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Ossifying Fibromyxoid Tumor of Maxilla: A Case Report and Review of Literature

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Abstract: ***Objective: Ossifying fibromyxoid tumor is a rare soft tissue tumor of uncertain differentiation with varied presentation at different sites. They behave in a benign fashion, however a small percentage of tumors may locally recur or metastatize. This paper reports an unusual site of ossifying fibromyxoid tumor. In the maxillofascial region, this tumor is rarely reported. Case report: A 45years female presented with a right cheek swelling since 3 months which was clinically diagnosed as carcinoma of hard palate. Radiological examination showed expansile lytic lesion of alveolar process of right maxilla bulging into maxillary sinus. FNAC features were suspicious of odontogenic tumor. Biopsy tissue from this site showed features of Epulis. Patient underwent hemimaxillectomy. On histological examination, tumor showed oval to spindle shaped cells in abundant fibromyxoid stroma with focal areas of interstitial calcification. Tumor was covered by a thin rim of fibrocollagenous capsule along with focal areas of lamellar bone. Immunohistochemistry stain for Vimentin was positive. Final diagnosis of ossifying fibromyxoid tumor was made. Conclusion: Ossifying fibromyxoid tumors are diagnosed with a combination of clinical, radiological and pathological criteria. The rarity of the lesion may lead the pathologists to misdiagnose. These tumors should be completely excised for thorough management as some of them show local recurrence after excision.***

Keywords: ossifying fibromyxoid tumor, soft tissue, cheek swelling, maxilla, calcification

1. Introduction

Ossifying fibromyxoid tumor is a rare soft tissue tumor of uncertain differentiation which was first described by Enzinger et al in 1989. It is usually a slow growing, well defined small subcutaneous mass commonly arising in extremities and virtually always behaving in benign fashion [1].

Males are more frequently affected than females. Age of presentation is 14- 79 years with a median age of 50 years, almost exclusively affects adults with rare examples seen in young children [2-4]. 70% cases arise in extremities while diverse sites of presentation are seen including head and neck, trunk, mediastinum and retroperitoneum [5-9].

The proportion of bone tissue in these tumors varies. 80% cases are ossifying tumors whereas 20% cases are non ossifying variants [10]. Histogenesis remains controversial. Although several studies suggest that this is an unusual tumor, possibly of Schwann cell or cartilaginous origin [1,5,11,12], other authors have proposed a partial myofibroblastic origin [12-15]. Although the tumor is considered benign, local recurrence after excision is frequent [1], and cases with metastasis have also been reported [1, 13, 16-18].

2. Case Report

A 45 years female patient presented to department of ENT, KIMS, Hubballi with the complaint of toothache and loosening of right upper teeth since 4 months, swelling in the right cheek since 3 months, difficulty in chewing food since 1month . Swelling was insidious in onset, rapidly increased in size and was associated with pain.

Clinical examination: An ulceroproliferative growth measuring 6 x 5 cm, firm to hard in consistency was involving right side of hard palate extending upto gingivolabial sulcus, involving gums and displacing the second molar tooth. Right side level I b lymph node was enlarged measuring 1x1cm. Clinically it was diagnosed as malignancy of hard palate, $T_3N_1M_X$, stage III.

Radiological examination: Contrast enhanced CT scan revealed an expansile lytic lesion of right maxilla involving the alveolar process of second molar tooth, bulging into maxillary sinus. Inhomogenously enhancing soft tissue mass in the inferior portion of maxilla, beneath the hard palate, extending into oral cavity. However there was no evidence of erosin of hard palate (Figure 1).

FNAC features were suspicious of odontogenic tumor. Biopsy from the growth showed features of Epulis. Patient underwent right side hemimaxillectomy surgery.

Gross examination: Specimen consisted of right hemimaxilla with attached tumor measuring 7x4x4 cm. Growth was 3cm from anterior end, grey white to grey tan in color, firm in consistency, measuring 4x4x3 cm, involving floor of maxillary sinus and roof of hard palate and
a separately sent tumor mass from maxillary sinus measured 4x3x1cm (Figure 2). Cut surface showed solid grey white to grey tan areas and focal gelatinous areas .

Microscopy: Multiple histopathological sections showed a well demarcated lesion comprised of oval to spindle shaped tumor cells in an abundant fibromyxoid stroma. Tumor was covered by a thin fibrocollagenous capsule along with focal

areas of lamellar bone (Figure 3). Stroma showed areas of interstitial calcification (Figure 4).

Immunohistochemistry: Tumor cells were positive for Vimentin (Figure 5). Final diagnosis of ossifying fibromyxoid tumor of maxilla was given.

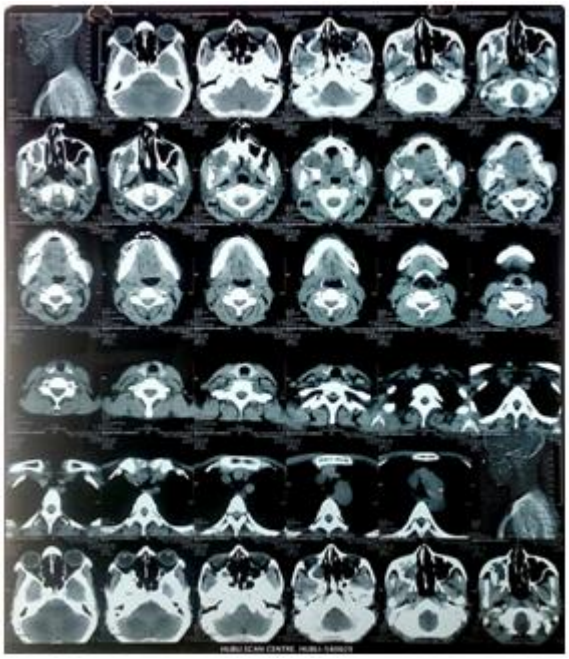
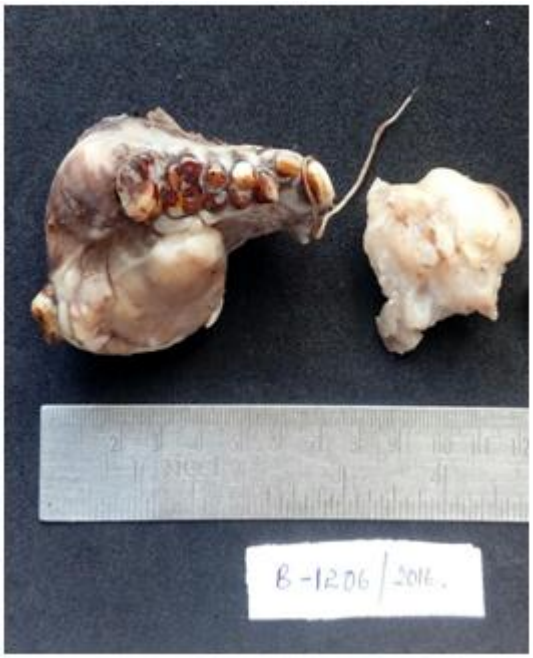


Figure 1



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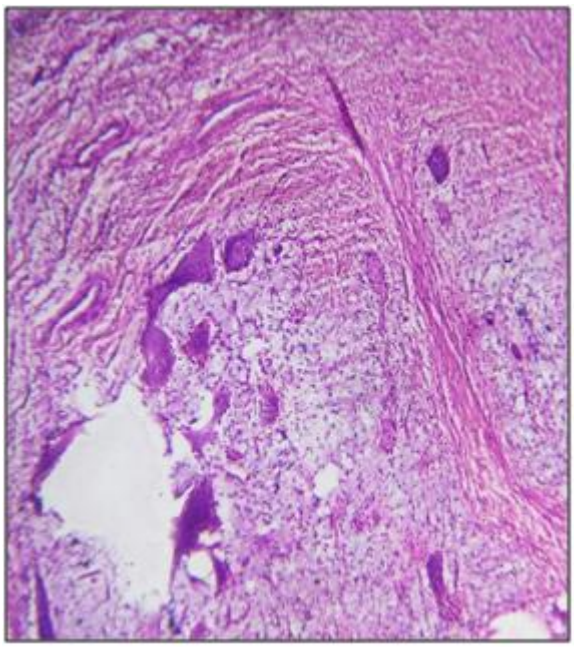

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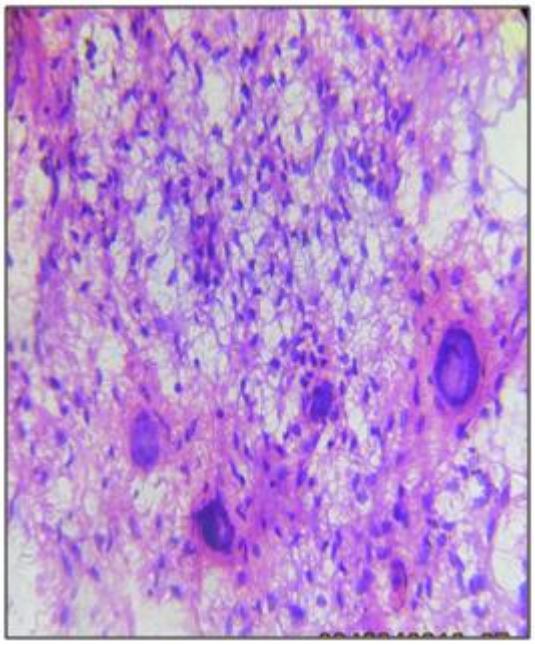

Figure 4

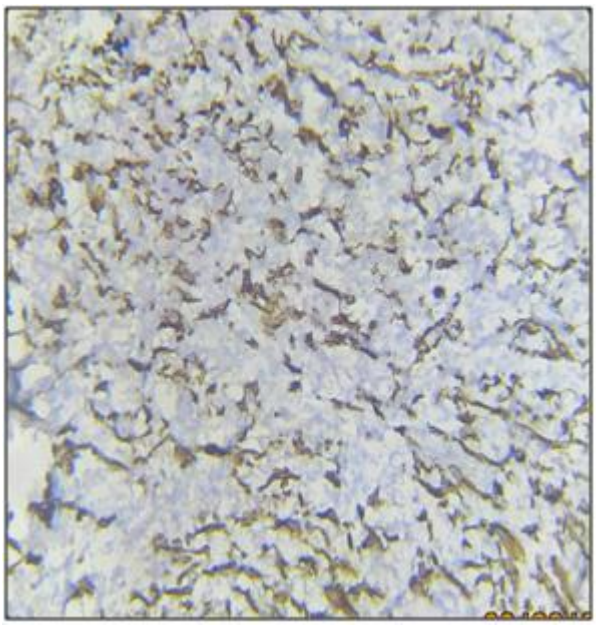

Figure 5

Figure 1: Contrast enhanced CT scan showing an expansile lytic lesion of right maxilla involving the alveolar process of second molar tooth, bulging into maxillary sinus and extending into oral cavity, beneath the hard palate.

Figure 2: Hemimaxillectomy specimen measuring 7x4x4 cm with tumor involving roof of hard palate and floor of maxillary sinus and separately sent tumor in the maxillary sinus measuring 4x3x1 cm.

Figure 3: Microscopy shows tumor composed of oval to spindle shaped cells in abundant fibromyxoid stroma, covered by a thin fibrocollagenous capsule along with focal areas of lamellar bone (H&E 10x).

Figure 4: Microscopy showing interstitial areas of calcification (H&E 40x).

Figure 5: Immunohistochemicaly tumor cells are positive for Vimentin (40x).

3. Discussion

Enzinger et al described 59 cases of ossifying fibromyxoid tumors (OFMT) that had not been previously reported. A wide age range (from 14 to 79 years) of patients were affected and there was a clear male predominance. Some patients had antecedent trauma to the tumor site [1].

Radiologicaly, X-Ray and computed tomography may reveal an incomplete shell of calcification surrounding a nodular mass with foci of ossification inside [17,18]. On magnetic resonance images,
the mass is mostly isodense with muscle in T1-weighted images and isodense with gray matter in T2-weighted images [9]. Tc-99M bone scan reveals increased uptake due to intratumoral mature bone formation [19]. OFMT should be considered as a possibility in FNA cytology of myxoid soft tissue tumors, especially tumors with radiologic evidence of ossification [17].

Grossly, they are well-circumscribed, spherical, lobulated or multinodular masses covered by thick fibrous pseudocapsule measuring 3-5 cm arising from subcutaneous tissue or muscle tissue [1,2]. Cut surface shows grey tan areas and has gritty texture. They occur chiefly in the upper extremities (34%) and lower extremities (34%) and less frequently in the trunk (19%). Head and neck involvement occurs in around 13% of cases [1]. In the maxillofascial region, OFMT has been reported in tongue, mandibular region, soft palate, hard palate, buccal mucosa, mandible, and vestibules. It was cranially reported in ethmoid sinus in a newborn as well. Malignant transformation has been reported in the tongue. Other sites, such as the sinonasal tract [9,4], the zygomatic/parotid region [5], and the retroperitoneum [20] have been documented.

Microscopically, the tumor is located in subcutaneous tissue, some are attached to tendons, fascia or involve underlying skeletal muscle. Tumor is composed of uniform round, ovoid or spindle shaped cells with vesicular nuclei and eosinophilic cytoplasm. Typically with a high cellularity, uniform round, oval or spindle shaped tumor cells arranged in cords, sheets or nest-like patter in a variably myxoid and collagenous stroma [1]. Some are predominantly myxoid and composed of alcian blue positive, hyaluronidase sensitive acid mucopolysaccharides, occasionally forming microcysts [21] and frequently shows transition towards hyaline fibrosis and metaplastic bone formation [2].

An additional typical feature is the presence of an incomplete shell of mature bone in the capsular region of the tumor. 80% of cases have incomplete shell of lamellar bone at periphery of nodules, either within or immediately underneath a dense fibrous pseudocapsule and some extending into substance of tumor. Upto 20% cases are non-ossifying variants of OFMT, the term used to describe tumors that lack a discernable shell of lamellar bone [5,14].

Mitotic figures are usually infrequent [2]. Ossifying fibromyxoid tumors are classifiesd as typical and atypical tumors [13]. In typical OFMT cases, recurrence and metastasis rates are 17% and 5% respectively, making it a lesion of intermediate malignancy [2]. Rare ossifying variants have areas of hypercelllarity, increased mitotic figures or both , others show extensive osteoid deposition in the central portion of tumor. These cases are reffered to as atypical or malignant variants. It is estimated that 3%-5% of all OFMT are atypical variants [22]. Necrosis and vascular invasion are rare [2, 17]. Some unusual histopathologic features that may make diagnosis of this tumor difficult include satellite micronodules, mucinous microcysts, absence of myxoid areas, crush artifact, multiple microcalcifications, epidermoid cysts, atypical chondroid differentiation with binucleate lacunar cells, and pericytic growth patterns [21].

Immunohistochemically, a significant number of tumors show variable staining. Tumors are positive for vimentin, some tumor cells display immunoreactivity for S-100 protein (70% of cases)[2]. whereas others are positive for smooth muscle actin, desmin, glial fibrillary acidic protein (GFAP), Leu-7 and neuron specific enolase (NSE) [23], with a lesser degree of reactivity for type II and IV collagen, but they do not express cytokeratins [8].

To conclude, Ossifying fibromyxoid tumors are rare tumors usually associated with benign behavior . They are diagnosed with a combination of clinical, radiological and pathological criteria. The rarity of the lesion may lead the pathologists to misdiagnose. These tumors should be completely excised for thorough management as some of them show local recurrence after
excision.

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