

JUVENILE AGGRESSIVE CEMENTO-OSSIFYING FIBROMA OF LOWER JAW : A CASE REPORT



Medical Science

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ABSTRACT

Cemento-ossifying fibromas (COF) of the craniofacial skeleton are benign fibro-osseous neoplasms characterized by the replacement of normal bone by a fibrous cellular stroma containing foci of mineralized bone trabeculae and cementum-like material that vary in amount and appearance. The definitive diagnosis can rarely be rendered on the basis of histopathological features alone and is usually dependent upon assessment of microscopic, clinical and imaging features together. This case report will discuss the clinical, microscopic, radiological and therapeutic aspects of COF. COF commonly occurs between 30-40 years of age, mostly in males. It rarely occurs in children, as in our case of a 11-year-old female child.

Introduction:

Cemento-ossifying fibroma (COF) is a rare benign fibro-osseous jaw lesion. Other lesions in this category are fibrous dysplasia, osseous dysplasia, ossifying tumor, fibroma, cemento-ossifying fibroma and cemental dysplasia. WHO classified this tumor as non-odontogenic tumor derived from mesenchymal blast cells of periodontal origin that are able to form fibrous tissue bone and cementum with a combination of all the three elements. It is seen between the third and fourth decades of life and when it arises in children, it is called as juvenile aggressive COF. ¹ COF is more frequent in women than in men (4:1). The most common location is the mandible, with 70-90% of all cases.¹⁻²

Case History:

A 11-year-old female was admitted in department of plastic surgery with complaints of swelling over right side of lower jaw since last six years. Patient's parents noticed the swelling when patient grew older. She complained of deviation of mouth on mouth opening. She had no complaints of difficulty in deglutition, swelling elsewhere in body. No history of trauma, bleeding or pain. She had history of intermittent onset oral swelling with redness in the swelling.

On local examination there was a single swelling over right side of lower jaw with overlying smooth surface and diffuse margins bony hard in consistency. Mouth opening was 3.5cm adequate. There was deviation of mouth inferiorly and outwards on mouth opening towards right side. Swelling was present over right lower jaw's alveolar margin involving the gingivo-buccal sulcus extending up to midline. No evidence of any obvious growth or ulceration was seen. CT scan, coronal section (Fig. 1) showed expansion of right mandibular area with cystic areas and extensive dislocation of second and third right molars.



Fig. 1: CT scan, coronal section showed expansion of right mandibular area with cystic areas and extensive dislocation of second and third right molars.

Patient was posted for surgery and right segmental mandibulectomy with free fibula graft reconstruction was done and specimen was sent to histopathology section. We received specimen of right hemi-mandibulectomy. A single oval lesion is seen over antero-lateral border of mandible measuring 5x4cm, externally capsulated and hard in consistency. On cut surface multiple cysts are seen, largest measuring 4cm in diameter.

Multiple sections through tumor showed features of cemento-ossifying fibroma composed of connective tissue stroma consisting of bony trabeculae lined by surface epithelium. Bony trabeculae were of varying sizes with entrapped osteocytes in lacunae. Numerous basophilic spherical structures of varying sizes resembling cementum (psammomatoid bodies) were also noted. (Fig. 2 & 3) Section also showed developing tooth surrounded by connective tissue stroma.

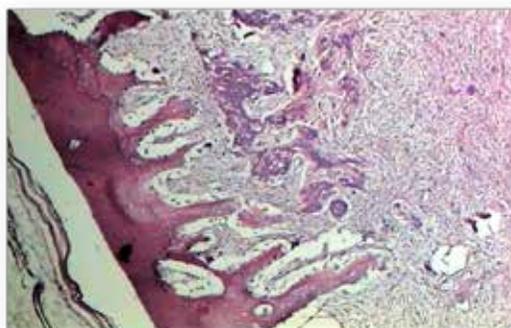


Fig.2: Microphotograph-Tumor composed of cellular fibroblastic tissue with basophilic cementum-like material and bony trabeculae containing entrapped osteocytes. (H&E, x 100)

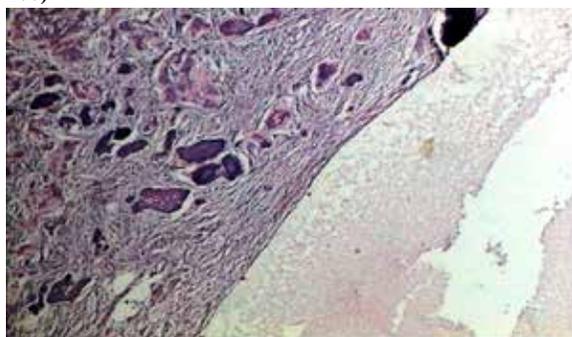


Fig.3: Microphotograph-Tumor composed of cellular fibroblastic tissue with cementum-like material. (H&E, x 400)

Discussion:

Maxillofacial fibro-osseous lesions comprise a group of face and jaw disorders characterized by the replacement of normal bone by a benign connective-tissue matrix with varying amounts of mineralized substances. A neoplastic etiology of ossifying fibroma is supported by examples of lesions that achieve a large size, exhibit aggressive behavior, and produce significant osseous destruction.² Additionally, recurrences, though rare, have been described in some studies of ossifying fibroma. Chromosomal translocations have been identified in a few cases of ossifying fibroma, however, the molecular mechanisms that underlie the development of this tumor remain unknown.²

The cemento-ossifying fibroma has caused considerable controversy because of confusion regarding terminology and the criteria for its diagnosis. The cemento-ossifying fibroma is odontogenic in origin, whereas ossifying fibroma is of bony origin. Cemento-ossifying fibroma is a fibro-osseous lesion that arises from the periodontal membrane.³ It contains multipotential cells that are capable of forming cementum, lamellar bone, and fibrous tissue.⁴

A close histogenetic relationship exists between the ossifying fibroma and the cementifying fibroma. It is based on the marked similarity between the two regarding predilection of age of occurrence, sex, race, location, roentgenographic appearance and clinical behavior, these two lesions represent the same basic neoplastic process. The only difference between the two being in the type of cell involved and its end product- bone in ossifying fibroma and cementum in COF. This has prompted many to use the term cemento-ossifying fibroma.^{2,4}

Cemento-ossifying fibromas are benign lesions of the jaw, which arise from the periodontal membrane. Trauma is commonly hypothesized to be the initiating feature of these lesions. This is commonly described in lesions in the jaw. Our patient did not have history of any such trauma at site of present mandibular swelling. Cemento-ossifying fibroma is a slow-growing lesion

composed of cellular fibroblastic tissue containing masses of cementum-like tissue. In addition, varying amounts of bony trabeculae are interspersed within the lesion, giving it its characteristic features.⁵

Cemento-ossifying fibromas are mostly reported in middle age group, between 30-40 years.⁶ This is not compatible with our patient's age of 11 years. So this case is called Juvenile COF. The conventional COF tumors are generally considered to be slow growing tumors. This is also proven by the six year long history of our patient. As Juvenile COF are aggressive than conventional COF, this case was thus an exception to this statement. Management of COF is by total excision. The prognosis is known to be fair, and recurrence after surgical removal seems to be unusual. Radiotherapy is contraindicated because of its radio-resistance and post-radiation complications.⁷⁻⁸

Conclusion:

Cemento-ossifying fibromas are common tumors in the jaw. We present an interesting case of juvenile cemento-ossifying tumor in a 11-year-old female child, which seems the youngest reported case through our literature search. This case may provide significant insight into the etio-pathogenesis, clinical, radiographic and microscopic features of COF. Also a proper surgical treatment is a must in order to eliminate the tumor completely, to avoid tumor recurrence and at the same time improve the patient's functional problems.

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