Aggressive Cemento-ossifying Fibroma of Maxilla: A Case Report

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Abstract

Fibro-osseous lesions of bone have evolved over several decades to integrate two major entities: Fibrous dysplasia and Ossifying fibroma along with the other entities such as periapical dysplasia, hyperparathyroidism and Paget's disease with hypercementosis in the late stages. The overlapping clinical, radiologic and histomorphologic features of these subtypes have led to diagnostic dilemma for the clinician, radiologist and the histopathologist. The biological characteristics of fibro-osseous lesions range from indolent to aggressive and from inflammatory to neoplastic. Cemento-ossifying fibroma is a mesodermal, slow-growing, benign fibro-osseous lesion of the jaws composed of lamellar bone, fibrous tissue and cementum. The tumor may grow quite extensively; thus, the term "aggressive" is sometimes applied. The aggressive local behaviour and high recurrence rate (30-58%) means that it is important to make an early diagnosis, apply the appropriate treatment and, especially, follow-up the patient over the long-term. We hereby report a case of aggressive cemento-ossifying fibroma in the maxilla.

Keywords: *Aggressive, cement ossifying fibroma, fibro-osseous lesion, maxilla*

Introduction

Fibro-osseous (FO) lesions are characterized by replacement of normal bone architecture by collagen fibers and fibroblasts that contain varying amounts of mineralized substances, which may be bony or cementum like in appearance. Many lesions contain an admixture of this calcification. FO lesions comprise fibrous dysplasia, periapical cemento-osseous dysplasia, focal cemento-osseous dysplasia, florid cement-osseous dysplasia and cemento-ossifying fibroma (COF). Ossifying fibroma (OF) is a fibro osseous tumour affecting both the jaws and composed of proliferating fibroblast and osseous products that include bone and cementum like material. The tumour is slow growing and well demarcated from the adjacent bone. Due to the presence of both cementum like products and bone these lesions are designated as ossifying fibroma, cementifying fibroma, cemento-ossifying fibroma.[1] According to the World Health Organization (WHO) classification in 1992, COF is classified within non odontogenic lineage of tumors.[2,3] COF occurs mainly in the second to fourth decades of life with a predilection for females. Occasionally, it may grow to a massive size and cause serious cosmetic and functional problems, and rarely, reports of lesions behaving in an aggressive fashion, diagnosed as "aggressive", "juvenile" or "active" COF, appear in the literature.[4] COF is a locally destructive, deforming tumor that can occur almost anywhere in the craniofacial region. Most osseous lesions have been reported in the premolar-molar region of the mandible, other sites include the maxilla, zygoma, paranasal sinuses, orbital and retromastoid regions.[5]

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Case report

A 30 year old female reported to the department of Oral Medicine and Radiology with a complaint of painless swelling in left side of face since two months. Patient noticed intraoral swelling initially which further progressed extraorally. There was no history of any trauma, pain on mastication or difficulty in swallowing.

Patient had no addictions, her medical and dental history was not significant.

Clinical examination revealed an extraoral diffuse swelling which was seen in the left middle third of face extending horizontally from lateral wall of nose to 1cm in front of the left tragus of ear, and vertically 1cm below inferior orbital rim involving left antrum region to the line joining the left angle of mouth and tragus of ear. Size was approximately 5x3 cm. Overlying skin was of normal colour and intact with smooth surface. On palpation it was hard and nontender. [Figure1]

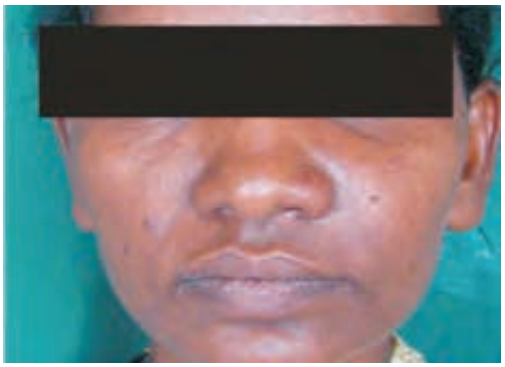

Fig1: Diffuse extraoral swelling seen on the left side in the middle third region

Intraorally obliteration of the buccal vestibule was seen in the upper left region of jaw extending from maxillary left central incisor to the third molar. A well defined swelling was seen in the left side of palate extending from midline to the marginal gingiva of all the teeth on left side, approximatly 5x3 cm in size. Overlying mucosa was normal and intact. On palpation swelling was hard in consistency and non tender. Expansion of the buccal cortical plate was present from canine to the second premolar region.[Figure 2] Grade I mobility was associated with first, second premolars and first molar. Bilaterally submandibular lymphnodes were palpable of approximately 1x1 cm in size and were firm, mobile and non tender. Thus a provisional diagnosis of benign tumor like adenamatoid odontogenic tumor with a differential diagnosis of cemento-ossifying fibroma and fibrous dysplasia was made.

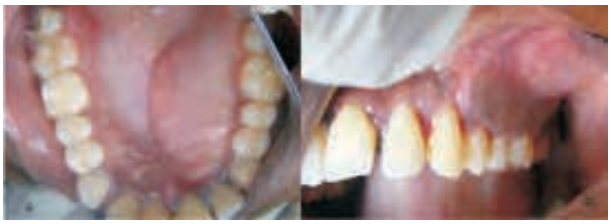

Fig 2: swelling seen on the palate (a) and expansion of buccal cortical plate (b)

Radiographic examination revealed opacification of left maxillary antrum with indistinct borders (OPG, PNS). Intraoral radiographs (IOPA and maxillary occlusal view) showed mixed radiolucent and radiopaque lesion extending from maxillary left central incisor to the third molar region. Radiopaque component was having ground glass appearance. Loss of lamina dura was seen with the teeth from maxillary left central incisor to first molar. Slight divergence of the roots was seen with maxillary left second premolar and first molar.[Figure 3, 4]

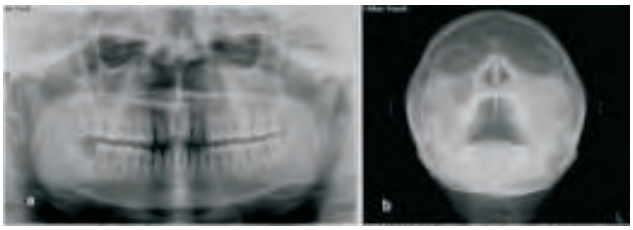

Fig 3: OPG (a) and PNS (b) shows opacification of the left maxillary sinus

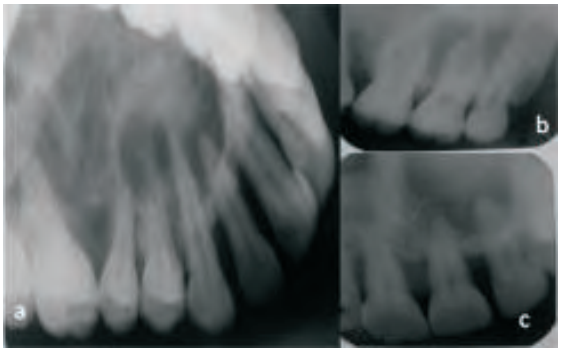

Fig 4: Maxillary occlusal view (a) showing mixed radiolucent and radiopaque lesion IOPA (b and c) showing ground glass appearance

Incisional biopsy was done. It showed highly cellular stroma with plump fibroblasts and myxomatous areas. The connective tissue showed highly vascular stroma composed of dilated engorged blood vessels and capillaries. Areas of osteoid tissue and trabeculae of new bone formation were noticed. Calcified bodies resembling cementum were noticed within stroma. Thus the diagnosis of aggressive cemento-ossifying fibroma was confirmed.

Discussion

Ossifying fibroma was first described by Menzel in 1872. Montgomery in1927 coined the term "Ossifying Fibroma".[3] The term ossifying fibroma is used if the

predominant component is bone which include ossicles, osteoids, fiber bone (woven bone) and mature bone[1,6] while cementifying fibroma is used if the predominant component is cementum like spherical calcifications. However, the microscopic appearance of an ossifying fibroma and a cementifying fibroma can be very similar, and the two are now thought to represent a spectrum of one disease and are combined under the name cemento-ossifying fibroma (COF).

COF is a disorder of unknown etiology. Marx and Stern have stated that ossifying fibroma occurs frequently in jaws, probably because these lesions are related to extensive mesenchymal cellular induction into bone and cementum required in odontogenesis.[1] Bernier hypothesized that COF in the bone might be caused by an irritant stimulus (such as tooth extraction) which may activate the production of new tissue from the remaining periodontal membranes.[3] Brademann et al. explained that ectopic periodontal membrane differentiating from primitive mesenchymal cells in the petrous bone may serve as a cause of development of COF in this area, and trauma such as severe whiplash may be a factor in the induction of proliferation of COF. The ethmoidal location of COF may also be explained by incomplete migration of mesenchyme and its differentiation into periodontal membrane.[3]

According to WHO classification (2005) O F most commonly occurs in the 3rd to 4th decades and shows a predilection for females [3] with female:male ratio of 2:1. The premolar–molar region of mandible is more commonly involved than maxilla although cases have been reported in the other craniofacial bones. 70% of cases of COF involve the mandible, but a significant percentage (22%) has been found in the molar region of the maxilla, ethmoidal and orbital regions and exceptionally in petrous bone.[2,3] Our case is that of a 30 year old female with the lesion seen in the maxilla.

The clinical features of COF can vary from indolent to aggressive behavior. The characteristics are more like those of a tumor than a bone dysplasia. Ossifying fibromas are initially asymptomatic until noticeable swelling or facial disfigurement is produced. The lesions are restricted to the tooth bearing areas of the jaw although in posterior region it can be extended into the ascending ramus. The lesion appears as hard localized and slow growing painless mass that may displace the adjacent structures.

According to Langlais, Aggressive ossifying fibroma (AOF) has some signs that are s/o AOF

- Occurrence of lesion in child or adolescent.
- H/o rapid increase in swelling reported by the patient.
- Pain reported by the patient or occuring on palpation.
- Occurrence of lesion in a maxillary location.
- Large size of the lesion radiologically (> 5cm in diameter)
- Expanded cortex visibly perforated on CT but not the burnt out appearance commonly seen in plain radiograph
- Recurrence of the lesion, many cases reported as aggressive actually did not recur.[9]

Juvenile Ossifying Fibroma (JOF) is a very aggressive form of COF that occurs in the first 2 decades of life. Johnson *et al* have reported JOF occur at any age between 3 months and 72 years. Among the many classification systems for this lesion, the classification by Slootweg *et al* is noteworthy and have classified JOF into two distinct groups, the JOF-WHO type and JOF-PO (psammoma-like ossicles) type, based primarily on the difference in the age of occurrence: the mean age of occurrence of JOF-WHO is 11.8 years and that of JOF-PO is 22.6 years.[8]

Radiologically, COF appears as a well-circumscribed, solitary radiolucency with scattered radiopaque foci. It shows distinct stages during its development. Radiographically, it is characterized by three stages: initial or early, mixed and mature stage.[3] In the early stages, the COF 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.[2,3] An important diagnostic feature of COF 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 and expands the surrounding cortical bone without perforation. A significant point is that the outer cortical plate, although displaced and thinned, remains intact. Large mandibular lesions may cause a characteristic thinning and downward “bowing” of inferior border.[3] Aggressive lesions in maxilla tend to have ground glass appearance.[9]

In our case, rapid growth was present in the maxilla with expansion of buccal cortical plate, the size of the lesion was 5x3cm. Radiographically also it showed ground glass

appearance. Hence considering the above mentioned criteria by Langlais it was diagnosed as an aggressive cemento-ossifying fibroma.

Microscopically, COF reveals many delicate interlacing collagen fibers, seldom arranged in discrete bundles, interspersed with large numbers of active, proliferating fibroblasts and cementoblasts. Although mitotic figures may be present in small numbers, there is seldom any remarkable cellular pleomorphism. As the lesion matures, the islands of cementum increase in number, enlarge, and ultimately coalesce.[3,10]

Differential diagnosis of cemento-ossifying fibroma includes fibrous dysplasia, florid osseous dysplasia, cementoblastoma. Aggressive lesions with marked destruction of adjacent structures may radiographically mimic osteogenic sarcoma.[1]

The treatment of choice for ossifying fibroma is surgical excision. Small well demarcated lesions can be excised by enucleation and curettage whereas larger lesion show a more aggressive pattern requires radical surgery with healthy pattern. In any case, decision to enucleate or resect radically depends on involvement of inferior border of mandible and spread of lesion in soft tissue or in maxillary sinus and nasal cavity.[1] Radiotherapy is generally contraindicated because of the risk of malignant transformation and the potentially harmful late effects in children.[5] The recurrence rate by either of the above treatment is low with a good prognosis, with the recurrence rate being 20-25% for ossifying fibroma.[1] The aggressive nature of the lesions with high rate of recurrence (30-58%) suggest that COF should be treated as the locally aggressive neoplasm.[3,8,11]

Clinical, radiographic and histopathologic features of COF and other fibro-osseous lesions are overlapping and may cause confusion in classification, diagnosis and treatment. The rapid growth rate often exhibited by these lesions can be quite alarming and cause the clinician to suspect the presence of a malignancy. Thus careful evaluation of the lesion is important.

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