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"A Rare Case of Central Ossifying Fibroma: Unmasked in the Maxilla"

Abstract:
Central ossifying fibroma (COF) is a rare benign neoplasm of the jaws characterised by the proliferation of fibroblasts and the formation of mineralised tissue. The purpose of this case report is to describe the diagnosis and management of a central ossifying fibroma of maxilla in a 17-year-old female. This report aims to increase awareness of COF among dental and medical professionals, also to highlight its clinical presentation, differential diagnosis, and treatment options.

Key-words: COC, Fibro-osseous lesions, Cemento-ossifying fibroma

Introduction:

Fibro-Osseous lesions are a group of poorly defined conditions affecting the maxilla and mandible along with the craniofacial bones. A prominent feature of these lesions is the replacement of bone by fibrous connective tissue, which has foci of mineralization as well as varying degrees of dispersed calcification in the form of immature bone and cementum type tissue depending on the type of condition involved [1]

Central Ossifying Fibroma (COF), is a pathological condition that affects the facial bones and craniofacial bones. COF is a relatively rare, benign tumor that primarily arises from the periodontal ligament or the periosteum. The periodontal ligament is believed to contain multi potent cells that forms the cementum, bone and fibrous connective tissue. It is characterised by the formation of fibrous tissue and the deposition of mineralised material within the lesion. [2,4]

COF most commonly occurs in the second to fourth decade of life. It has been observed that COF affects females predominantly. It can occur in any part of the jaw, but it more frequently involves the mandible and rarely seen in maxilla. It is relevant to mention here that it is seen frequently involving the premolar-molar region. [5,6]

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While the exact etiology of COF is uncertain, it is believed to be associated with local trauma, inflammation, or hormonal imbalances. Furthermore, genetic factors and certain systemic diseases have been suggested to contribute to the development of COF. [7]

Clinically, COF presents as a slow-growing, painless swelling in the affected region, often accompanied by expansion of the jawbone or displacement of adjacent teeth. Radiographically, COF exhibits well-defined radiolucent or mixed radiolucent-radiopaque features, depending on the stage of mineralization within the lesion. [8]

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However, it is crucial to differentiate COF from other entities such as cemento-ossifying fibroma, fibrous dysplasia, and odontogenic tumors through clinical, radiographic, and histopathological examinations.

Histologically, COF is characterised by the presence of a cellular fibrous connective tissue stroma, in which variable amounts of calcified tissue can be observed. The mineralised material may appear as spherical structures, resembling cementum or bone, and can be seen in association with fibroblastic cells, osteoblasts, and multi nucleated giant cells. [9]

The management of COF typically involves surgical excision of the lesion, followed by careful reconstruction of the affected area to restore optimal function and aesthetics. Recurrence rates for COF have been reported to be relatively low, although long-term follow-up is recommended due to the possibility of regrowth.

Case Presentation:

A 17-year-old female patient presented with a gradually increasing painless swelling in the left maxillary region since 2-3 years. The swelling extended from left maxilla involving the region wrt 21 to 28 up to floor of the orbit to alveolar process of maxilla, Superior-inferiorly involving left lateral wall of nose (Fig 1, Fig 2).

Extra-orally, the swelling extended from the commissure of lip to the inferior margin of eye.

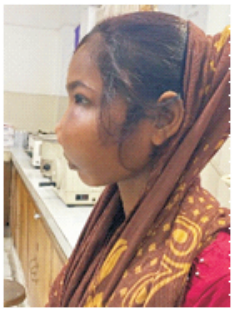

Fig 1 Photograph showing extra oral swelling in the patient (frontal view)

Fig 2 Photograph showing extra oral swelling in the patient (lateral view)

Intra-orally, vestibular swelling extending from 21 to 28 was seen. The swelling was 56.13mm x 61.64 mm in size, firm, non-tender, non-fluctuant. The overlying skin and mucosa was normal (Fig 3).

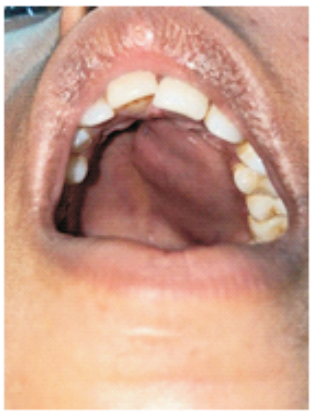

Fig 3 Photograph showing intra oral swelling in the patient

Radiographicimage revealed mixed radiolucency and radio-opacity with irregular borders and internal calcifications. An incisional biopsy was performed.

A gross specimen measuring 9mm x 8mm x 4mm in size, irregular in shape, pinkish in colour and firm in consistency was received.

After proper fixation and processing, microscopic examination revealed a well demarcated lesion, surrounding the bone. The stroma appeared loose, fibroblastic containing very fine lace-like trabeculae of immature osteoid entrapping plump fibroblasts suggesting 'trabecular' type of COF. Variable degrees of cellularity and scanty to well definedeosinophilic calcifications in a condensed stroma can be appreciated. Some basophilic areas representing presence of cementoid or dentinoid like material in the stroma which confirmed the diagnosis (Fig 4, Fig 5).

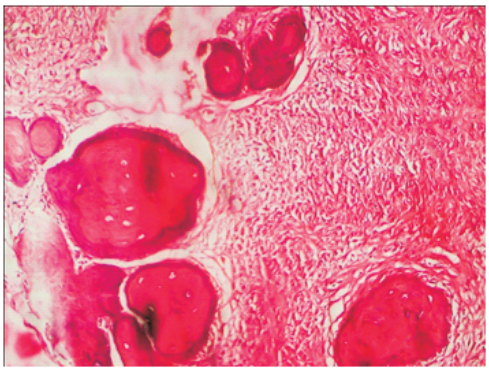

Fig 4 : H&E stained section (20X) showing woven bone formation

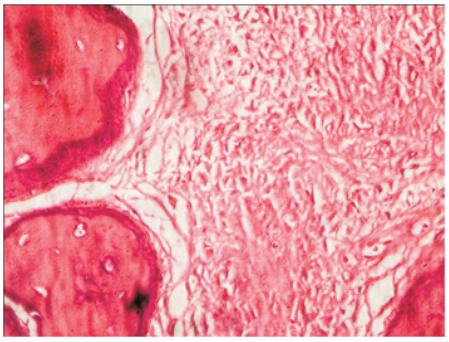

Fig 5 H&E stained section (40X) showing cementoid like deposits

On the basis of clinical, radio graphical and histopathological examination, a confirmatory diagnosis of COF was made. The lesion was surgically excised with adequate margins. One year follow-up of the patient showed satisfactory healing and no recurrence.

Discussion:

Fibro-osseous lesions represent a diverse group of benign bone disorders characterized by the replacement of normal bone with fibrous tissue and varying degrees of mineralization. Among the different fibro-osseous lesions, one that stands out for its clinical significance and unique features is COF.

COF is a relatively uncommon tumor, accounting for approximately 1-3% of all jawbone lesions. Differentiation of COF from other fibro-osseous lesions, such as cemento-ossifying fibroma and ossifying fibroma is important for appropriate management as each fibro-osseous lesion has a different biological behaviour and potential for aggressive growth[10]

The classification of central ossifying fibroma (COF) has evolved over time as our understanding of this condition has improved.

Terms like "periodontoma," "cementifying fibroma," and "ossifying-odontogenic fibroma," which refer to the conditions frequent relationship with teeth and the presence of cementum-like material, are examples of terminology modifications over time. The name "cemento-ossifying fibroma" was preferred by the WHO classification of odontogenic tumors in 1992, and it was the first to recognise a more aggressive subtype of these lesions known as "juvenile aggressive ossifying fibroma." [11]

The 2005 World Health Organisation (WHO) classification of odontogenic tumors includes central ossifying fibroma (COF) as a distinct entity. According to this classification, COF is classified under the category of "Odontogenic tumors derived from the ectomesenchyme," specifically under the subcategory of "Benign neoplasms"

Currently, COF is classified based on its histopathological features, which help differentiate it from other fibro-osseous lesions and provide insights into its behaviour. The two main histopathological variants of COF are as follows:

Central Ossifying Fibroma (Cementum-like Type):

This variant is characterised by the presence of cementum-like material within the fibrous stroma. Histologically, it shows the formation of acellularcementum-like tissue, which resembles normal cementum found on the root surface of teeth. This type of COF is less cellular and has a more well-defined appearance. It typically exhibits slow growth and a lower recurrence rate compared to the other variant.

Central Ossifying Fibroma (Osteoblastoma-like Type):

This variant of COF is characterised by the presence of woven bone trabeculae within the fibrous stroma. Histologically, it shows a highly cellular fibroblastic stroma with prominent osteoblastic activity, leading to the formation of woven bone trabeculae. This type of COF is more cellular and exhibits a more aggressive growth pattern, with a higher tendency for recurrence.

In addition to these two main variants, there may also be cases that exhibit features of both types, showing a combination of cementum-like material and woven bone trabeculae.

To better reflect the fact that these lesions develop in the tooth-bearing regions of the jaws and are benign mesenchymal odontogenic tumors that likely originate from the periodontal ligament, the WHO consensus panel agreed to restore the term "cemento-ossifying fibroma" (COF) in 2017. [12]

The term cemento-ossifying is still used in the most recent WHO classification (2022) for a variant that develops in the tooth bearing areas and is assumed to have an odontogenic origin [13]. Due to the wide age range of the psammomatoid

variation, the term "juvenile" is also removed in the new categorisation. Cemento-ossifying fibroma, juvenile trabecular ossifying fibroma, and psammomatoid ossifying fibroma are the three varieties recognised by the WHO.

The classification of COF based on its histopathological features helps guide treatment decisions and provides prognostic information. The cementum-like type of COF tends to have a more favourable prognosis, with a lower recurrence rate, while the osteoblastoma-like type may exhibit a more aggressive behaviour and a higher recurrence rate.

Clinical, radiographic, and histopathological findings aid in distinguishing COF from entities such as cemento-ossifying fibroma, ameloblastoma, and fibrous dysplasia.

COF commonly presents as a slow-growing, painless swelling in the affected jaw. Radiographic features typically include a well-defined radiolucent lesion with variable calcifications.

COF displayed a 2:1 preference for females. Although, in a case study given by Eversole et al., this female preference was significantly higher (5:1). [14]

Liu et al. discovered a male predominance in their retrospective case series among individuals under the age of 18. [15]

Clinically, the lesions typically manifested as asymptomatic swellings without any additional symptoms, though there have been rare reports of pain, tenderness, sinus discharge, facial deformity, and eye protrusion.

The ability of the tumour to enlarge is more in maxilla as it may extend in the maxillary sinus.

Mac Donald Jankowski [16] described 66 cases and discovered that they were all clearly defined unilocular, round, or oval forms. Larger tumors may seem multilocular on radiographs. The inference was that the radiographic appearance goes through three stages. The lesion starts out radiolucent (osteolytic picture), then gradually turns radiopaque as the stromamineralises, resulting in a mixed lesion. Individual radiopacities eventually combine to the point that the mature lesion can take on the appearance of a sclerotic or radiopaque lesion. Additionally, he provided a summary of the radiological characteristics in 177 cases of cemento-ossifying fibromas that had been described in the literature and in his personal files, showing that 42% of them were radiolucent, 24% were radiopaque, and 34% had a mixed appearance. Our case was categorized radiologically as a mixed type.

They exhibit aggressive behaviour locally, have a high recurrence rate, especially after partial and incomplete excisions, and the best course of treatment is full removal. In our case, surgical resesction followed by bone grafting was done. After 1 year of treatment, the patient did not turn up for follow-up due to financial constraints.

Treatment Modalities:

During the past few years, there has been a marked advancement in the treatment landscape for COF. It is evident that refining treatment strategies for COF is highly important because of its potential for local aggressiveness and associated morbidity.

In the past, surgical excision has stood as the cornerstone of COF treatment, seeking to remove the entire lesion and minimize the chances of recurrence. There has been a challenge in finding a balance between the eradication of lesions while preserving functionality and aesthetics at the same time. It should be noted that, however, that contemporary imaging modalities such as cone-beam computed tomography (CBCT) have revolutionized preoperative planning, enabling surgeons to plan precise surgical approaches based on high-resolution 3D images.

The use of minimally invasive surgical techniques, guided by CBCT, has emerged as one of the most significant advances in the management of COF. Using this technique ensures that not only can the lesion be removed accurately, but also that as much damage as possible to adjacent critical structures is minimized, which leads to improved postoperative outcomes and fewer complications.

Furthermore, the integration of adjuvant therapy, such as bisphosphonates and calcitonin, in the treatment of COF appears to have a promising effect in reducing the rate of recurrence.

In recent years, molecular studies have shed light on the pathways leading to COF pathogenesis and opened the door to targeted interventions. Various therapies are available for treating COF that aim to disrupt the signaling pathways that

are responsible for the aberrant osteogenic activity. As of late, various studies have indicated that cytogenetics might provide an ancillary method of diagnosing benign FO lesions due to the fact that they possess distinct karyotypic abnormalities (for example, ossifying fibromas possess a characteristic chromosomal distribution). Toyosawa et al. [20], in another study, suggest that while the diseases of FO (especially fibrous dysplasia and ossifying fibroma) may seem similar, their differences can be found using immunohistochemical detection of osteocalcin expression as well as PCR analysis with PNA to determine whether they have the same mutations as the GNAS codon at Arg (201) [18].

Although in nascent stages, such approaches hold potential to revolutionize COF treatment, ushering in personalized, effective interventions.

The recurrence rates for these lesions can vary significantly. Cemento-ossifying fibromas, for example, tend to have a higher recurrence rate compared to ossifying fibromas which is around 27% [17]. Understanding the specific lesion type helps in planning long-term patient care and monitoring for potential recurrences.

To reduce the risk of recurrence in COF and other fibro-osseous lesions, careful preoperative planning, comprehensive surgical excision with an adequate safety margin, and appropriate reconstruction techniques are essential.

Conclusion:

Central ossifying fibroma (COF) is a rare benign neoplasm that predominantly affects the mandible and rarely seen in maxilla. It is characterised by the proliferation of fibroblasts and the formation of mineralised tissue.

COF in our case typically presents as a slow-growing, painless swelling in the maxillary region. Radiographic imaging reveals a mixed radiolucent and radio-opaque lesion with irregular borders and internal calcifications. The differential diagnosis should include other benign and malignant jaw tumors, such as ameloblastoma, odontogenicmyxoma, and osteosarcoma.

Histopathological examination is necessary for the definitive diagnosis of COF. Microscopic analysis reveals fibroblastic proliferation and the presence of variable amounts of mineralised tissue. Surgical excision with wide margins is the treatment of choice for COF in the maxilla. Preservation of vital structures and reconstruction of the defect may be necessary, depending on the extent of the lesion. Long-term follow-up is essential to monitor for recurrence or complications.

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