Cementifying fibroma—A case report

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ABSTRACT

The etiology and pathogenesis of fibro-osseous lesions remain a subject of investigation. Various explanations are offered- congenital anomaly of bone, developmental defect caused by faulty embryogenesis ; hamartoma, tumor of periodontal membrane origin, mesenchymal tumor arising in spongiosa and an abnormal repair of bone after injury. Fibro-osseous lesions of the jaws form a group of conditions, which are remarkable for their clinicopathological similarities. On occasions clinician may find himself in the position of being the arbiter in the face of equivocal histological evidence. Some pathologists use the same terminology for apparently quite dissimilar lesions, and seemingly others to render the same diagnosis use variable histologic criteria. By analyzing the clinical, radiographic, gross/surgical and histological features of all lesions coded as fibro-osseous lesions we should be able to separate a clinicopathologic entity. A case of cementifying fibroma is presented here along with discussion.

KEY WORDS: Fibro-osseous lesion, *jaw bone, Histopathology*.

Case Report

A 30 year old patient who was about to commit suicide because of her appearance was rescued and brought to the hospital for further treatment of her maxilla. She has a history of rapidly growing lesion on left side of maxilla Since 3 years. Initially it was asymptomatic, Since 7 months she has pain, dysphagia and difficulty in respiration due to the size of the lesion(Fig.1). The lesion was involving complete left side of the maxilla and extending to the right side of the maxilla. The size of the lesion was 22 x 15 cms and the shape was irregular. The maxillary anterior teeth were protruded from the face 5 cms away from the nose. Occlusion was deranged. Radiograph was showing mixed radiolucent and Radio opaque lesion (Fig.2.) . Case was discussed inter departmentally between ENT Surgery, Plastic Surgery, Oral Surgery and Oral Medicine and Radiology and the diagnosis was pointing towards fibro osseous lesion, giant cell lesion and Sarcomatous lesion. The Rapidity of the growth of the lesion frightened the surgeon and the provisional diagnosis was towards the sarcoma (Osteo Sarcoma). Incisional biopsy was ruled out and total excision of the lesion was planned.

Surgical excision of the entire maxilla was done. After the surgical removal the entire maxilla was weighing 3.2 kgs. The histopathological report of the lesion was towards fibro osseous lesion (CEMENTYFYING FIBROMA). Which is a benign neoplasm. If that was the provisional diagnosis some kind of surgical recontouring would have been done to save the patients maxilla.

Discussion

Fibro-osseous lesions have been a subject of discussion as controversies prevail regarding their etiology. Histopathology is also confusing as similar microscopic picture may be seen in different lesions. Diagnosis of fibro-osseous lesions of the skeleton particularly the bones of the jaws present a considerable problem to clinicians and pathologists. Giant cell reparative granuloma must be considered in the differential diagnosis of fibro-osseous lesion because of the presence of giant cells in both the lesions. Chronic scalloping osteomyelitis, osteogenic sarcoma, chondrosarcoma, Ewing's sarcoma, must be differentiated from mottled type of fibro-osseous lesion. Other conditions which may resemble fibro-osseous lesion include osteoblastoma, osseous dysplasia, odontogenic myxoma, osteogenesis imperfecta. Paget's disease.[1,2]

Fibrous dysplasias are basically non-pathologic entity with unknown etiology. However looking at their age of appearance, clinical behavior, coupled with microscopic picture, it would not be out of place

to claim that these lesions are the outcome of disturbances in normal growth pattern[1].

Review

The term fibro-osseous lesion is a generic designation of a group of jaw disorders that microscopically exhibit a connective tissue matrix and islands/trabeculae of bone. Fibro-osseous lesions have been a subject of controversy and a matter of discussion amongst both pathologists and clinicians. Waldron in 1985 came out with a better-detailed grouping of fibro-osseous lesions based on their pathogenesis, histopathology and clinical features [1].

Classification of fibro-osseous lesions[1,2]:

1. Fibrous dysplasia
   a) polyostotic
   b) monostotic
2. Fibro-osseous or cemental lesions presumably arising in the periodontal ligament.
   a) periapical cemental dysplasia
   b) localized fibro-osseous cemental lesions
   c) florid cento-osseous dysplasia (gigantiform cementoma)
   d) Ossifying and cementifying fibroma.
3. Fibro-osseous neoplasms of uncertain or debatable relationship to those arising in the periodontal ligament.
   a) Cementoblastoma, osteoblastoma and osteoid osteoma.
   b) Juvenile active ossifying and the so called active ossifying cementifying fibroma.

I.Ossifying fibroma

Menzel[3] in 1872 first described the entity known as ossifying fibroma. Ossifying fibroma is designated as a benign form of fibro-osseous lesion with a well circumscribed, slow growing and sharply defined margins with a radiolucent peripheral component.

Etiology [2,3]: The etiology of ossifying fibroma remains unknown. It has been suggested however that those ossifying fibromas associated with trauma seem to behave more aggressively than the typical benign ones. Ossifying fibroma should be considered as a tumor of the periodontal membrane origin. They arise from the multipotent mesenchymal blast cells present in the periodontal membrane and have a capacity to produce cementum, alveolar bone and fibrous tissue. Some ossifying fibromas behave in an aggressive fashion, reaching massive proportion with extensive cortical expansion. Most of these aggressive lesions occur in children and have prompted the term 'Juvenile aggressive or active ossifying fibroma'[4]. No definitive histopathologic findings have been uncovered in determining the potential for aggressiveness. The lesions are most commonly seen in the third and fourth decades of life with a predilection for females, ratio of Female: Male is 5: 1. Mandible is most commonly involved, seen 89% of all cases. Sites of involvement are molar region 52%, premolar region 25%, incisor region 13%, cuspids 11%. Patients with clinically progressive lesions may approach for treatment due to pain, paresthesia and facial asymmetry. The lesions are usually firm, non-tender on palpation, well encapsulated and may reach size up to 10 X 12 cm. If seen in maxilla, they may cause cortical expansion obliterating the buccal sulcus extension into the nasal cavity and orbital floor may lead to epistaxis and even epiphora. Mandibular lesions are usually seen posterior to the canines. Expansions of both cortical plates may also seen. Extension of tumor mass into the ramus of the mandible, and involvement of the inferior border may cause paresthesia of the inferior alveolar nerve. Bilateral lesion involving the maxilla and the mandible has also been noted.

Radiographic features:

Two major types

a) Expansive multilocular configuration.
b) Expansive unilocular radiolucency with or without opacification.

All six variations

1) radiolucent superimposed or residing in edentulous region – 28%
2) radiolucent superimposed or residing in edentulous region with opacification 42%
3) radiolucent interradicular with root divergence – 5%
4) Radiolucent interradicular with opacification – 9% with root divergence.
5) Multilocular 7%
6) Aggressive with opacification more than 5 – 6 cms in size – 9%

Fibrous dysplasia is poorly marginated radiographically whereas ossifying fibroma has a well-defined lesional border.

Histological features: Microscopically presents as evenly spaced speckles of bone rimmed with osteoblasts and osteoclasts within fibrous stroma. Most of the spicules are centrally composed of woven bone with lamellar bone formation at the periphery. Cementum like numerous ovoid and heavily calcified masses are seen in stroma.

Ossifying fibromas are also fibro-osseous lesions seen in growing children, usually involving single bone and having a slow painless growth pattern. These are benign tumors showing variable clinical behavior and must be differentiated from monostotic fibrous dysplasia. Rapidly growing lesions attaining sizeable dimension have been reported under the juvenile variety of ossifying fibroma.

Lesions are well circumscribed and well delineated and hence can be differentiated form fibrous dysplasia. Mandible shows increased predilection as opposed to maxillas seen in fibrous dysplasia. Disturbances in occlusions not uncommon, though paresthesia of the inferior alveolar nerve is rarely reported under the microscope. The cementifying fibroma may occur at any stage, but is more common in older subjects, either jaw may be involved, but it usually occurs in the mandible. Histological pattern varies with the stage of development, being predominantly cementum like as the rounded masses enlarge and fuse together. Bhaskar[3] has warned that cementifying fibromas are in reality, ossifying fibromas, in which the bone tissue appears basophilic and resemble cementum superficially. Cementifying fibromas and ossifying fibromas are two distinct benign neoplasms representing two facets of the same tumor (Shafer etal)[2]. The presence of cementicles and osteoid structures in ossifying cementifying fibromas has raised a big controversy regarding their origin

II. Cementifying fibroma

Cementifying fibroma, a benign fibro-osseous lesion of the jaws consists of cellular fibroblastic tissue containing rounded or lobulated masses of calcified cementum like tissue. It is usually well circumscribed, generally grown slowly and occurs more frequently in females. A review of literature [3,4,5] disclosed a small number of cases that showed aggressive behavior and recurrences, the recurrences were related to the inherent biologic behavior of the tumor rather than to insufficient surgical removal. Although WHO and Shafer [2] regard the cementifying fibroma as an odontogenic tumor and consider ossifying fibroma separately as non-odontogenic neoplasms, this seems arbitrary and unnecessary separation, as the clinical radio logic and prognostic features of the lesions are identical.

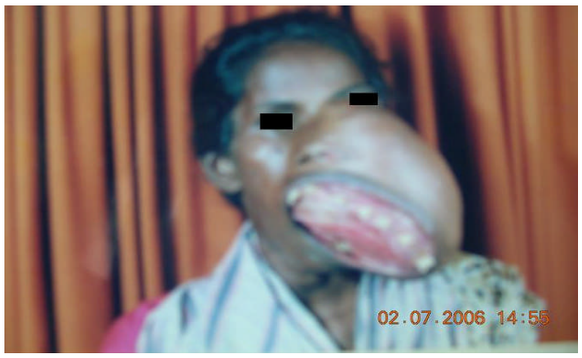

Fig 1. **A massive fibrous osseous lesion**

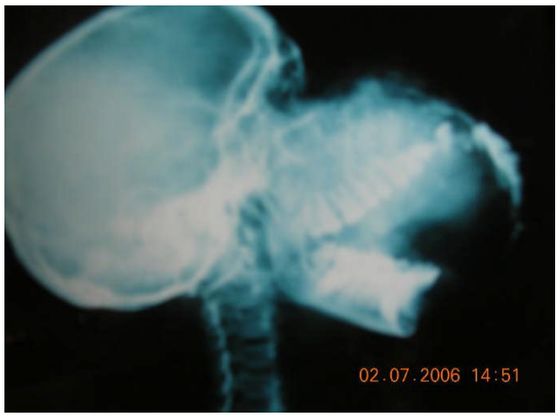

Fig2. Radiograph showing mixed radiolucent and radioopaque lesions.

Histologically the calcified product in some cases consists of almost entirely of amorphous, basophilic, usually rounded calcifications commonly considered to be cementum. These lesions often designated as cementifying fibromas. A significant number however contain an admixture of the two types of calcifications and are often grouped as “cemento-ossifying fibromas”. Cementifying and ossifying

fibroma may be seen with a similar mottled appearance to that seen in fibrous dysplasia. The following differences are recognized radiologically.

1. **Shape**: the cementifying and ossifying fibromas are predominantly round while those of fibrous dysplasia are more rectangular.
2. **Jaw expansion**: jaw expansion caused by cementifying and ossifying fibroma is usually nodular or dome shaped whereas the jaw expansion of fibrous dysplasia is usually of the elongated fusiform type.
3. **Margins**: cementifying and ossifying fibroma has sharply defined radiographic margins. In contradistinction, the margins of fibrous dysplasia are indistinct, blending imperceptible with normal bone.
4. **Predominant jaw**: approximately 70% of cementifying and ossifying fibromas occur in the mandible. Fibrous dysplasia shows a slight predilection for the maxilla.
5. **Age** : The age range for ossifying fibromas is from 7 – 58 years. The majority of active case of fibrous dysplasia is found in patients under 20 years of age.

Monostotic fibrous dysplasias are more commonly encountered by maxillofacial surgeon as they are predominantly seen in craniofacial areas. These conditions remain asymptomatic with painless slow bony expansion ultimately altering the facial morphology and thereby drawing both the patients' as well as the clinicians' attention. Disability and dysfunction is not seen in the jaws and occlusion is rarely disturbed though obliteration of the palatal vault and maxillary sinus is not uncommon. Involvement of frontal bone may result in dystopia, diplopia, disfigurement and disturbances in vision. A sudden spurt in an indolent lesion is not uncommon.

.**Summary and conclusions**

Fibro-osseous lesions appear in the jaw bones with a variety of clinical and radiographic characteristics. The fibro-osseous nature of the lesion can only be confirmed by biopsy. As it has varied radiographic appearance, a fibro-osseous lesion must be considered in the differential diagnosis of almost all radiolucent, radiopaque or radiopaque-radiolucent lesions of the jaw bones. Conversely many serious bone diseases and lesions are manifested radiographically with characteristic "fibro-osseous" bone changes. Even clinically asymptomatic lesions that show radiographic changes should have a biopsy. But differentiation between the various fibro-osseous lesions i.e., fibrous dysplasia, ossifying fibroma and cementifying fibroma is difficult on the biopsy results alone. A review of the clinical and radiographic features together with the histopathologic changes and surgical findings will allow for a definitive diagnosis. The variability of fibro-osseous lesions are so great that can present markedly separate clinical and radiologic entities, depending on the proportion of fibrous to osseous tissue within the lesion. Historical, clinical, radiographic and histopathologic data combined with surgical findings are though essential, but still causing a lot of confusion in diagnosis and treatment pattern. Probably it requires further studies to determine the exact nature of these lesions,to pin point the characteristics of its clinical features, radiological and histological features for a definitive diagnosis and treatment planning.

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