Case Report

A rare case of undifferentiated pleomorphic sarcoma affecting the mandible – A case report

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Abstract

Undifferentiated pleomorphic sarcoma (UPS) is a rare entity found in the oral cavity which involves the mandible in only 3% of all UPS bone lesions. The correlation of the clinical presentation of the neoplasm with the histopathological and immunohistochemistry findings is very important for appropriate diagnosis. Its incidence in membranous bone like mandible is rare. According to our literature search in the PubMed database, there are only four reported cases from India some of which does not have any immunohistochemistry findings to confirm the diagnosis. We present a rare case of UPS of mandible which was clinically mimicking squamous cell carcinoma. Upon immunohistochemical analysis, we found vimentin, KI67, CD68 positive and desmin, Melan A, LCA, S100 and myogenin negative, which lead us to the diagnosis of UPS.

Keywords: Immunohistochemistry, mandible, undifferentiated pleomorphic sarcoma (UPS)

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Submitted: 20-Mar-2023, **Revised:** 13-Oct-2023, **Accepted:** 13-Oct-2023, **Published:** 15-Apr-2024

INTRODUCTION

High-grade soft-tissue sarcomas include undifferentiated pleomorphic sarcoma (UPS), which was previously named as malignant fibrous histiocytoma (MFH).[1] O' Brian and Stout originally identified MFH as a novel malignant tumour in 1964. It is one of the rare soft tissue sarcomas of adults encountered in the oral cavity.[2] UPSs represent about 10% of adult soft tissue sarcomas and are frequently diagnosed as an exception.[3] The prevalence of these tumours rises with advancing age, with the largest risk of occurring during the sixth decade of life, and white males are more likely to develop UPSs.[4] The nasopharyngeal tract, craniofacial bones, larynx and soft tissue structures of the neck are the most often affected areas in the craniofacial area.[5] Here, we report a case of UPS with an unusual presentation in the right mandibular buccal mucosa.

Access this article online

Quick Response Code:

Website: https://journals.lww.com/JPAT/

DOI: 10.4103/jomfp.jomfp_142_23

CASE REPORT

A 68-year-old male Hindu patient reported to a private clinic with the chief complaint of growth in the lower right back tooth region with cheek bite and difficulty in having food since 1 month. The growth was gradually increasing in size and was associated with pain. The pain was of gradual onset and vague aching in nature. It was not associated with any discharge. The medical history of the patient revealed that he was hypertensive and was under medication. His dental and family history were not relevant. No other anomalies

How to cite this article: Mahapatra M, Panda A, Kumar H, Bhuyan L. A rare case of undifferentiated pleomorphic sarcoma affecting the mandible – A case report. J Oral Maxillofac Pathol 2024;28:130-3.

were found during the routine physical examination. Upon extraoral examinations, the face was bilaterally symmetrical, temporomandibular joint (TMJ) was normal and the lymph nodes were not palpable. Intraoral examination on inspection revealed a nonhealing ulceroproliferative, sessile growth of size 5 cm × 3.5 cm in the mandibular right buccal mucosa extending from 46 to the retromolar region [Figure 1]. On palpation, the growth was tender, firm in consistency and sessile. It was nonreducible, noncompressible and nonfluctuant in nature. There was no purulent discharge observed. The growth was extending into the labial and lingual vestibular mucosa adjoining the alveolar mucosa. A provisional diagnosis of malignancy alveolobuccal complex? was given with the differential diagnosis of squamous cell carcinoma, tuberculosis, rhabdomyosarcoma and fibrosarcoma. Routine blood and biochemical investigations were advised, which were found to be within the normal limits. Radiographical findings revealed generalised horizontal loss of bone with respect to 45 46 regions [Figure 2]. Followed by which incisional biopsy was advised to the patient. On gross examination, we received multiple bits of soft tissue with the largest tissue measuring 3.5 cm × 2.2 cm and the smallest tissue measuring 0.5 cm × 0.4 cm in dimension, brownish in colour and firm in consistency. The haematoxylin and eosin stained section, in 40× view it showed a fibropurulent membrane lining the connective tissue suggestive of surface ulceration. 100× view showed some areas of dense connective tissue component, which was an admixture of Spindle-shaped and ovoid neoplastic cells which were arranged in a storiform pattern as well as in a fascicular pattern along with a few myxoid areas. In 400× view, neoplastic cells with features of malignancy such as cellular and nuclear pleomorphism, multiple nucleoli, altered nuclear cytoplasmic ratio were seen. Few aberrant mitotic figures were present [Figure 3]. The histopathological differential diagnoses were fibrosarcoma, pleomorphic sarcoma, desmoplastic melanoma, leiomyosarcoma and synovial sarcoma. Further immunohistochemistry was advised to the patient in order to rule out the fibrous histiocytic nature of the lesion and for an accurate diagnosis. Upon immunohistochemistry (IHC), the tissue was strongly positive for vimentin, CD68 and KI67 confirming the presence of histiocytes and fibroblast like cells in the connective tissue stroma [Figure 4]. It showed CK AE1, AE3, CK 5 and CK6 positivity only in the epithelium and negativity in the connective tissue region. It also showed negativity for dsmin, Melan A, LCA, S100 and myogenin, which helped us to differentiate it from myogenic, melanotic, haematopoietic, neurogenic and muscular tumours. Correlating the clinical findings to the histopathology findings and immunohistochemistry findings we reached onto the diagnosis of UPS. A localized

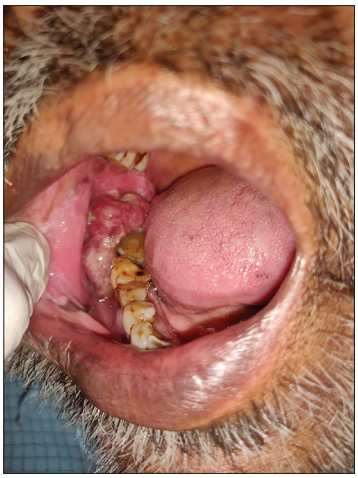

Figure 1: Ulceroproliferative growth extending from 46 to the retromolar region

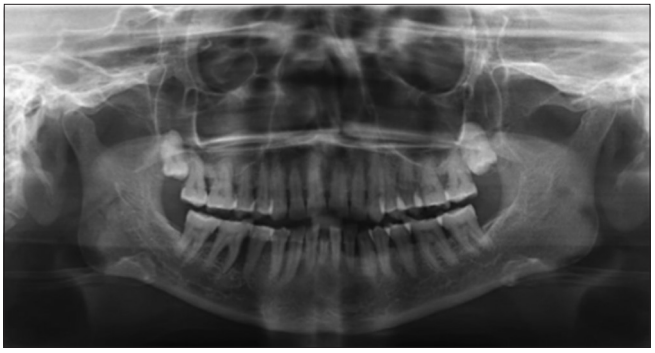

Figure 2: Orthopantomogram showing generalised horizontal loss of bone in 45 46 regions

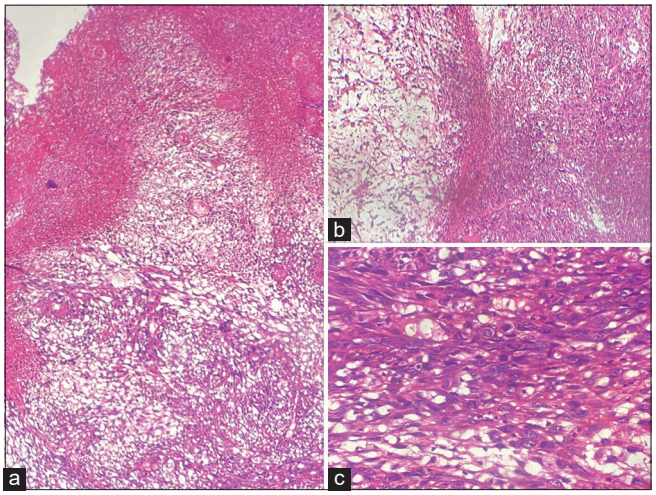

Figure 3: Photomicrograph of haematoxylin and eosin stained section showing (a) fibrinopurulent membrane lining the connective tissue (40× magnification), (b) spindle shaped neoplastic sells arranged in fascicular and storiform pattern with adjoining myxoid arears (100x magnification) and (c) neoplastic cells showing features of malignancy (400×)

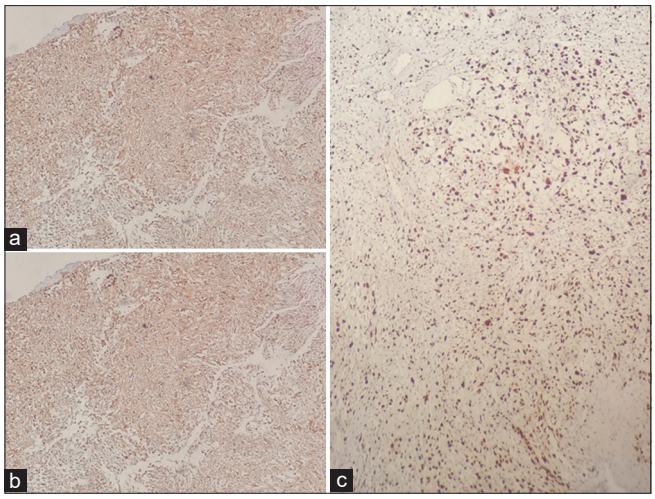

Figure 4: Photomicrograph showing strong positive expression of CD68 (a), vimentin (b), and KI67 (c) in immunohistochemistry under 100× magnification

excision with 1 cm tumour-free boundaries was carried out under general anaesthesia. The patient had been followed up at regular intervals and there was no evidence of recurrence in the past 3 months.

DISCUSSION

In 2002, the World Health Organization reclassified MFH as an UPS and discontinued it as a standardized diagnostic entity.[6] According to the new nomenclature, MFH is the last prevalent route in tumours that proceed to undifferentiation.[7] The most prevalent soft-tissue sarcoma in adulthood is UPS, which makes up 20%–30% of all soft-tissue sarcomas.[2] Pleomorphic sarcoma, also known as UPSUPS or MFH, can involve various tissues including soft tissue, muscles and in some cases bone. However, it is primarily considered a soft tissue sarcoma. When pleomorphic sarcoma affects bone, it is often referred to as a "malignant fibrous histiocytoma of bone". Bone involvement by pleomorphic sarcoma is relatively rare, and most cases of pleomorphic sarcoma occur in soft tissue. When it does occur in bone, it can lead to bone destruction, fractures and other complications.[8] In all UPS bone lesions, the mandible is only involved in 3% of cases. We conducted a literature search in the PubMed database for the documented cases of UPS of mandible using the following Mesh terms malignant fibrous, undifferentiated pleomorphic, histiocytoma, sarcoma and mandible. We found only four documented cases from India. listed in [Table 1] Other case reports were excluded as they were of different variants than our case, some had no full-text article and some belonged to various different countries other than India. According to the review of literature, we can say that the occurrence of UPS in the oral cavity specifically in the mandibular region is rare. Here, we have reported a nonhealing ulceroproliferative growth of size 5 × 3.5 cm in the mandibular right buccal mucosa extending from 46 to the retromolar region in a 68-year-old male who has a habit of paan chewing, since 10 years. UPS of the bone, initially identified by Feldman and Norman in 1972, can happen at any age; however, most instances affect individuals between the sixth and seventh decades of life which coincided with our literature review. We found that various authors reported the cases in different age groups like Sarode *et al*.[9] reported a case of a 32-year-old male, Lambade *et al*.[7] reported a case of a 14-year-old male, Datarkar *et al*.[10] reported a case of a 16-year-old male and Archana *et al*. reported a case of a 33-year-old male. The neoplasm has a male predilection.[11] According to the review of literature, the tumour is most commonly found in the posterior mandibular region. In our case, the lesion was slow growing in nature and had a history of trauma from occlusion; according to the literature review, only 20% of cases have a history of trauma.

UPS is typically observed clinically as a painful lumpy growth with muscle spasms; however, it can occasionally be asymptomatic.[12] Histopathologically, there are different five subtypes: (1) storiform/pleomorphic, (2) myxoid, (3) giant cell, (4) inflammatory and (5) angiomatoid.[7] In our case, storiform arrangement of malignant spindle cells are seen with few myxoid areas.

Most of the UPSs (60%–70%) have been reported to be of the storiform/pleomorphic type in the review of literature. The next most frequent form is the myxoid type, which has a better prognosis than the other varieties (10%–20%).[13] In the diagnosis of UPS, immunohistochemistry plays a crucial role. In our case, cytokeratin negativity and vimentin were positivity pointed towards mesenchymal origin of the tumour. The positivity of CD68 confirmed the histiocytic nature of the tumour cells. Upon KI67 staining, the proliferative index showed to be 90%.

Early and thorough surgical excision employing broad or radical resection and at least 3 cm tumour-free margin constitutes the course of treatment.[14] The use of radiotherapy and chemotherapy is advised for tumours that have a high risk of metastasising to distant organs.

According to the reported cases of UPS in the literature, early diagnosis with radical resection with or without radiotherapy and chemotherapy is the choice of treatment. Regular periodic follow-ups are absolutely necessary as it has a high chance of recurrence.

The present case mimicked an oral squamous cell carcinoma clinically. Yet the histopathological presentation

Table 1: Review of literature

Author	Age/sex	Location	Histopathology	IHC	Treatment/follow up
Sarode *et al*. 2018[9]	32 yrs/ male	Left mandibular buccal mucosa, 3rd molar to retromolar pad region	Spindle-shaped cells arranged in storiform pattern	Positive for Vimentin, CD68 Negative for S100, EMA, CK	Hemimadibulectomy and disarticulation of chondyle. Recovered after 5 yrs of surgery
Lambade *et al*. 2013[7]	14 yrs/ male	Left mandibular gingivobuccal sulcus	Spindle-shaped cells arranged in storiform pattern	NA	Chemotherapy and radiotherapy. Recurrence after 5–6 months and death
Datarkar *et al*. 2009[10]	16 yrs/ male	Left mandibular molar and ramus	Fibroblast like cell, fibrosis, and hyalinization	NA	Hemimandibulectomy Recurrence after 36 months
Archana *et al*. 2021[6]	33 yrs/ male	Left mandibular 1st premolar upto retromolar region	Spindle shaped fibroblast and histiocytes, multinucleated giant cells	NA	Segmental mandibulectomy with supraomohyoid neck dissection

diverted the diagnosis towards a spindle cell malignancy. Immunohistochemistry was empirical in reaching onto a final diagnosis.

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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