Case Report

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Dentistry Section

Juvenile Psammomatoid Ossifying Fibroma: A Rare Case Report

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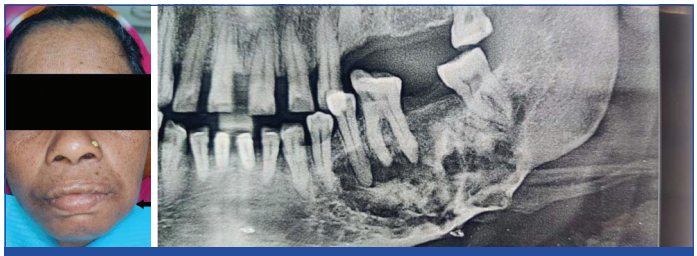
ABSTRACT

Juvenile Ossifying Fibroma (JOF) is a rare, non cancerous overgrowth of bone in the face or jaw. There are two subtypes of JOF: Juvenile Psammomatoid Ossifying Fibroma (JPOF) and Trabecular Juvenile Ossifying Fibroma (JTOF). JPOF shows a modest male predominance (1.2:1), an age range of 3 to 49 years, with a mean age of 17.7 years. The majority of instances are associated with the orbital bones and paranasal sinuses. Proptosis is the most typical clinical sign of JPOF. Other features include nasal blockage, headaches, facial oedema, discomfort, recurrent sinusitis, and missing teeth. The lesion is identified under a microscope by a fibroblastic stroma that contains small ossicles resembling psammoma bodies. The preferred first-line treatment for JPOF is total resection due to the disease's potential for rapid growth and recurrence. Hereby, the authors present a rare case of JPOF ossifying fibroma occurring in the left mandibular molar region of a 50-year-old female who complained of continuous expansion of the lower jaw. Although benign, the potential for recurrence necessitates follow-up and can impact treatment planning. Early diagnosis and intervention are crucial for a favourable outcome.

Keywords: Cemento ossifying fibroma, Juvenile trabecular ossifying fibroma, Ossicles, Psammoma bodies

CASE REPORT

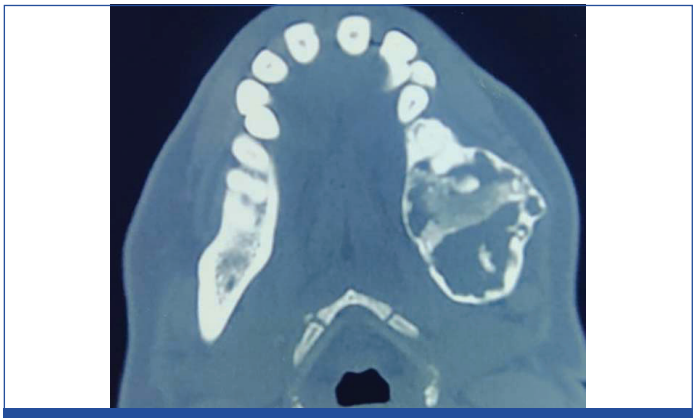
A 50-year-old female reported to a private dental clinic with a chief complaint of a slow-growing swelling in the lower left back tooth region for 15 years. The lesion was steadily expanding in the left mandibular molar region and was not associated with pain or discharge. She did not have any significant family or medical history. On extraoral examination, the swelling was diffuse, extending from the parasymphyseal area to the posterior edge of the mandibular ramus. The overlying skin appeared normal, and no signs of lymph node involvement were observed. Upon intraoral examination, a diffuse bulge extending from the distal region of the left mandibular first premolar to the left mandibular third molar was noted. The second molar was absent. Upon palpation, the swelling was hard, non tender, obliterating the left buccal vestibule, with no evidence of any discharge [Table/Fig-1].



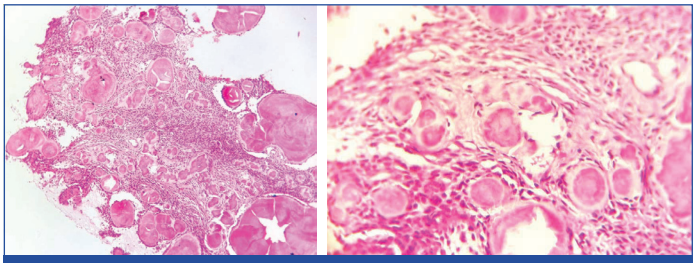
[Table/Fig-1]: Showing swelling in the left mandibular molar region.
[Table/Fig-2]: Orthopantomograph showing mixed radiolucent radiopaque lesion extending from 34 until distal aspect of 38 with well-defined borders. (Images from left to right)

An Orthopantomograph (OPG) revealed a mixed radiolucent-radiopaque lesion extending from tooth 34 to the distal aspect of tooth 38, with well-defined borders. Thinning of the lower left border of the mandible was observed [Table/Fig-2]. Axial sections of a Computed Tomography (CT) scan showed a multilocular lesion with multiple internal septa, an intact lower border of the mandible, and formation of internal septa along with areas of homogeneous calcification [Table/Fig-3]. No root resorption was noted. Based on the clinical and radiographic features, a provisional diagnosis of a benign odontogenic lesion was made.

An incisional biopsy of the lesion was performed, and the tissue was sent for histopathological examination. The Haematoxylin and Eosin (H&E) stained tissue section of the lesional tissue revealed a fibrocellular connective tissue stroma composed of bony spicules and ossicles [Table/Fig-4]. The ossicles varied in appearance, ranging from small, round shapes to large, irregular shapes resembling psammoma bodies. The intervening connective tissue was composed of polyhedral to spindle-shaped cells with prominent basophilic nuclei. Blood vessels of varying sizes and shapes were also observed. Correlating with the clinical, radiographic, and histopathological features, a final diagnosis of JPOF was made [Table/Fig-5]. The patient was referred to a higher centre for further management of the lesion.



[Table/Fig-3]: Axial sections of computed tomography scan showing multilocular lesion with multiple internal septa.



[Table/Fig-4]: Photomicrograph showing fibro cellular connective tissue stroma composed of bony spicules and ossicles (H&E, 10X).
[Table/Fig-5]: Photomicrograph showing ossicles resembling psammoma bodies and intervening connective tissue composed of polyhedral to spindle shaped cells (H&E, 40X). (Images from left to right)

DISCUSSION

The English term "psammoma" comes from the Greek word "psammos," which means "sand [1]." The various terminologies used in the literature are noted [Table/Fig-6] [2-9]. The cause of JPOF, an unusual type of aggressive ossifying fibroma, remains unknown.

Name of the author	Year	Terminology
Benjamins CE [2]	1938	Osteoid fibroma with atypical ossification
Gögl H et al., [3]	1949	Psammomatoid ossifying fibroma
Johnson LC et al., [4]	1952	Juvenile active ossifying fibroma
Makek M and S Karger AG [5]	1983	Psammous desmo-osteoblastoma (variant of osteoblastoma)
Wenig BM et al., [6]	1995	Aggressive psammomatoid ossifying fibromas
Hartstein ME et al., [7]	1998	Psammomatoid ossifying fibromas
Barnes L et al., [8]	2005	Juvenile Psammomatoid Ossifying Fibroma (JPOF)
Soluk-Tekkesin M and Wright JM [9]	2022	Juvenile Psammomatoid Ossifying Fibroma (JPOF)

[Table/Fig-6]: Different terminologies used for the lesion [2-9].

According to Sarode SC et al., the abundance of myxofibrous cellular stroma, which is often implicated in the formation of septa in the paranasal sinuses during their enlargement and pneumatisation, is assumed to contribute to the formation of POF [10]. These stromal cells release connective tissue mucin, which initiates the cystic areas, and hyaline material, which ossifies [10]. Makek M, examined 86 instances of JPOF, with a modest male predominance (1.2:1), an age range of 3 to 49 years, and a mean age of 17.7 years [5]. This contrasts with present case, which occurred in an elderly female aged 50 years [5]. The majority of instances (61.6%) were associated with the orbital bones and paranasal sinuses; the remaining cases were associated with the maxilla (19.7%), mandible (7%), parietal bones (4.6%), temporal bones (3.5%), and frontal bones (3.4%) in that order of incidence. Johnson LC et al., also noted a slight male predilection [4]. Total 70% of instances were located in the paranasal sinuses, with the remaining cases in the maxilla (20%), mandible (10%), and calvaria (10%) [11]. Conversely, Slootweg PJ et al., reported 23 cases, 16 of which included the maxilla (6 cases), the paranasal sinuses (2 cases), and the rest in the mandible (1 case); the lesion occurred in the posterior region of the mandible [1].

Proptosis is the most typical clinical sign of JPOF. The eyeball may occasionally move laterally. There have also been reports of vision loss, with progression to blindness. Other clinical observations include headaches, facial oedema, discomfort, and recurrent sinusitis. There are numerous reports of missing teeth in the literature. Local invasion can take many forms, such as "bowing" or "pushing" into neighbouring bony boundaries or penetrating through osseous delimiting walls and extending into nearby anatomic compartments. When it affects the jaw, cortical plate enlargement and painless swelling are the hallmarks. With an extension of the buccal and lingual cortical plates, the neighbouring area in the mandible most frequently appears in the ramus rather than elsewhere in the body [12]. Similar oral findings were observed in the present case. Some studies indicate that the tendency toward aggressive growth and recurrence is age-related, with younger age groups experiencing it more frequently [12]. The aggressive behaviour may originate from the lesion's future progression, resulting in bone erosion and extension to nearby critical structures, considering the small anatomical boundaries of the sinonasal tract [12].

The JPOFs exhibit thin sclerotic rims around radiolucent and radiodense regions on radiography. They resemble various bone lesions such as osteoblastoma, aneurysmal bone cysts, cemento-ossifying fibromas, fibrous dysplasia, and osteogenic sarcoma [11]. On the basis of neighbouring location, ground-glass lesions, and peripheral margin identification, a large number of differential diagnosis can be ruled out. However, a ground-glass appearance could also Indicate a Fibrous Dysplasia (FD) lesion; therefore, other characteristics should be used when differentiating between FD and JPOF. JPOF is well-circumscribed, while FD tends to spread with ill-defined margins. Moreover, FDs typically maintain the general shape of the affected bone, whereas JPOFs take on a spherical shape. Mucoceles initially appear as sizable, well-defined lesions that extend into the nearby soft tissues. While osseous dysplasias are frequently observed in the periapical area, mucoceles begin in the nasal sinuses, and JTOF typically develops from the maxilla or mandible. Psammomatous bodies may not be present in JTOF, and osseous dysplasias and Odontogenic Fibromas (OF) have mixed features. Since JPOFs typically affect children, other aggressive bone lesions in children, such as chondrosarcoma and Ewing sarcoma, should be considered, as the treatments for these illnesses differ from those for JPOFs [13].

The lesion is identified under a microscope by a fibroblastic stroma containing small ossicles that resemble psammoma bodies. Gögl H was the first to refer to these distinct spherical formations as "psammoma-like bodies [3]." Ultrastructurally, it was found that psammoma-like structures in PJOF had a black crystal rim from which tiny spicules and needle-like crystalloids protruded toward the edge. The stroma can range in texture from loose and fibroblastic to highly cellular with little to no collagen in between. The mineralised material comprises a cellular or sparsely dispersed spherical or curved ossicles. PJOF occasionally includes irregular thread-like or thorn-like calcified strands over a hyalinised background, in addition to profoundly basophilic concentrically lamellated particles. Aneurysmal bone cyst-like regions are caused by additional characteristics such as haemorrhages, pseudocystic stromal degeneration, and trabeculae of lamellar bone alongside woven bone [1].

The preferred first-line treatment for JPOF is total resection due to the disease's potential for rapid development and recurrence. Complete excision, however, may be challenging for functional as well as aesthetic reasons. If there is a recurrence, more surgery might be required. Nowadays, autologous bone grafts are regarded as the best alternative for reconstruction. Autologous bone transplants can be utilised to repair skeletal defects, promote bone healing, and provide supporting structures. When autologous bone grafts are used, a combination of osteoconductive, osteoinductive, and osteogenic effects has been observed. The recurrence rate of JPOFs after surgery varies from 30% to 56% [12].

CONCLUSION(S)

The JPOF is a rare and aggressive type of ossifying fibroma with an unclear aetiology, often presenting in younger individuals but also occurring in older patients. The primary treatment approach is total surgical resection, although this can be challenging due to functional and aesthetic considerations. Recurrence rates postsurgery are significant, highlighting the need for ongoing monitoring and potential additional interventions.

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