A Rare Entity of Undifferentiated Pleomorphic Sarcoma of the Mandible - A Case Report

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Abstract

The Rationale: Undifferentiated pleomorphic sarcoma originally known as malignant fibrous histiocytoma was declassified by the World Health Organization in 2002 as a formal diagnostic entity and renamed as an undifferentiated pleomorphic sarcoma. It accounts for <1% of malignant tumours of the long bone. **Patient Concerns:** A 33-year-old male patient reported with swelling and pain in the lower left posterior jaw region for 3 months with a history of fall on the floor 3 months back. **Diagnosis:** On examination, diffuse solitary swelling was present on the left lower third of the face and was diagnosed radiographically and histopathologically as undifferentiated pleomorphic sarcoma of the mandible. **Treatment and Outcomes:** Selective neck dissection, followed by reconstruction with fibula osteomyocutaneous flap and then referred for adjuvant radiotherapy. **Take-away Lessons:** Vimentin staining plays a substantial role in the diagnosis of undifferentiated pleomorphic sarcoma. A long-term follow-up after treatment is required to increase the chances of disease-free survival for the patients.

Keywords: Fibrous histiocytoma, histiocytes, multinucleated giant cells, pleomorphic sarcoma

Introduction

Undifferentiated pleomorphic sarcoma earlier known as malignant fibrous histiocytoma arises predominantly in soft tissues with a peak incidence from fifth to seventh decades of life.[1] Head-and-neck malignant fibrous histiocytoma usually manifests as smaller in size and lower in grade, hence for this reason head-and-neck malignant fibrous histiocytoma has a more favorable survival prognosis in comparison with trunk and other extremity regions.[2]

The nasal cavity and the paranasal sinuses are more common sites in the head-and-neck region.[3] The tumour occurs only 3%–10% of cases in the maxillofacial region and only 3% of the cases in the mandible.[4] Aggressive surgical resection is the principal treatment. Local recurrence noted is about 13%–42% and distant metastasis could be about 31%–35%.[5] Genetic background, environmental factors such as trauma, radiotherapy, and malignant transformation from benign lesions can be involved in the etiology of this malignant tumour.[6] A history of antecedent trauma is seen in about 20% of the cases suggesting that some of these tumours may represent as an initial proliferative response to trauma.[4]

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We present a case of undifferentiated pleomorphic sarcoma in a 33-year-old patient with a history of trauma to the mandible. This reinforces the possibility of tumour being an initial proliferative response to trauma.

Case Report

A 33-year-old man reported to the department with a chief complaint of pain and swelling in the lower left jaw region for 3 months and had no significant medical history. The patient gave a history of slip and fall on the floor 3 months back after which swelling and pain developed. On extraoral examination, mild asymmetry was noted with no trismus or lip paresthesia.

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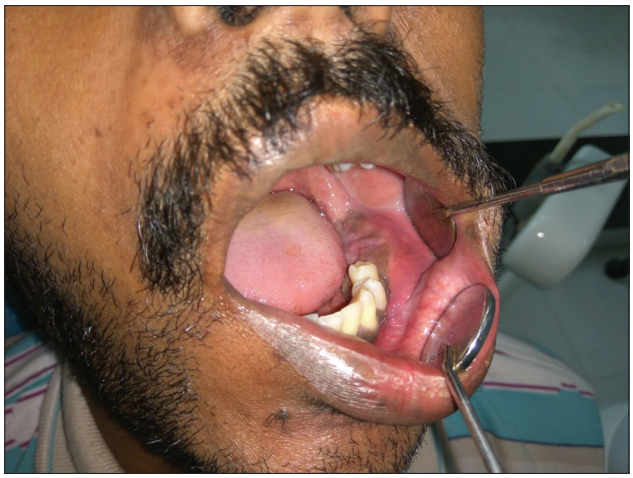

Figure 1: Intraoral photograph showing lesion *in-situ*

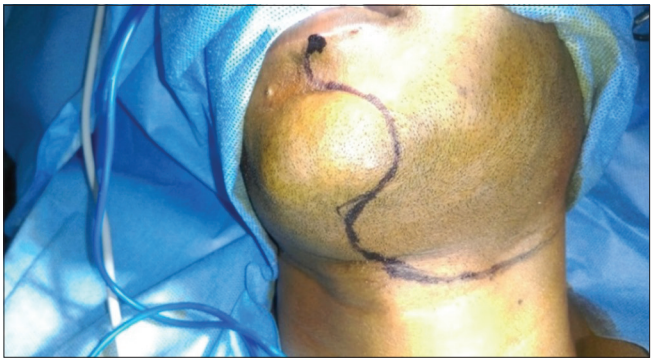

Figure 3: Midline split incision given

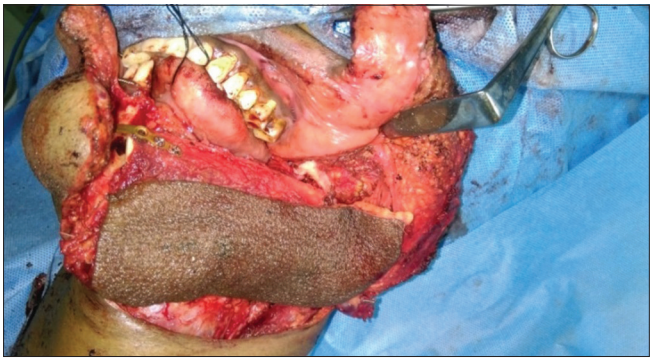

Figure 5: Reconstruction using fibula flap

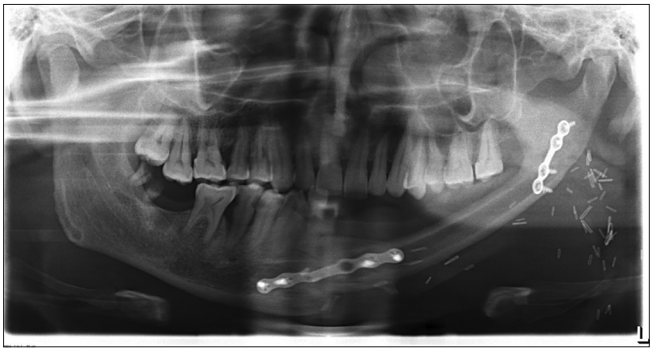

Figure 7: Postoperative orthopantomogram

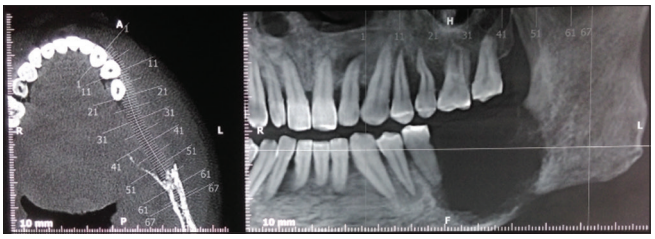

Figure 2: Cone-beam computed tomography revealed radiolucent lesion with ill-defined border

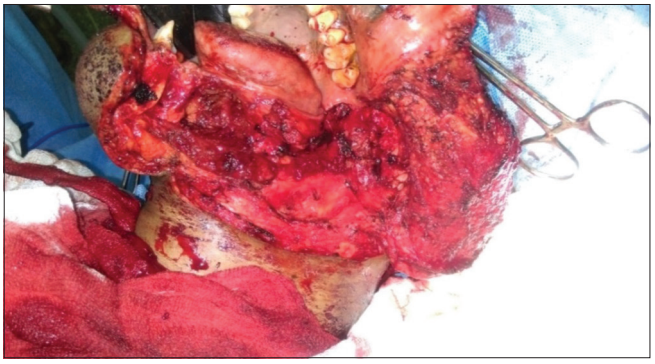

Figure 4: Segmental mandibulectomy done

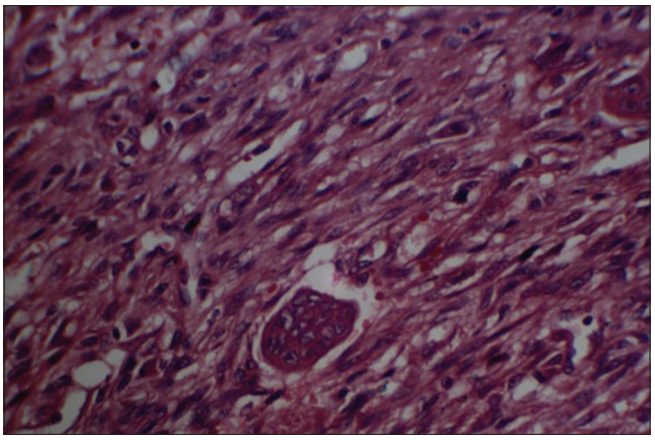

Figure 6: Histopathology of the patient showing high cellular mesenchymal component that consists of numerous monomorphic spindle-shaped fibroblast and histiocyte like cells in varying proportions, multinucleated giant cells seen interspersed in the connective tissue stroma, and the tumour cells are invading the osseous tissue at the periphery (×40)

Diffuse solitary swelling was seen in the left lower third of the face measuring 5 cm × 5 cm. Anteroposteriorly swelling extended from 1 cm from the corner of the mouth till the angle region of the mandible and superoinferiorly extending 2 cm below the ala tragal line extending up to the inferior border of the mandible.

Two submandibular lymph nodes were palpable measuring around 0.5 cm × 0.5 cm oval in shape, fixed to the underlying tissue, and tender on palpation.

On intraoral examination diffuse swelling measuring around 4 cm × 5 cm was seen extending from mesial aspect of mandibular left first premolar and extending up to the retromolar region.

Mucosa over swelling was normal and mildly tender on palpation. Vestibule was obliterated. The patient gave a history of spontaneous exfoliation of teeth in that region [Figure 1].

Cone-beam computed tomography revealed radiolucent lesion with ill-defined border extending from mandibular left first premolar up to the angle region [Figure 2]. Chest X-ray did not reveal any metastatic deposits.

Incisional biopsy was performed under local anaesthesia. Histopathology suggested malignant fibrous histiocytoma. Immunohistochemical stains showed strong reactivity to vimentin. Ultrasound of the neck showed large soft-tissue mass on the left side of the mandible, showing heterogeneous texture with increased flow measuring around 3.5 cm × 3 cm. Two or three enlarged lymph nodes were seen in the left submandibular region measuring 0.8–1.2 cm.

Segmental mandibulectomy with supraomohyoid neck dissection was planned with reconstruction with fibula osteomyocutaneous flap. Midline split incision was given extending till the left neck crease [Figure 3]. Segmental mandibulectomy was done from midline extending up to ramus. Left level IIa and III neck nodes were cleared [Figure 4]. Fibula flap was harvested and used to reconstruct the left side of the mandible with reconstruction plates and microvascular anastomosis was carried out [Figure 5].

Histological examination [Figure 6] of the primary tumour revealed highly cellular mesenchymal component that consisted of numerous monomorphic spindle-shaped fibroblast and histiocyte-like cells in varying proportions. Multinucleated giant cell was seen interspersed in the connective tissue stroma. At the periphery, the tumour cells were invading the osseous tissue. Postoperative orthopantomogram was taken [Figure 7]. The patient was referred for adjuvant radiotherapy.

Discussion

The World Health Organization in 2002 declassified malignant fibrous histiocytoma as a formal diagnostic entity and renamed it as an undifferentiated pleomorphic sarcoma. This new terminology suggests that malignant fibrous histiocytoma represents a final common pathway in tumours that undergo progression toward undifferentiation.[4] Pathological analysis of primary tumour tissue demonstrates the presence of myxoid, fibrous, and osseous tissue, the isolated cells had a spindle cell-like morphology.[7] Various studies suggest that malignant fibrous histiocytoma is a sarcoma which shows features of both fibroblastic and histiocytic differentiation.[8]

Oral soft tissue is more commonly involved than the oral hard tissue. From the clinical aspect, malignant fibrous histiocytoma is mostly seen as a painful nodular mass with trismus, but however, in some cases, it can also be painless.[9]

Immunohistochemistry plays a substantial role in the diagnosis of UPS. Vimentin positivity indicates that the tumour cells originate from mesenchymal cells. In the present case, tumour showed positive reaction to vimentin.[10]

There are five histologic subtypes (1) storiform/pleomorphic, (2) myxoid, (3) giant cell, (4) inflammatory, and (5) angiomatoid. The giant cell variant is considered to be aggressive and has poor prognosis.[4] High recurrence rate is seen due to aggressive infiltration of adjacent tissues or between muscle fibers.[6]

Majority of the undifferentiated pleomorphic sarcoma (60%–70%) in the literature has been shown to be of storiform-pleomorphic type. This type is essentially composed of a mixture of spindle cells admixed with polygonal or rounded cells, arranged in a storiform pattern. Myxoid type is the next most common type (10%–20%) with a better prognosis compared to the other types.[11]

Treatment consists of early and complete surgical removal using wide or radical resection with a minimum of 3 cm tumour-free margin.[1,6] Elective neck dissection should be considered for patients with lymph node metastasis, advanced-stage disease, or evidence of aggressive histopathological features.[4.6]

Based on the follow-up data from the reported Malignant Fibrous Histiocytoma (MFH) cases involving the mandible, it is seen that an early-stage diagnosis and aggressive treatment with radical surgery with or without adjunct postsurgical chemo/radiotherapy is the formulated treatment plan.[12]

Systemic chemotherapy is indicated for those tumours with a high risk of distant metastasis. Anticancer agents such as methotrexate, cisplatin, doxorubicin, cyclophosphamide, actinomycin-D, vincristine, and ifosfamide have been routinely used. Adjuvant radiotherapy is generally recommended for high-grade sarcomas, large tumours, close or positive surgical margins, and certain histologic variants.[4]

Conclusion

Undifferentiated pleomorphic sarcoma with its questionable origin, unusual occurrence in mandible, varying pathological features, multiple subtypes, aggressive nature, and distant metastasis, is a challenge for the surgeon. The diagnosis is a combination of clinical, radiographic, and histopathological features to formulate a customized treatment plan. This tumour presents as a response to proliferation after trauma and vimentin staining plays a substantial role in diagnosis. After the treatment, long-term follow-up is required to increase the chances of disease-free survival for the patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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