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Case Report

Ossifying fibromyxoid tumour in mandibular region: A case report

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ABSTRACT

Ossifying fibromyxoid tumour is a distinctive mesenchymal neoplasm of uncertain differentiation which has a potential for local recurrence and metastasis.
This case report represents the ossifying fibromyxoid tumour in mandible which is an uncommon site. OFMT needs to have early follow-up to detect local recurrence and metastasis.
OMFT presents as a fairly circumscribed cellular neoplasm arranged in lobules with incomplete peripheral shell of woven/lamellar bone. Appropiate diagnosis of ossifying fibromyxoid tumour is important to recognize because, even though it has a low grade morphology, it can have aggressive behaviour with potential for recurrence and metastasis. Hence, the patient should be kept under strict follow-up.

1. Introduction

Ossifying fibromyxoid tumors (OFMTs) are rare, soft tissue neoplasms of uncertain differentiation. Most commonly occur in extremities. Uncommon sites are head and neck, mandible and gingival regions.

They were first described in 1989 by Enzinger et al. in an article detailing 59 cases of a histologically unique neoplasm.[1] Histologically, they are characterized by lobulated chords of bland, round cells organized in a fibromyxoid hyaline matrix with a peripheral shell of woven bone. Although OFMTs can present in all age groups, they usually arise in adults with a median age of 50, with a slight predilection towards males (1.5:1).[2]

Clinically, an OFMT tends to present as a small, painless mass that persists over years most commonly arising on the extremities. Most OFMTs are benign and can be treated with excision; however, local recurrence has been seen in 17% of cases. Furthermore, malignant OFMTs have been identified in 5% of cases and have a metastatic potential of 60%.[2] In this report, we present a case of a atypical variant OFMT, localized in the mandibular region.

2. Case Presentation

A 59- year-old female presented with a slow growing painless swelling in the right mandibular region of 4 years duration. She also gives a past H/O Invasive breast carcinoma NST for which chemotherapy was given. CECT-mandible showed a lesion with cortical breakdown and bone destruction with infiltration into surrounding structures measuring 3.6x2.4x2.2cm. Curettage was done, and we received grey brown soft to firm tissue pieces aggregate measuring 3x3x2cm. Cut section showed grey white with myxoid, gritty and glistening areas. No bony tissue identified.

Microscopy revealed a fairly circumscribed cellular neoplasm arranged in lobules with incomplete peripheral shell of woven/lamellar bone. Cells were arranged in cords, nests and sheets embedded in fibromyxoid stroma. (Figure 1) Cells were round to oval / spindle cells with scant to moderate cytoplasm and bland round to oval nuclei.

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(Figures 2 and 3). Mitosis <2/50 hpf. Satellite nodules were present.

Immunohistochemical stains were done which showed that the tumour cells were strongly positive for CD 10, S100, & Vimentin, GFAP showed weak focal positivity. p63, SMA, ER, PR, HER2neu, GATA-3 were all negative. Ki67 Proliferation index was < 1%.

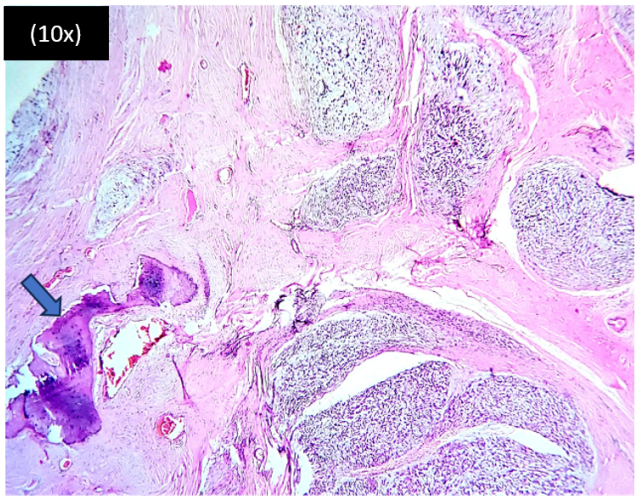

Figure 1: 10 x view shows fairly circumscribed neoplasm and cells arranged lobules. Arrow in this figure indicates woven bone in the peripheral shell

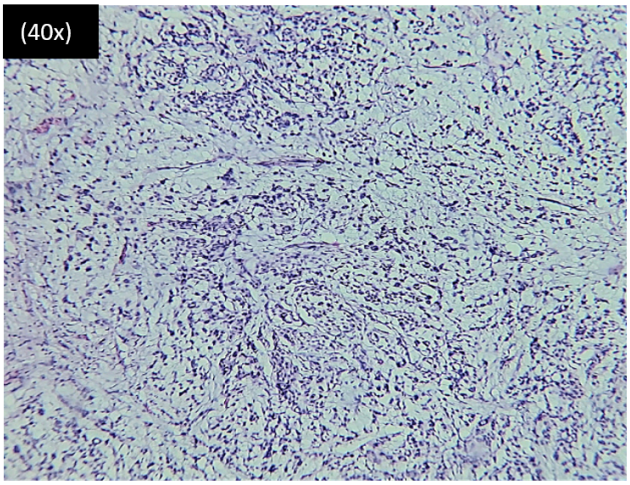

Figure 2: Low magnification image shows round to oval or spindle cells embedded in fibro myxoid stroma

2.1. Diagnosis

Histopathology and immunohistochemical findings favoured the diagnosis Ossifying fibromyxoid tumour with atypical presentation.

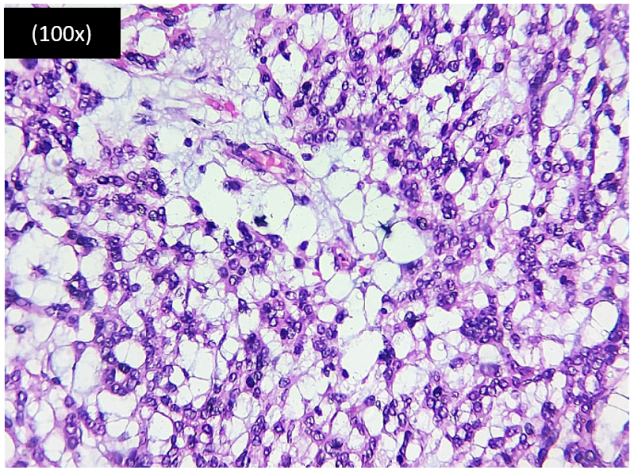

Figure 3: Higher magnification image shows round to oval cells with moderate eosinophilic cytoplasm with high N/C ratio with round to oval nuclei with few cells shows conspicuous nucleoli

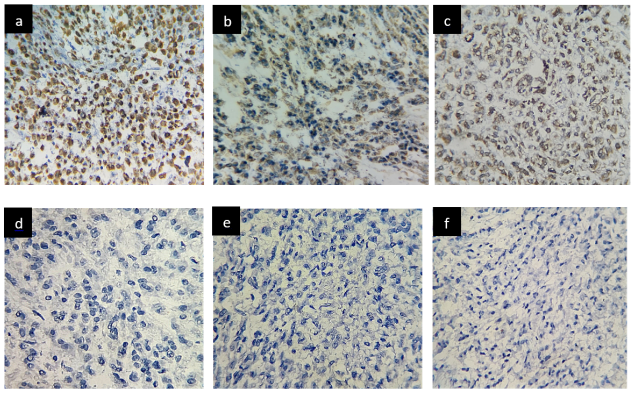

Figure 4: Immunohistochemical stains: **a)**: S100; **b)**: Vimentin; **c)**: CD10. **d):** p63; **e)**: SMA; and low proliferation index; **f)**: Ki67

3. Discussion

OFMT is a rare mesenchymal neoplasm of uncertain histogenesis demonstrating intermediate malignant potential.[3] It arises more commonly in the extremities and trunk and only 10 to15% of the cases reported in head and neck region.[4] OFMT is usually occurs as well circumscribed mass arising in subcutaneous or deep soft tissue, characterised by proliferation of small epithelioid cells with round vesicular nuclei arranged in cords or nests in a myxoid and hyaline stroma with an incomplete shell of lamellar bone in a fibrous capsule.

Folpe and Weiss[5] in 2003, proposed a risk stratification system for OFMT in which cases with a high nuclear grade, high cellularity and mitotic activity >2 mitotic figures/50 HPF should be classified as “malignant OFMT”.

Cases with atypical features, but not with all the criteria listed above, may be classified as “atypical OFMT” and considering the others as “typical OFMT”.[6]

OFMT was considered to be of cartilaginous or neural origin (supported by S100 protein expression).[7] The immunohistochemical profile of intraoral OFMT is similar to cases arising in extraoral sites[3] with frequent positivity for vimentin, S100 and occasionally positive for CD10, negative for SMA and p63.

The most common genetic rearrangement found in OFMT is that of inactivation of INI-1 gene and deregulation of PHF1 gene in 6p2.[8] INI-1 is a tumour suppressor gene located on chromosome 22q11.2 that encodes a protein expressed essentially in all nucleated cells.[9] The mosaic loss inactivation of this tumour suppressor gene in OFMT, including typical and malignant forms, has also been suggested as a role in tumorigenesis. Because of which it is placed in the category of soft tissue tumour with recurrent translocation lacking distinct histogenesis.[7]

OFMTs have been characterized as tumours with intermediate clinical behaviour, although the majority behave in a benign manner. But atypical tumours present with local recurrence or rarely metastasizing to distant locations. OFMT is typically treated by local excision and post-surgical surveillance.

In this case report tumour was excised and patient kept under strict follow-up for any recurrence or metastasis.

4. Conclusion

OFMT is important to recognize because even though has a low-grade morphology, it can have aggressive behaviour with potential for recurrence and metastasis renders the differential diagnosis a true challenge. Hence, the patient should be kept under strict follow-up.

5. Source of Funding

None.

6. Conflict of Interest

None.

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