Journal of Craniomaxillofacial Research

Vol. 12, Issue. 2 Spring 2025

Oral Undifferentiated Pleomorphic Sarcoma: A Case Report

Saeedeh Khalesi [1], Mohammad Parsa Ghazi [2*], Bijan Movahedian Attar [3], Seyed Mohammad Razavi [3]

1. Dental Materials Research Center, Department of Oral and Maxillofacial Pathology, School of Dentistry, Isfahan University of Medical Sciences, Isfahan, Iran.

2. School of Dentistry, Isfahan University of Medical Sciences, Isfahan, Iran.

3. Dental Implant Research Center, Department of Oral and Maxillofacial Pathology, School of Dentistry, Isfahan University of Medical Sciences, Isfahan, Iran.

ARTICLE INFO

Article Type:
Case Report

Received: 25 December 2024
Revised: 19 January 2025
Accepted: 13 February 2025

**Corresponding author:*
Mohammad Parsa Ghazi

School of Dentistry, Isfahan University of Medical Sciences, Isfahan, Iran.

Tel: +98-21-84903747
Fax: +98-21-84903747
Email: dr.m.ghazi@khuisf.ac.ir

ABSTRACT

Undifferentiated pleomorphic sarcoma is a common soft tissue sarcoma in the human body, but it is rarely reported in the oral cavity. This article aims to report a case of UPS in the mandible. An 83-year-old female patient was referred to the Department of Oral and Maxillofacial Pathology at the Faculty of Dentistry in Isfahan, Iran, with a complaint of sudden, painless swelling under her removable complete denture, which had grown gradually over three months. The size of the lesion was approximately 5cm, and there were no radiographic defects present. Neoplastic and malignant proliferation of cells, which is necessary for UPS final diagnosis, was present in the H&E microcopy evaluation. Furthermore, immunohistochemical examination proved that the lesion is an undifferentiated pleomorphic sarcoma. Because of the late diagnosis of the disease, the patient died due to lung metastasis before any treatment.

Keywords: Case report; Surgery; Undifferentiated pleomorphic sarcoma; Malignant fibrous histiocytoma.

Please cite this Article as:
Khalesi S, Ghazi MP, Movahedian Attar B, Razavi SM. Oral Undifferentiated Pleomorphic Sarcoma: A Case Report. J Craniomaxillofac Res 2025; 12(2): 118-122. DOI:

Introduction

Undifferentiated pleomorphic sarcoma (UPS), formerly known as malignant fibrous histiocytoma (MFH), arises mainly in the soft tissues [1]. This tumor has been established as an entity in the World Health Organization (WHO) classification of soft tissue sarcoma (STS) since 2002 [2]. In general, the occurrence of head and neck sarcoma is rare. The nasal cavity and paranasal sinuses are the most common locations of UPS in the head and neck region, and only 0.1% of them appear in the oral cavity, which is very uncommon [3]. Although its etiology is still unknown, genetic predisposition, environmental factors such as trauma, history of radiotherapy, and the development of malignant tumors from benign lesions have been proposed as its etiology [4]. Symptoms usually depend on the involved anatomical site, but the neoplasm usually presents as rapid tissue growth with or without pain. Due to the highly invasive potential of this tumor, the frequency of metastases is very high at the time of initial tumor diagnosis [5]. Moreover, head and neck UPSs with bone invasion have a more aggressive invasion and poor prognosis than tumors confined to soft tissue [6]. Unlike carcinoma, several subtypes of sarcomas exist, which do not have a specific tissue or cell origin. UPS is one of the tumors whose diagnosis is based on the rejection of other tumors, and immunohistochemical staining plays an essential role in its diagnosis [7]. Due to the rarity of this tumor in the oral cavity and the shortage of oral UPS cases reported in the English literature, the current study presents a rare case of oral UPS in an old patient with rapid growth and poor prognosis.

Case Report

An 83-year-old female patient was referred to the Department of Oral and Maxillofacial Pathology at the Faculty of Dentistry in Isfahan, Iran, with a chief complaint of sudden swelling under her complete removable denture. The patient reported that the lesion appeared 3 months ago under her removable complete denture, and since then, she has not been able to use her denture. In clinical examination, a reddish-brown lesion with an indistinct border was seen in the left alveolar mucosa of the mandible. The patient did not report any history of systemic disease, smoking, alcohol use, or trauma. Extra-oral examination revealed a tender lesion, a 5 to 6cm-sized mass in the left alveolar mucosa of the mandible. Cervical lymph nodes were not palpable. In the radiographic examination, there was no change or evidence of intraosseous lesions in the mandible (Figure. 1).

An incisional biopsy was performed on the patient under local anesthesia and sent to the laboratory for microscopic examination. Examination of the histopathological sections (Hematoxylin and eosin staining) of the lesion showed the appearance of neoplastic and malignant proliferation of plump, spindle-like cell clusters in a myxoid background arranged in a storiform pattern and showing scattered pleomorphism with remarkably active mitosis (Figure. 2). Moreover, there were multinucleated giant cells in the connective tissue stroma. The differential diagnosis includes spindle cell carcinoma, carcinosarcoma, and fibrosarcoma. In immunohistochemical staining, vimentin was strongly positive. CD68 was focally positive, and Ki-67 was positive in 60% of the tumoral cells. But CD34, desmin, S-100, and cytokeratin were negative (Figure. 3). These results led to a final diagnosis of UPS. After the final diagnosis, the patient was referred to an oral and maxillofacial surgeon for final treatment and removal of the lesion. Unfortunately, due to the rapid growth of the lesion and the delay in treatment, the patient died due to lung metastasis without any further treatment after 2 months of early diagnosis.

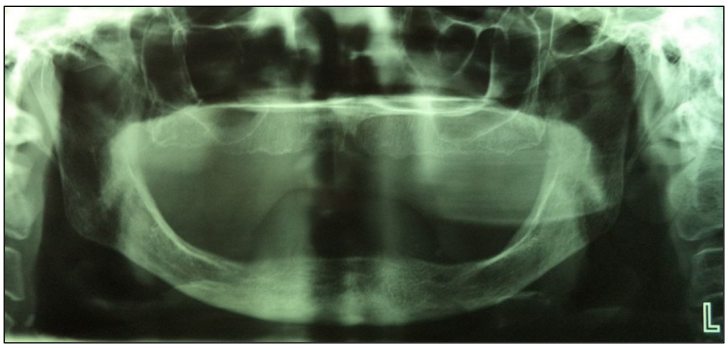

Figure 1. Panoramic image of the patient without intraosseous lesion.

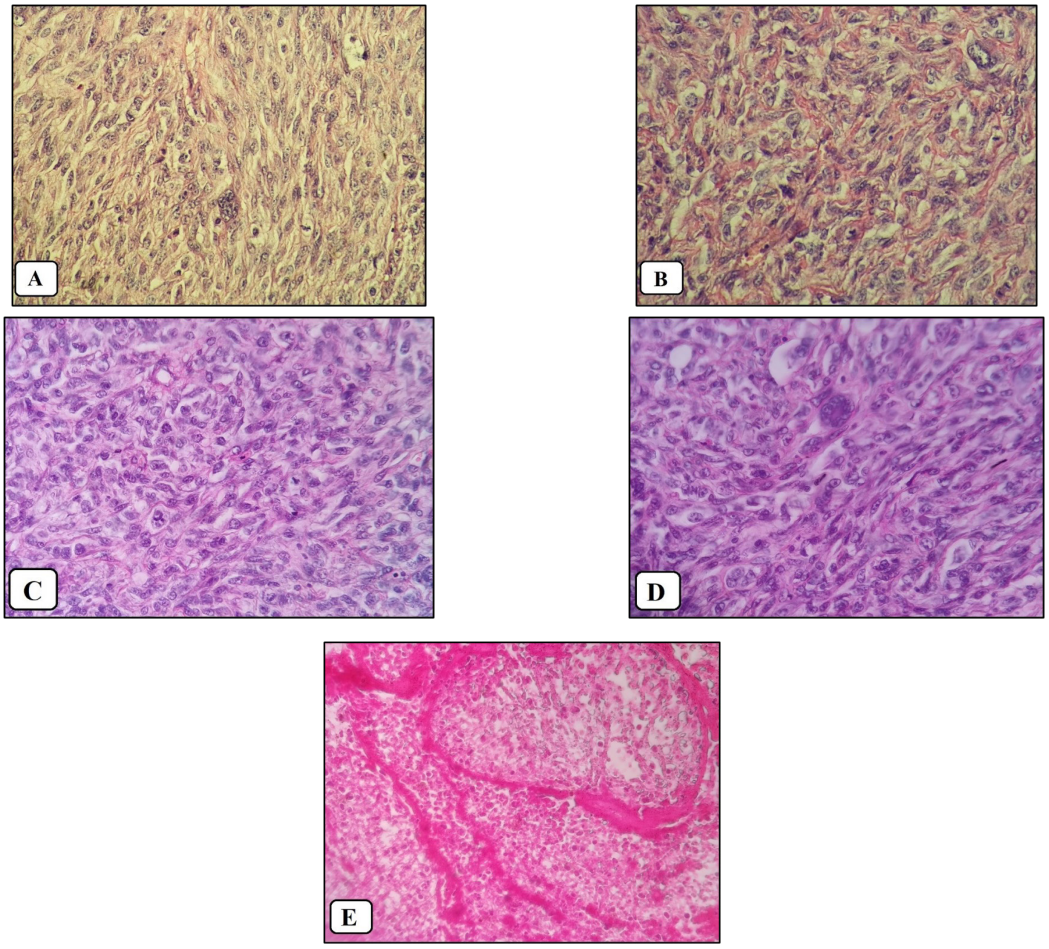

Figure 2. (A) Histo-pathological view revealed a hypercellular tumor mass with spindle-like cells and multinucleated giant cells, (B) Storiform pattern and active mitosis, (C) Atypical mitosis, (D) Nuclear pleomorphism, (E) Necrosis (H&E, ×400).

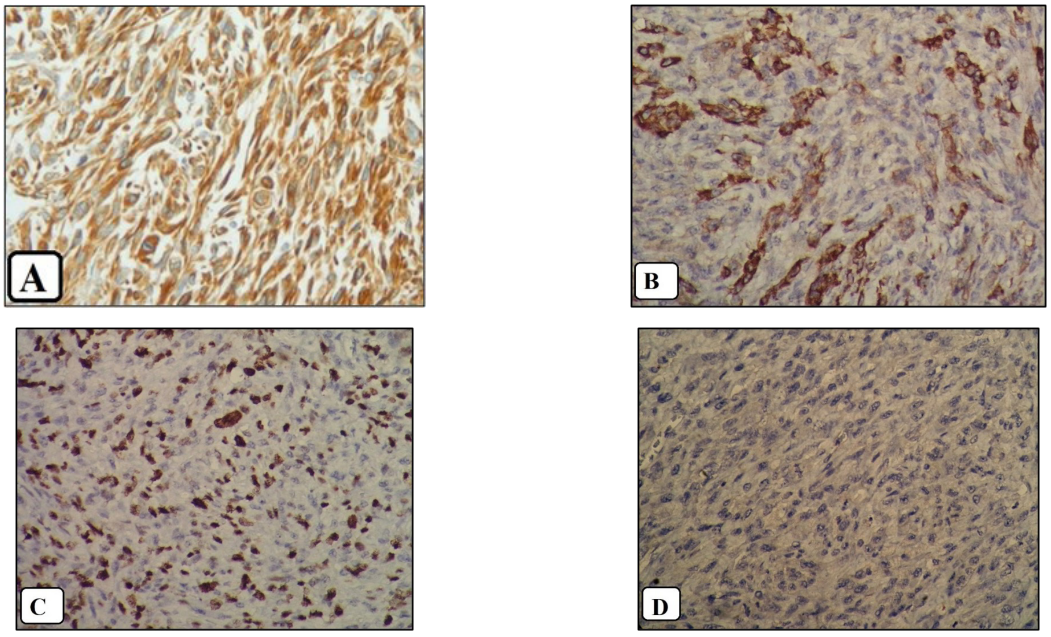

Figure 3. (A) Vimentin reactivity is detected. (B) Focally positive for CD68. (C) Ki67. (D) Negative results for cytokeratin, desmin, S100, and CD34 (Immunohistochemical staining, ×400).

Discussion

The renaming of malignant fibrous histiocytoma to undifferentiated pleomorphic sarcoma by the WHO represents an eventual common pathway in tumors that progress to undifferentiated forms [8]. According to most studies, UPS shows features of both histiocytic and fibroblastic differentiation [7]. Predominant spindle cell components with cytologic and nuclear pleomorphism and peripheral histiocyte-like cells with storiform pattern have been observed. Atypical mitosis and necrosis have also been observed in many cases [9]. Histopathologically, five subgroups of this tumor have been identified, including storiform/pleomorphic, myxoid, giant cell, inflammatory, and angiomatoid. The giant cell type exhibits a more aggressive clinical behavior and a poorer prognosis. In contrast, the myxoid type is the most common and has a better prognosis than other types [10]. The current case had a giant cell type of histology. Maybe this is one of the reasons for early death and high initial invasion of this tumor. This tumor typically demonstrates only vimentin in immunohistochemical staining. Vimentin positivity indicates the mesenchymal origin of the tumor [6]. As observed in this study, the majority of the tumoral cells studied expressed only vimentin with high intensity in immunohistochemical staining [2,3,5-7]. According to the standard stains histopathological view, many differential diagnoses such as carcinosarcoma, leiomyosarcoma, angiosarcoma, Liposarcoma, malignant peripheral nerve sheath tumor, rhabdomyosarcoma, malignant melanoma, malignant lymphoma can be suggested that the negativity of this tumor for cytokeratin, SMA, CD31, CD34, S100, desmin, HMB45, LCA causes the correct diagnosis [6]. In some cases, there has been a variable reaction to CD68 [5,11,12]. Similar to Benites's study, CD68 was locally positive in this study [5].

According to the reported cases, the oral soft tissue is more involved than the jawbone. It is often a nodular mass with or without pain that expands over a few weeks or months [6,7]. The mandible alveolar mucosa was involved in 70% of Oral UPSs [5], and only 30% of patients complained of pain in the head and neck region. However, other symptoms such as trismus, dysphagia, and nasal obstruction were observed [13]. In this case, the patient had complained of a rapidly growing painless soft tissue mass for three months. Oral UPS is commonly observed in older adults aged 50 to 70 years and is more prevalent in males than females [5,6]. In this report, an oral UPS in an 83-year-old female patient was presented, which is a rare case. Complete surgical removal of the neoplasm with clear margins is the treatment of choice. Regional metastasis or cervical lymph node metastasis is rare, occurring in only 15% of cases. Furthermore, radiotherapy can be recommended in unresectable instances or in cases where a margin may be defined [5,6]. However, reported head and neck UPSs have higher rates of recurrence and distant metastasis (25-35% of cases) to lung, liver, and bone, as observed in this patient and Vivek's and Benites's cases [3,5]. Therefore, adjuvant chemotherapy may be warranted for histologically high-grade tumors with a history of local recurrence [1,6]. The prognosis of oral UPS depends on tumor depth, size, stage, and whether it's primary or recurrent. The 5-year survival rate varies from 30% to 70% [3,7,8]. This case had no lymph node metastasis at the initial examination and before surgery. Eventually, due to a delay in referral and incomplete obliteration of the lesion, lung metastasis was diagnosed one month after the first surgery.

Conflict of Interest

There is no conflict of interest to declare.

References

[1] Widemann BC, Italiano A. Biology and management of undifferentiated pleomorphic sarcoma, myxofibrosarcoma, and malignant peripheral nerve sheath tumors: state of the art and perspectives. J Clin Oncol 2018; 36(2): 160-7.

[2] You Y, Shi R, Dai M, Du Z, Tian Z, Xu G, et al. Primary undifferentiated pleomorphic sarcoma in oral-maxillary area: retrospective study and molecular analysis. Histol Histopathol 2021; 36(8): 889-98.

[3] Vivek N, Magesh KT, Sivachandran A, Kumar MS. Undifferentiated pleomorphic sarcoma of maxilla presenting like a periapical pathology – a case report. J NTR Univ Health Sci 2018; 7: 70-3.

[4] Shahoon H, Esmaeili M, Nematollahi M. Eight year follow up of malignant fibrous histiocytoma (undifferentiated high grade pleomorphic sarcoma) of the maxilla: case report and review of the literature. J Dent Res Dent Clin Dent Prospects 2009; 3: 32-5.

[5] Benites BM, Miranda-Silva W, Fonseca FP, de Oliveira CRGCM, Fregnani ER. Undifferentiated pleomorphic sarcoma of the mandible. J Korean Assoc Oral Maxillofac Surg 2020; 46: 282-7.

[6] Kim CH, Jang JW, Kim MY, Kim YH, Kim HG,

Kim JH. Undifferentiated pleomorphic sarcoma in mandible. Maxillofac Plast Reconstr Surg 2014; 36(6): 303-7.

[7] Archana TS, Shetty A, Imran M, Ahmed N, Kumar P, Shruthi TM. A rare entity of undifferentiated pleomorphic sarcoma of the mandible - a case report. Ann Maxillofac Surg 2021; 11(1): 156-9.

[8] Lambade PN, Lambade D, Saha TK, Bande CR, Ramakrishana A. Malignant fibrous histiocytoma: an uncommon sarcoma with pathological fracture of mandible. J Maxillofac Oral Surg 2015; 14: 283-7.

[9] Amm HM, DeVilliers P, Srivastava AR, Diniz MG, Siegal GP, MacDougall M. Mandibular undifferentiated pleomorphic sarcoma: Molecular analysis of a primary cell population. Clin Exp Dent Res 2020; 6: 495–505.

[10] Chen S, Huang W, Luo P, Cai W, Yang L, Sun Z, et al. Undifferentiated pleomorphic sarcoma: long-term follow-up from a large institution. Cancer Manag Res 2019; 11: 10001-9.

[11] Thompson LD. Pleomorphic sarcoma of the neck. Ear Nose Throat J 2015; 94: 376-7.

[12] Rajan ST, Elengkumaran S, Narasimhan M, Ravindran C. Pleomorphic sarcoma of the oral cavity: a rare case entity. J Pharm Bioallied Sci 2021; 13(Suppl 2): S1747-S1749.

[13] O'Neill JP, Bilsky MH, Kraus D. Head and neck sarcomas: epidemiology, pathology, and management. Neurosurg Clin N Am 2013; 24: 67-78.