins. Lesions are oval, spherical, or multilocular and are clearly separated from the surrounding bone by osteolytic borders the cemento-ossifying fibroma appears as a radiolucent lesion with no evidence of internal radiopacities. As the tumor matures, there is increasing calcification so that the radiolucent area becomes flecked with opacities until ultimately the lesion appears as an extremely radiopaque mass. Displacement of adjacent teeth is common.[2] One additional important diagnostic feature is that there is a centrifugal growth pattern rather than a linear one and therefore the lesions grow by expansion equally in all directions and present as a round tumor mass. Other lesions (fibrous dysplasia) expand the cortex linearly and the outline of the expanded mandible is not in continuity with the remainder of the outline of the lesion.[11,12] Three radiographic patterns have been described depending on radiographic borders:

1. Defined lesion without sclerotic border (40%)
2. Defined lesion with sclerotic border (45%)
3. Defined lesion with illdefined border (15%)

In early stages of COF, appears as radiolucent lesion with no internal radiopacity. With maturity of lesion there is increasing calcific flecks progressing ultimately an radiopaque mass. The growth pattern of mass is centrifugal so grows equally in all direction. Differential diagnosis depends on radiographic feature of the lesion. It includes Fibrous dysplasia, Cemento-osseous dysplasia, Condensing osteitis, Pindborg’s tumor, Retained root, Odontoma. The boundaries of COF are usually weldefined surrounded by soft tissue capsule whereas fibrous dysplasia is usually diffuse and blends with surrounding with bone. FD has a characteristic groundglass appearance whereas not seen in COF. Also FD resorbs the teeth and
the expanded bone still resembles the normal morphology. Cemento-osseous dysplasia is usually multifocal whereas COF is not. Presence of simple bone cyst and a wide sclerotic border is a characteristic of cemental dysplasia. Vitality test can help in differentiating COF from condensing osteitis. Pindborg’s tumor have a high association with impacted teeth. Odontoma can be differentiated by presence of tooth like structures. Retained root have a root canal in root fragment. Multiple COF are rare and there pathogenesis is not yet understood. There are few cases of multiple COF. Familial COF has also been reported.[13, 14,15]

Histologically a fibro-osseous pattern is typically seen, yet akin to the imaging features, subtle changes are seen at various stages of the disease’s natural history. In the early formative phase, pronounced osteogenesis is seen with thin osteoid anastomosing trabeculae that are rimmed with osteoblasts. The stromal fibroblastic element is proliferative and hypercellular although no pleomorphism can be seen. With ensuing weeks, the trabeculae thicken, yet the osseous collagen pattern remains woven and the trabeculae assume the classic “Chinese figure” characteristics. The fibrous element continues to be hypercellular. In later stages of the disease woven bone is replaced by lamellar bone trabeculae; extensive remodeling may result in a mosaic pattern of resting and reversal line.

Yih et al suggested the possibility of a hereditary element. Treatment is surgical in the form of complete resection. since it is low vascularized it is easy to remove from the surrounding bone. Prognosis is good and recurrence is rare.[15]

CONCLUSION
The definitive diagnosis can rarely be rendered on the basis of histopathologic features alone; rather, procurement of a final diagnosis is usually dependent upon assessment of microscopic, clinical and imaging features together.

REFERENCES