CASE REPORT

JUVENILE PSAMMOMATOID CEMENTO- OSSIFYING FIBROMA: A Case report and Review of Literature

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INTRODUCTION:
Cemento-ossifying fibroma (COF) is considered a osseous tumor, very closely related to other lesions such as fibrous dysplasia, cementifying periapical dysplasia or cemento-osseous florid dysplasia, however, forming its own entity according to the 1992 classification of the WHO. COF is believed to be derive from the cells of the periodontal ligament. Consequently, one of its main characteristics is the massive formation of cementum, cementoid substance or calcified material in the interior of a predominantly fibrous tissue.¹

The COF is subdivided into conventional and juvenile clinicopathologic subtypes.² Juvenile ossifying fibroma (JOF) is also called aggressive ossifying fibroma due to its aggressiveness and the tendency to recur, unlike other fibro-osseous lesions.³ A recent study by El-Mofty identified two histopathological variants, trabecular JOF (TrJOF) and psammomatoid JOF (PsJOF).³

CASE REPORT:
A thirteen year old male patient came to the department of Oral Medicine and Radiology with a chief complaint of swelling on the Right side of the face since the age of 2 years. History revealed that the swelling was first noted when the patient was 2 years old, which was of a size of a marble (1 x 1 cm) and has gradually increased to a size of a lemon (3 x 3 cm) over the period of next 3 years. Patient went to a dentist who informed that swelling was due to decayed tooth and extracted a right lower back tooth. Swelling did not reduce post extraction and he was referred to this hospital (5 years back) for further treatment. Patient was operated for the same four times at different intervals (due to recurrence). Last operation was done 4 years back. Presently the swelling has again grown to the present size of 8 cm x 10 cm.

It was associated with pain and discomfort for the past 1 week. Pain was of moderate intensity, continuous and increased during night. It did not subside even after talking medications. His Medical history was non – contributory and review of systems revealed no abnormality. A solitary right submandibular lymph node was palpable measuring 1×1.5 cm, oval, firm, non-tender and freely mobile in all planes. On extra oral examination (figure 1 & 2), a solitary, diffuse swelling was seen on the right side of face measuring 8 cm x 10 cm, oval, with smooth surface extending anteriorly from the midline of mandible, posteriorly up to the angle of mandible, superiorly from 0.5 cm below the ala-tragus line, inferiorly extending into the submandibular region up to the

Abstract:
The term Juvenile (aggressive) ossifying fibroma was used in the World Health Organization classification of odontogenic tumors to describe a lesion affecting the jaws of children under the age of 15 year. Juvenile ossifying fibroma is a variant of cemento-ossifying fibroma which is classified under Fibro-osseous neoplasm by Charles A Waldron in the year 1993. Although JOF can occur anywhere in the skeleton, its highest incidence is in the facial bones, most commonly the maxilla. Mandibular and extracranial involvement are rare. The juvenile forms of ossifying fibroma are uncommon but they should be recognized and managed appropriately in view of their distinctive behaviour. Here we report a rare case of Juvenile Psammomatoid Cemento-Ossifying Fibroma of right side of mandible in a 13 year old male patient.
midline. Borders were diffuse, & no discharge was noted. Colour over the swelling was normal with that of the surrounding. On palpation, there was mild increase in local temperature and it was non-tender. Periphery of the swelling was soft and the central area was bony hard which merged into the underlying bone (Mandible). No paresthesia was elicited in the facial region. Intra orally (figure 3) a diffuse swelling was seen in the right lower buccal vestibule, obliterating the vestibule, extending from 44 up to retromolar region. Clinically missing 45, 46, 47 were noted. Swelling was non-tender & bony hard. Based on the history and clinical examination a provisional diagnosis of Odontogenic tumor was arrived and patient was subjected to further investigations.

Mandibular occlusal radiograph (figure 4) revealed a multilocular radiolucency in the region of missing 45, 46, 47 expanding both buccal and lingual cortical plates.

OPG (figure 5) revealed multilocular radiolucency in the right body of the mandible extending anteriorly from distal aspect of 44, posteriorly up to edentulous 48 region. Lower border of mandible is expanded but no discontinuity of the inferior border of the mandible was seen. Absence of trabecular pattern of bone is noted. There was increased radiodensity in the posterior part of the lesion whereas it is more radiolucent in the anterior part of the lesion.

CT (figure 6 & 7) revealed a destructive and expansile bone lesion in right side of body and ramus of mandible with a mixed hypodense and hyperdense lesion measuring 44 mm (anterioposteriorly) × 28 mm (mediolaterally) × 59 mm (superoinferiory). Presence of intact cortical borders was seen.

Incisional biopsy of the bone and the content of the lesion was done. H&E stained soft tissue section (figure 8) showed a connective tissue devoid of epithelium. The connective tissue consisted of dense irregularly arranged collagen fibres with spindle shaped and plump fibroblasts. Numerous basophilic irregular shaped islands of varying sizes were present, suggestive of cementum like calcifications; also seen were few islands contain cementocytes within lacunae. A moderate chronic inflammatory cell infiltrate was present, predominantly of lymphocytes and macrophages. Vascularity was increased. Diffuse areas of numerous extravasated RBCs were seen.

Based on all the investigation a final diagnosis of Juvenile Psammomoid Cemento-Ossifying Fibroma was made.

DISCUSSION:
The term juvenile ossifying fibroma is used to describe 2 distinct histopathologic variants of ossifying fibroma of the craniofacial skeleton. 2 The most characteristic feature of JOF, as the name suggests, is its higher incidence in children and young adults. It is most often seen in patients who are between 5 and 15 years of age (60% to 80% of cases), although examples have been diagnosed in older patients. Johnson et al have reported JOFs occurring at any age between 3 months and 72 years. Considerable debate has been going on among pathologists regarding the precise nature and classification of the juvenile ossifying fibromas, resulting in a confusing proliferation of competing nomenclatures. 2 there are many classification systems for this lesion, among them the classification by Slootweg et al is noteworthy. They have classified juvenile ossifying fibroma into two distinct groups, the JOF-WHO type and JOF-PO (psammoma-like ossicles) type, primarily based on 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. The most recent classification is by El-Mofty in which he identified two categories, trabecular JOF (JTOF) and psammomoid JOF (JPOF), based on histologic criteria. The mean age also differs in that in JPOF, it is approximately 20 years, compared with an age range of 8.5 to 12 years in JTOF.

JPOF was first reported by Benjamins, in 1938, and designated it as osteoid fibroma with atypical ossification of frontal sinus. It was later termed as psammomoid ossifying fibroma of the nose and paranasal sinuses by Golgi, in 1949. The same lesion was later termed as juvenile active ossifying fibroma by Johnson et al. He reviewed 112 cases and described juvenile active ossifying fibroma as a “cellular mass which generates many small uniform-sized osteoid bodies”. Markek reviewed 86 cases and he considered the lesion to be variant of ostoblastoma and termed it psammomatoid desmo-osteoblastoma. Clinically, the JOF often presents as a progressive and sometimes rapid expanding lesion. Although there are significant variations the histopathology of COFs is distinctive. The lesions are usually well defined together with a surrounding outer border of cellular fibrous tissue, whereas in fibrous dysplasia the lesionsal bone (that usually features in the differential diagnosis) usually has some points of continuity with adjacent normal bone. Even in the same tumor, the lesions of COF contain fibrous tissue with high cellularity or alternatively zones of almost acellular collagen. The mineralized tissue consists of woven and lamellar bone, and acellular, approximately spherical, calcified deposits resembling cementum. The diagnosis of juvenile forms of COF is made on the distinctive histology. JPOF which is more frequent, is characterized by ossicles which are roughly spherical and resemble the psammoma bodies of meningiomas set in cellular fibrous tissue. The ossicles in JPOF have a thick irregular collagenous rim compared to OF particles which have smooth contour. JTOF is recognized by presence of swollen osteocytes, trabeculae of woven bone with coarse lacunae and lining of plump osteoblasts. Bands of cellular osteoid are also found. Both JPOF and JTOF may occasionally lack a distinct fibrous rim. 6 Cystic structures apparently resulting from degeneration of myxoid stroma are often encountered in older lesions and may indicate regression of the tumor. 5 The radiologic features are variable and depending on the tumor’s location and the amount of calcified tissue produced by the tumour, the lesion will show varying degree of radiolucency and radiopacity. It may be fairly well demarcated or show invasion and erosion of the surrounding bone. 5 Entities considered in the radiographic differential diagnosis of this lesion include mixed odontogenic tumors and fibrous dysplasia. Fibrous dysplasia is excluded on the basis of the well demarcated border with a radiolucent rim instead of a blended, indistinct margin even though it also presents as a ground glass radiopacity. The overall configuration fibrous dysplasia is typically fusiform where as that of JPOF is more rounded outline. The ameloblastic fibroodontoma, mixed odontogenic tumors and odontomas also occur in younger patients and present as mixed density mass lesions in the jaws. These tumors are well demarcated,
often intimately associated with an unerupted tooth and coarse opacities similar in radiodensity to tooth structure.² Majority of the authors have suggested complete removal of the lesion at the earliest stage. Irrespective of its aggressive nature, the appropriate treatment for a benign fibro-osseous lesion, includes either curettage or enucleation, until the healthy bony margins are reached. The clinical management of JOF remains uncertain. Small lesions can be treated conservatively (curettage or enucleation). For large and irregular shaped tumors that has infiltrated sinuses and fronto nasal bones, resection with an open surgical approach, such as transfacial, is ideal.³ Radiotherapy seems contraindicated because the tumors are considered to be radio-resistant and because of the adverse effects of radiotherapy.⁶ Recurrence rate ranging from 30% to 58% have been reported.⁷

References: