Case Report

A 35-year-old male patient reported to the dental hospital with the complaint of pain and pus discharge in the right maxillary molar region for two weeks.

History revealed that the patient noticed a small swelling in the right side of the face eight years back which had gradually grown to attain a huge size, on the face as well as intra-orally. There was no pain, tenderness or any discharge associated with the swelling but 15 d back the patient developed pain in the right maxillary molar teeth with pus discharge from the intra-oral swelling which did not subside on taking medications. He also had nasal stuffiness and restricted mouth opening for some months.

On clinical examination a diffuse swelling was seen in the right side of the face, ovoid in shape extending from the right inferior orbital margin to the angle of the mouth supero-inferiorly and from the bridge of the nose up to 3cm from the tragus of the ear antero-posteriorly, about9x5cm in size, bony hard in consistency and non-tender on palpation [Table/Fig-1,2a,b]. Due to the swelling there was obliteration of the nasolabial sulcus and displacement of the bridge of the nose and the angle of the mouth. Mouth opening was restricted; the inter-incisal distance being 32mm. Intra-orally a large swelling was seen extending from the canine to second molar region, obliterating the buccal sulcus and with expansion of the buccal cortex [Table/Fig-3]. There was the presence of a pus discharging sinus in the periapical region of maxillary first premolar with flaring of the buccal root, severe gingival recession, mobility and tenderness. Correlating the history of a long standing swelling, presence of a bony swelling with considerable expansion and an intra-oral pus discharging sinus led to provisional diagnosis of a benign bony lesion with secondary infection.

Clinical Differential Diagnosis
1. Craniofacial fibrous dysplasia
2. Infected dentigerous cyst
3. Adenomatoiododontogenic tumour
4. Central Ossifying Fibroma

Investigations

Periapical radiograph of maxillary premolar-molar region showed indistinct trabeculae with patchy radio-opacities.

Radiologic Differential Diagnosis
1. Craniofacial fibrous dysplasia
2. Calcifying odontogenic cyst
3. Calcifying epithelial odontogenic tumour
4. Central ossifying fibroma
5. Osteosarcoma

The differential diagnosis was based on radiologic findings. Presence of cotton wool appearance filling the antrum unilaterally is a feature of fibrous dysplasia, while well-defined expansive mixed radio-opaque-radiolucent appearance is a finding in calcifying odontogenic cyst, calcifying epithelial odontogenic tumour, central ossifying fibroma and osteosarcoma.

Incisal Biopsy: showed the presence of a cellular connective tissue stroma spindle and stellate shaped cells and numerous areas of calcification resembling bone along with osteoblastic rimming and cementum like areas. There was evidence of moderate vascularity. [Table/Fig-10].

A final diagnosis of COF was made but the patient denied treatment due to financial constraints and could not be followed-up.

DISCUSSION

Central ossifying fibroma (COF) is a benign neoplasm designated as a fibro-osseous lesion by Eversole [1]. A bewildering variety of neoplasms, both benign and malignant, are encountered in the maxillofacial region which pose a diagnostic challenge owing to the complexity of the anatomical structures and diversity of odontogenic and non-odontogenic lesions arising in the region. A pathologic

Keywords: Benign fibro-osseous lesion, Huge tumour, Non-odontogenic neoplasm, Radiopaque-radiolucent lesion, Secondary infection

process in which the normal bone architecture is substituted by fibrous tissue containing variable amounts of mineralized material leads to the formation of Fibro-osseous lesions (FOL) [1]. FOL of the maxillofacial bones comprise of a diverse group of lesions which may be developmental, reactive or neoplastic [Table/Fig-11]. Irrespective of the type of FOL, all exhibit replacement of normal bone by fibrous connective tissue with some type of mineralized substance which may be mature or immature bone, cementum or osteo-cementum like calcifications.

In 1872 Menzel first described Cemento-ossifying fibroma but it was Montgomery in the year 1927 who coined the term OF. It was thought that Fibrous dysplasia (FD) and OF were variants of the same entity till 1948 when Sherman and Sternberg described OF in detail and since then OF has been considered as a distinct lesion [2].

The WHO first classified cementum containing lesions to be of four types: Fibrous dysplasia, ossifying fibroma, cementifying fibroma and cemento-ossifying fibroma. The next WHO classification categorized fibro-osseous lesions of the maxillofacial region into two types: osteogenic neoplasms and non-neoplastic bone lesions. In 1992, the WHO further revised the nomenclature of the separate neoplasms of ossifying fibroma and cementifying fibroma as a single entity of “cemento-ossifying fibroma”. The latest 2005 classification of the WHO now classifies cemento-ossifying fibroma as a benign fibro-osseous neoplasm which is included among the non-odontogenic tumours, arising from the mesenchymal blast cells of the periodontal ligament, with a potential to form either fibrous tissue, bone, cementum, or a combination of such tissues [3]. The terms “cementifying, cemento-ossifying and ossifying fibroma” were reduced to a single term Ossifying Fibroma.

The origin of OF has been attributed to the periodontium as it has the potential to form both cementum and bone. Chronic inflammation and fibrosis of the periodontium due to periapical infection or injury to the periodontium may stimulate the formation of the lesion. But the pathogenesis of this lesion is controversial as neoplastic lesions of similar histology have been reported from other bones of the face and the long bones which lack the periodontal ligament and sometimes referred to as ‘cementiform fibrous dysplasia’. It has been hypothesized by Brademann et al., that primitive mesenchymal cells in the other bones may differentiate into periodontal ligament and local trauma may be a factor in the induction of proliferation of OF [4].

All the FOL share some common clinical characteristics like slow growth and lack of any symptoms. As the lesions enlarge, they cause cortical expansion leading to facial asymmetry. If the tumour mass impinges on any nerve there may be pain or paraesthesia. Teeth
in the vicinity of the lesion retain their vitality and root resorption is uncommon. COF commonly occurs between the third and fourth decades of life and shows a strong female gender predilection of 5:1. In about 70-90% of all cases the mandible is affected [5]. We have used the term ‘giant ossifying fibroma’ as the tumour in our case measured more than 8cm in greatest diameter and is the largest to be reported in the maxilla of a male patient [6]. The presence of an intra-oral pus discharging sinus was suggestive of secondary bacterial infection as the tumour mass was in close vicinity of the oral mucosa.

The radiologic appearance of COF is variable, depending on the degree of mineralization of the lesion. A well-defined unilocular or multilocular lesion is seen with an internal structure which is entirely radiolucent or a mixed radiopaque-radiolucent appearance [7]. Sometimes a radiolucent line is seen in the periphery of the lesion suggestive of a capsule and is referred to as ‘Rind sign’ [8]. Generally teeth or roots are displaced but root resorption is uncommon. COF commonly occurs between the third and fourth decades of life and shows a strong female gender predilection of 5:1.

Histological picture exhibits hypercellularity of fibrous stroma interspersed with calcified islands of osteoid, bone or cementum. The bony trabeculae are of variable sizes showing both lamellar and woven patterns. Peripheral osteoid and osteoblastic rimming are seen [9]. Peripheral brush borders are seen blending into the connective tissue from the spherules of cementum like material. But, there is controversy regarding the histopathologic features of COF and FD [10]. The only distinct feature of OF is presence of a fibrous capsule. As the lesion matures the calcific deposits increase but no cellular atypia or mitotic figures are seen.

As COF and FD resemble each other in their clinical, radiological as well as histopathologic features both need to be differentiated during diagnosis. [Table/Fig-12] summarizes the comparison between both entities.

Since COF is well circumscribed and well demarcated from the surrounding bone, complete surgical removal poses no problems in case of small lesions, especially in the mandible. In case of maxillary tumours as the present case filling the antrum, complete surgical removal is challenging [11]. The recurrence rate after surgical removal is about 6-28% which further increases for maxillary lesions. Despite their tendency for local invasion and recurrence, COF carries a good prognosis.

A comparison was done with five recently published case reports of COF as detected in a PubMed search [12-16]. It was seen that presence of a swelling and facial asymmetry was the common complaint, the age ranged from 11 to 80 y with a definite female gender predilection with a ratio of 4:1 and the mandible was the commonly affected site. Clinically there was the presence of a well circumscribed, non-tender bony hard swelling and radiographic appearance was similar revealing a well-defined expansile lesion consisting of a mixed of radiolucent-radiopaque internal structure. Though two cases had huge lesions of more than 10cm diameter [15,16], none had secondary infection with the presence of an intra-oral pus discharging sinus which makes the present case unique.

CONCLUSION

Central ossifying fibroma is a benign fibro-osseous neoplasm which occurs as a mixed radiopaque-radiolucent lesion. Rarely such large lesions as the present case are encountered. However, it is essential to diagnose differentiating it from other odontogenic lesions and follow up over a long term.

REFERENCES


PARTICULARS OF CONTRIBUTORS:
1. Reader, Department of Oral Medicine & Radiology, Institute of Dental Sciences, Bhubaneswar, Odisha, India.
2. Reader, Department of Oral Medicine & Radiology, Dasmesh Institute of Research & Dental Sciences, Talwandi Road, Faridkot, Punjab, India.
3. Reader, Department of Oral Pathology & Microbiology, Kalika Dental College, Meerut, Uttar Pradesh, India.
4. Senior Lecturer, Department of Oral Medicine & Radiology, Institute of Dental Sciences, Bhubaneswar, Odisha, India.
5. Senior Lecturer, Department of Oral Medicine & Radiology, Institute of Dental Sciences, Bhubaneswar, Odisha, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Satya Ranjan Misra,
Prasanti Kathogola Road, Mangalabag, Cuttack-753001, Odisha, India.
E-mail: drsatyaranjandds@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.