OSSIFYING FIBROMYXOID TUMOR OF ORAL CAVITY

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ABSTRACT
Ossifying fibromyxoid tumor is a rare tumor of mesenchymal origin with varied presentation at different sites including head and neck. Clinically these are slow growing lesions and patients have a variable age at presentation. A 14 years old girl presented with a slowly enlarging gingival swelling, which on radiological examination showed increased rarefaction in the mandible with a provisional diagnosis of an inflammatory lesion. Microscopically, the tumor had spindle to oval shaped cells in a fibromyxoid background with a peripheral shell of lamellar bone. Histological diagnosis of ossifying fibromyxoid tumor was made after immunohistochemical stains for vimentin and S-100 protein. Recurrence, metastasis and histologically increased mitotic count are indicative of atypical or malignant ossifying fibromyxoid tumors.

Key words: Ossifying fibromyxoid tumor. Oral cavity. Gingival swelling.

INTRODUCTION
Ossifying fibromyxoid tumor (OFT) is a rare and recently described mesenchymal neoplasm by Enzinger et al.1 They present as slow growing lesions and are generally regarded as benign but cases with atypical morphology showing recurrence and metastasis have also been reported.2 The presentation in oral cavity is extremely rare and only a few cases have been mentioned in the head and neck region.3 The reported case is the first of its kind seen at our institute.

CASE REPORT
A 14 years old girl presented with 3 months history of a nodular swelling between the buccal and gingival mucosa in the left anterior canine region, which was gradually increasing in size. The swelling was 4 cm in diameter and had red overlying mucosa. A provisional clinical diagnosis of epulis was made.

Radiologically, a left anterior oblique view of the mandible showed an area of rarefaction in the anterior mandibular region with a differential diagnosis of osteomyelitis. Excisional biopsy was done and the tumor was subsequently submitted for histopathological examination.

Grossly, the tumor was nodular with grayish white cut surface and a rubbery texture measuring 3.5cm in greatest diameter. Microscopically, there were ovoid to spindle shaped cells with clear lacunae around the cells. The cells were arranged in cords and nests with a scanty cytoplasm in a fibromyxoid background (Figure 1). The nuclei had inconspicuous nucleoli. The mitotic count was low with less than 2 mitosis per 10 high power field. There was perivascular hyalinization with a focus of metaplastic cartilage. Osteoid formation (Figure 2a) with fibrous connective tissue and metaplastic lamellar bone was found at the peripheral areas of the lesion (Figure 2b).

Immunohistochemistry showed strong vimentin and S-100 positivity by the tumor cells (Figure-3), whereas EMA and cytokeratins were negative confirming the diagnosis of OFT.

Figure 1: Photomicrographs showing stellate cells arranged in cords with a scanty cytoplasm in a fibromyxoid background. (Hematoxylin and eosin stain at x 200 and x 400 magnification).

Figure 2: Photomicrographs showing (a) stellate cells arranged in cords with a scanty cytoplasm in a fibromyxoid background and focal osteoid formation. (b) There is peripheral rim of fibrocollagenous tissue and fibrolamellar bone. (Hematoxylin and eosin stain at x 200 magnification).
Ossifying fibromyxoid tumor is a rare mesenchymal neoplasm of uncertain lineage presenting histologically as cords and trabeculae of ovoid cells in a fibromyxoid matrix surrounded by partial shell of lamellar bone. Males are affected more frequently than females. The age of presentation is 14-79 years with a median age of 50 years. About 70% cases arise in the extremities while diverse sites of presentation are seen including head and neck, trunk, mediastinum and retroperitoneum. Occasional cases have also been reported in oral cavity. In a case series reported about oral cavity lesions, there were 7 lesions which occurred in a subcutaneous site, while two lesions occurred intraorally beneath the gingival and palatal mucosa. The patients present clinically with a long-standing painless mass attached to the underlying tissue. Radiologically, there is well circumscribed lobulated mass with irregular calcifications and surrounded by an incomplete shell of lamellar bone. Erosion of the underlying bone with periosteal reaction mimicking osteomyelitis may also be seen in this case.

On gross examination of the resected specimen, the tumors vary in size from 3 to 5 cm with a median size of 4 cm. However, cases as large as 17 cm have also been reported. The tumors are circumscribed with a fibrous pseudocapsule having an incomplete shell of bone at the periphery. Microscopically, there are lobules of uniform, round to fusiform shaped cells arranged in nests and cords set in a fibromyxoid stroma. The neoplastic cells have scanty eosinophilic cytoplasm with round to oval nuclei having inconspicuous nucleoli. Incomplete shell of hypocellular metaplastic bone is seen at the peripheral edges of the tumor. Calcifications and/or nodules of metaplastic cartilage are also occasionally identified. A histological differential diagnosis with benign lesions as myxoma and fibroma can be made. A histological diagnosis of OFT is more favourable than the above-mentioned lesions, which are more common at this site clinically, when a cellular tumor with fibromyxoid background, metaplastic bone and a shell of lamellar bone at the periphery is seen.

DISCUSSION

Frequently, there is random deposition of osteoid by the neoplastic cells within the centre of the lesions. Atypical and malignant ossifying fibromyxoid tumor are hypercellular with increased mitotic figures. In the absence of metastasis, the lesion is regarded as atypical variant while in the presence of metastasis, it is classified as malignant.

Immunohistochemical examination of the tumor shows positivity for vimentin, while 70% cases are positive for S-100 protein. Variable expression for desmin, glial fibrillary acid protein and smooth muscle actin is seen which further augments the uncertain lineage of the tumor. Based on immunohistochemistry and ultrastructural findings, there is preponderance of evidence to suggest a Schwann cell or cartilagenous origin.

Recurrence is noted in 27% cases. The clinical features and histology of the recurrent tumors is not any different from the non-recurrent lesions. Increase in cellularity and higher mitosis have been seen in some of the recurrent tumors which are regarded as atypical or malignant tumors.

REFERENCES