Case Report

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Dentistry Section

A Case of Junvenile Psammomatoid Ossifying Fibroma in Mandible: An Unconventional Variant

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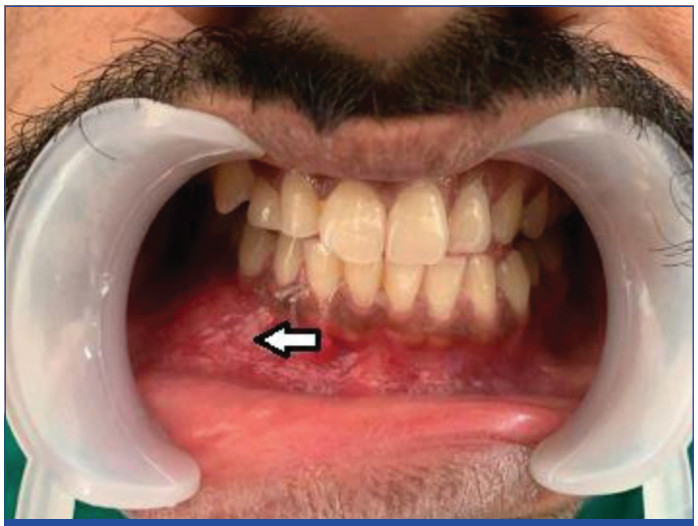
ABSTRACT

The uncommon fibro-osseous tumour known as ossifying fibroma is composed of both mature bone and fibrous tissue. It is divided into subgroups known as conventional and juvenile. Both variations affect the craniofacial bones, with the juvenile trabecular form being more common in the jaws and the juvenile psammomatoid variety being more prevalent in the paranasal sinuses. Hereby, the authors present a case report of 21-year-old male patient with extremely rare case of mandibular Juvenile Psammomatoid Ossifying Fibroma (JPOF), in which a patient reported a slow-growing, painless swelling of the mandible. After surgical removal of the tumour, histological analysis revealed that it was a psammomatoid variation of juvenile ossifying fibroma with numerous psammoma-like ossifications in the fibrous stroma. In this case study, authors discussed JPOF occurring at an unusual site, emphasising the importance of early detection, clinical findings, imaging results, differential diagnosis, histological characteristics, and conservative treatment of JPOF without evidence of recurrence.

Keywords: Craniofacial bones, Psammoma, Paranasal sinuses

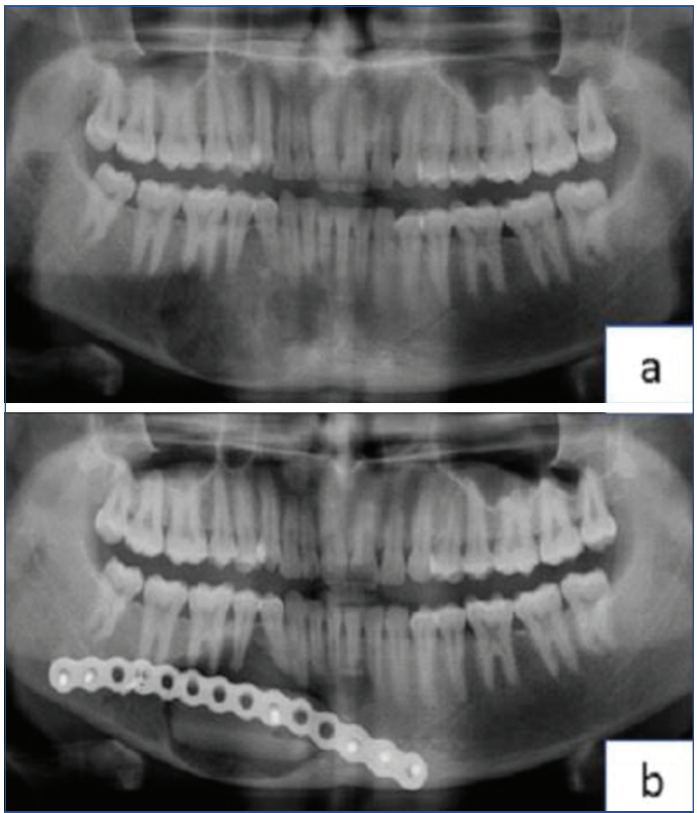
CASE REPORT

A 21-year-old male patient presented with a progressive, painless swelling over the right half of his face for one and a half years. He had no significant medical history or family history. Upon extraoral examination, the skin appeared typical in appearance. During intraoral examination, no gross swelling was noted, but expansion of the buccal and lingual cortical plates was observed, extending from tooth 42 to 47; the mucosa over the lesion appeared stretched [Table/Fig-1]. The swelling was firm to hard, non tender, non fluctuant, and non compressible upon palpation. The associated teeth showed no abnormalities. The right submandibular lymph nodes were palpable and tender. A provisional diagnosis of benign odontogenic tumour was made based on the clinical examination. Fibrous Dysplasia (FD) and ossifying fibroma were considered as differential diagnosis.



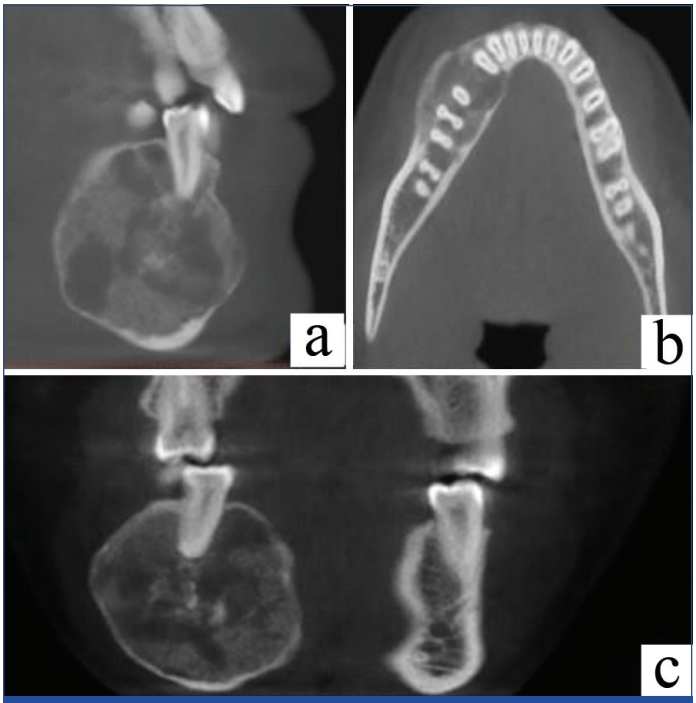
[Table/Fig-1]: Diffused swelling on right mandibular region showing buccal cortical plate expansion.

An Orthopantomogram (OPG) revealed a well-defined, circular-shaped lesion extending from the distal surface of the root of the right lateral incisor to the mesial aspect of the right second molar. The interior of the lesion exhibited a mixture of radiolucent-radiopaque structures, giving it a typical cottonwool appearance. Displacement and resorption of the apical one-third of the roots of teeth 44 and 45 were observed [Table/Fig-2a,b]. Cone Beam Computed Tomography (CBCT) demonstrated a multilocular lesion with a mixed hypodense and hyperdense interior. Expansion of the buccal and lingual cortical plates was evident without any signs of perforation [Table/Fig-3a-c]. Haematological and urine investigations yielded results within normal ranges.

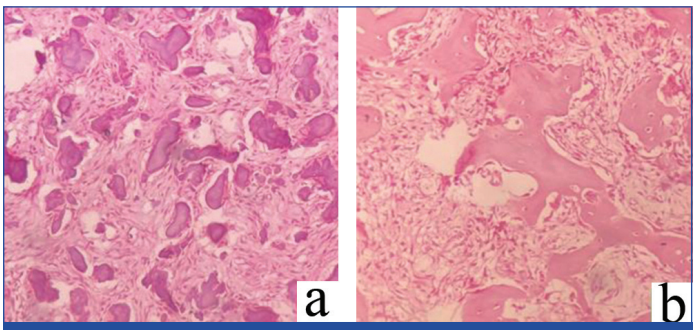


[Table/Fig-2]: Orthopantomogram (OPG): a) Preoperative OPG shows a well-defined, circular-shaped lesion, interior of the lesion showing mixed radiolucent-radiopaque structures, giving it a typical cotton wool appearance; b) Postoperative OPG shows the defect space filled with a fibula graft support and secured using a reconstruction plate and locking screws.

The lesion was scraped, healthy bleeding was induced, and the defect space was filled with a non vascular fibula graft to provide additional support, secured using a reconstruction plate and locking screws [Table/Fig-2b]. Routine histopathological examination of an incisional biopsy with Haematoxylin and Eosin (H&E) stain revealed tissues composed of connective tissue stroma with fibroblastic proliferation and the presence of numerous basophilic irregular and spherical calcified ossicles. These structures were relatively acellular, with a concentric pattern of lamination resembling psammoma bodies, spread homogeneously within the entire tissue section [Table/Fig-4a,b]. Consequently, the diagnosis of JPOF was confirmed. Follow-up at 6-month and one-year intervals showed no signs of recurrence.



[Table/Fig-3]: Cone Beam Computed Tomography (CBCT) section: a) Sagittal section; b) Axial section; c) The coronal section shows the expansion of buccal and lingual cortical plates with the interior of the lesion showing a multilocular pattern with a mixed hypodense and hyperdense structure without evidence of any perforation.



[Table/Fig-4a,b]: Multiple bits of tissues composed of connective tissue stroma with fibroblastic proliferation. Numerous calcifications are seen in the form of basophilic small round bodies (Psammomatoid) (H&E stained 10x, 40x, respectively).

DISCUSSION

The unusual form of ossifying fibroma, known as juvenile ossifying fibroma, exhibits more aggressive behaviour than the typical kind. It arises in the orbit and paranasal sinuses of the craniofacial skeleton; in contrast, the traditional ossifying fibroma usually occurs in the jaw [1]. According to the World Health Organisation (WHO) classification 2017, ossifying fibroma is classified into three types: cemento-ossifying fibroma, Juvenile Trabecular Ossifying Fibroma (JTOF), and JPOF [2]. Juvenile ossifying fibroma is a rare, benign, bone-forming tumour with aggressive growth [3]. Most of them affect the extragnathic bones; a few may occasionally affect the craniofacial bones and maxilla but infrequently the mandible [4].

"Psammos" is derived from the Greek word "psammos," which signifies grains of sand. The term "juvenile" was dropped earlier because these lesions are seen into adulthood as well, as presented in present case. However, it is proposed that these lesions start in adolescence and only become noticeable in adulthood when they have grown to a significant size [3,5]. According to Johnson LC et al., JPOFs are caused by an excess of myxofibrous cellular stroma, which is involved in the formation of the paranasal sinus septa, so this variety typically occurs in the craniofacial bone but has an unusual site of involvement as in the above case [2,6].

The fibroblastic stroma in POF is characterised by spherical calcifications that resemble psammoma bodies. Cases have shown that it is typical to see aneurysmal bone cyst-like development and cystic degeneration [7]. Grossly, JPOF is tan-white, greyish-white, or greyish-brown in colour, firm to hard in consistency, and separated from the surrounding bone, albeit not encapsulated [5]. Depending on the degree of calcification and the extent of the cystic alterations, the internal structure of JPOF might be radiolucent, mixed, or radiopaque. While resorption is uncommon, it can happen, and root displacement is frequent, as seen in the above case. Severe cases can induce perforation, cortical thinning, and expansion [5,8]. Difficulty in breathing, swallowing, proptosis, and eye-watering have all been reported in cases with JPOF involving the maxilla and other cranial bones [2,4]. The mandibular lesion exhibits a centrifugal development pattern, resulting in bowing of the inferior cortex of the jaw [8].

The JPOF appears multiloculated internally on Computed Tomography (CT) and is bounded by a thick layer of bone density with varied densities within. Psammomatoid ossicles resemble ground glass or are radiolucent [5]. As per Owosho A.A et al., there exist three distinct patterns of JPOFs on CT scans: 1) a solid, homogeneous radiopacity; 2) a solitary ground glass mural nodule; or 3) a radiolucent core surrounded by an outer thick mantle [2]. The T1 isointense and T2 hypointense appearances of the sclerotic rim on Magnetic Resonance Imaging (MRI) exhibit enhancement upon contrast delivery. The solid component of the lesion appears isointense to muscle on T1 imaging and isohypointense on T2 images [1]. According to reports, JPOF's Positron Emission Tomography (PET) scan results indicate a significant uptake that is greater than the surrounding anatomical structures [9]. Imaging and histology are used to make the diagnosis. All expanding lesions originating from the facial bones, such as FD, osseous dysplasia, JTOF, mucocele, or other types of ossifying fibroma, are included in the differential diagnosis of JPOF. Based on the tumour's location, characteristics of the tumoural margins, and the presence of a ground-glass pattern, the differential diagnosis can be narrowed [2,5]. FD can be distinguished from other expanding lesions by its poorly defined boundaries. Depending on the lesion's maturity, FD can occur in any part of the skeleton and exhibit ground glass opacities. Since these conditions do not have this composition, ground glass opacity and diffused border make it possible to rule out JPOF and ossifying fibroma [10]. Mucocele can grow into sizable lesions with smooth remodeling walls, alterations in the bone, and expansion into the surrounding tissue. Additionally, the lesion's location aids in the diagnosis [2]. Even though it is uncommon to discover JPOF in the mandible, in present case, it was associated with the mandible only. There have only been 13 cases of JPOF affecting the mandible recorded since the year 2000 [11]. A case involving the mandible in a seven-year-old boy showed a pre-existing lesion of JPOF developing into an aneurysmal bone cyst [12]. JPOF also needs to be differentiated from osteoblastoma, osteosarcoma, and primary aneurysmal bone cysts, in addition to other fibro-osseous disorders. The radiologic and histologic similarities between eosinophilic granuloma and intraosseous cavernous haemangioma should also be considered [7]. Conservative excision or curettage is

the treatment for JOF; more aggressive treatments may be required for specific lesions. Continuous follow-up is crucial because the recurrence rate for JPOF varies from 30% to 58% [4].

Early discovery of a lesion can limit the size of the defect. Examination and treatment of these lesions may benefit from a multidisciplinary approach involving a radiologist, pathologist, neurosurgeon, craniofacial surgeon, and orbital specialist. Partial or incomplete resection can lead to recurrences. Because the tumour is believed to be radioresistant, radiation therapy is not advised. Early detection and complete surgical excision are the preferred course of treatment.

CONCLUSION(S)

The JPOF are rare benign fibro-osseous tumours; they do not have distinct clinical or radiologic features, resulting in a diagnostic challenge, but judicious histopathologic findings provide confirmation. Considering the aggressive nature of the tumour, large lesions are treated with surgical resection along with adequate margins of normal tissue to avoid recurrence. Meticulous reconstruction of the bony defect and frequent follow-ups are required for rehabilitation.

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