

CASE REPORT

Maxillary undifferentiated high-grade pleomorphic sarcoma: A case report and review of the literature

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ABSTRACT

Undifferentiated high-grade pleomorphic sarcoma is a tumor of mesenchymal cells without clear cell differentiation and represents a diagnosis of exclusion. It is located mainly in extremities, so its location in the head and neck is extremely rare. We report the case of an undifferentiated high-grade pleomorphic sarcoma located on the hard palate.

Key Words: Sarcoma, Undifferentiated, High-grade, Pleomorphic, Lung, Metastases

1. INTRODUCTION

Undifferentiated high-grade pleomorphic sarcoma is a soft tissue sarcoma composed of undifferentiated mesenchymal cells that have a fibrohistiocytic morphology without differentiation to a specific cell line.^[1] They represent a diagnosis of exclusion.^[2,3]

They correspond to 5% of soft tissue sarcomas in adults, occurring mainly in the lower extremities in patients during the sixth and seventh decade of life.^[2] The location in the head and neck is very rare with an incidence of 4% to 10%, being the most frequent sites the neck and the parotid gland.^[4] Clinically they are presented as a mass of progressive and rapid growth that may be associated with pain of the compromised site. Histologically they present a heterogeneous cell pattern with pleomorphism and aberrant giant cells with nonspecific immunohistochemical markers due to the lack of cellular differentiation, being only positive for vimentin.^[5]

Their presence in oral cavity is extremely rare^[6] and it is difficult to estimate the cases previously reported in the literature given the recent change in the classification of the World Health Organization of these tumors.^[2,6] The aim of this paper is to present a case of undifferentiated pleomorphic sarcoma of the oral cavity emphasizing the clinical, imagenological and histological presentation.

2. CASE REPORT

A 47-year-old patient without a significant medical history, consulted in the Maxillofacial Surgery Service of Hospital Guillermo Grant Benavente of Concepción with a one-year history characterized by the appearance of a lesion in the hard palate that was biopsied in multiple occasions being diagnosed as fibrous dysplasia. However, during the last month the lesion presented a painful progressive and fast growth associated with bleeding episodes. Physical examination showed a reddish brownish-friable lesion of 5 cm ×

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3 cm in the hard palate, different from the previously biopsied lesion. An incisional biopsy of the lesion was performed that informed the presence of undifferentiated spindle and pleomorphic cells with a vaguely storiform growth pattern. Immunohistochemistry was positive for vimentin, CD99 and BCL2. It was negative for p53, keratin AE1/AE3, keratin 5/6,

S100, CD 31, CD 34, actin, desmin, myogenin and TLE1 (see Figures 1-2). An undifferentiated high-grade pleomorphic sarcoma was diagnosed and the patient was sent to the Otolaryngology and Head and Neck Surgery Service of the same hospital for extension study and management.

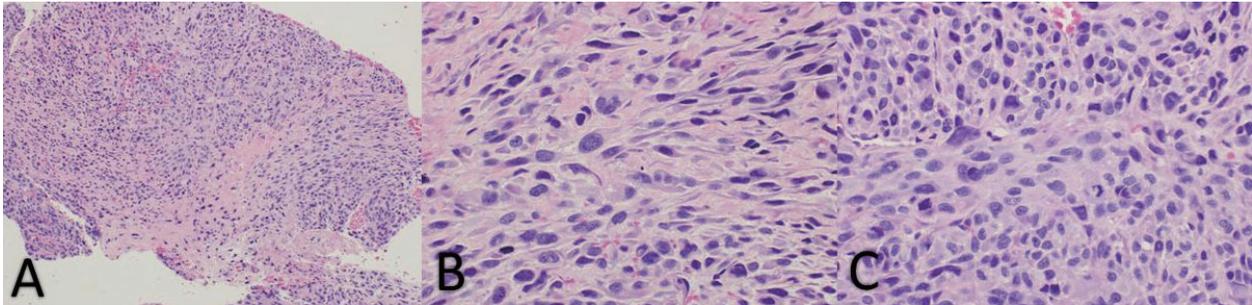


Figure 1. Hematoxylin and Eosin stain

A: epithelioid and spindle cell malignant tumor with marked cellular pleomorphism; B: spindle cell tumor area; C: epithelioid tumor area.

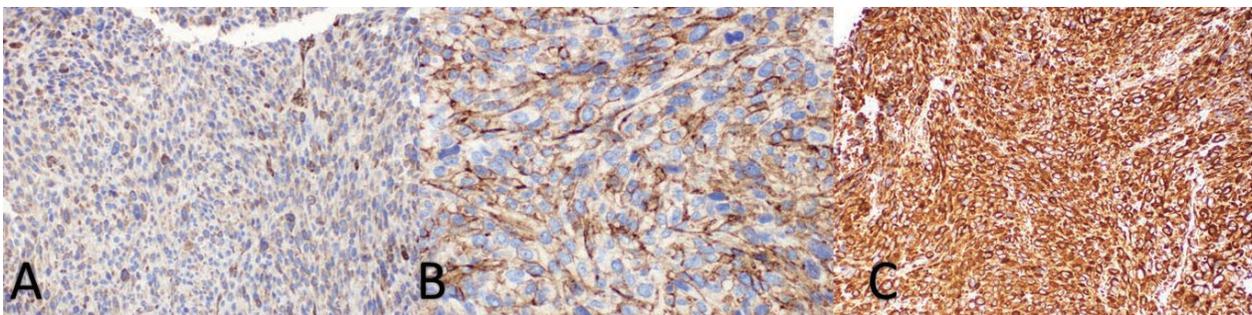


Figure 2. Immunohistochemistry study showed positive antigens

A. BCL2; B. CD99; C. Vimentin.

After the biopsy, the patient presented rapidly growing of the lesion associated with pain and bleeding events up to 800 ml, requiring transfusion of red blood cells in two opportunities. A computed tomography of paranasal sinuses, neck, thorax, abdomen and pelvis was requested demonstrating a destructive bone lesion of the right maxillary bone with irregular and heterogeneous contours, infiltration of the alveolar ridge, total occupation of the ipsilateral maxillary sinus, with infiltrative commitment of the inferior turbinate, the nasal septum and floor of the orbit, with obstruction of the nostril on the same side and destruction and invasion of the hard palate in its two anterior thirds. In the extension study highlights the presence of multiple bilateral pulmonary nodules suggestive of metastases (see Figures 3-5).

The case was presented and discussed to the Head and Neck Tumor Board (HNTB), where it was staged as a T4N0M1 maxillofacial tumor. The HNTB recommended palliative

chemotherapy and palliative care because of the presence of lung metastases in a patient with an undifferentiated high-grade pleomorphic sarcoma. The patient was considered a bad candidate for surgery and radiotherapy due to the extension of the tumor. Due to the size of the lesion and possible airway compromise, it was decided to do a tracheostomy, gastrostomy and installation of central venous catheter with subcutaneous reservoir.

Currently the patient is in palliative care, pending the start of palliative chemotherapy. He presented two bleeding episodes requiring emergency radiotherapy (two doses of 8 Gy) with which it was possible to control the bleeding (see Figure 6).

3. DISCUSSION

Squamous cell carcinoma and lymphoma are the most common cancers of head and neck representing 80%-90% of the total cases, while sarcomas are relatively rare in this area

representing between 1% to 10% of the neoplasm's.^[4]

Undifferentiated high-grade pleomorphic sarcoma is currently a diagnosis of exclusion and represents 5% of soft tissue sarcomas in adults.^[2,8] It is a soft tissue sarcoma composed of undifferentiated mesenchymal cells.^[1] It usually occurs in extremities, being more common in lower extrem-

ities.^[4] Its location in the head and neck is rare, with an incidence of 4% to 10%. The most frequent location in head and neck is the neck and parotid gland, followed by the scalp, face, anterior skull base and orbit, there are few reports in upper aerodigestive tract, lateral skull base and ears.^[1,7,9,11] Their presence in oral cavity is extremely rare.



Figure 3. CT bone window

A axial plane, destructive and expansive process eroding maxillary sinus walls. B coronal plane, infiltration of the alveolar ridge, nostril, floor of the orbit and hard palate

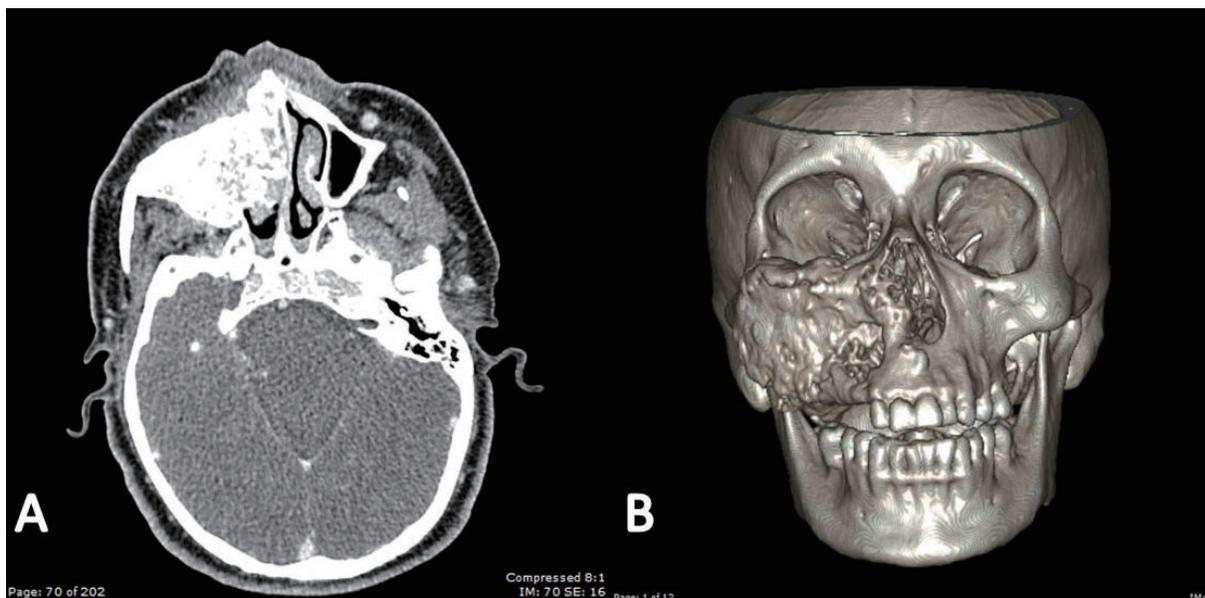


Figure 4. A. Axial CT showing moderate enhancement after intravenous contrast administration; B. Three-dimensional CT image with osseous volume rendering.

There have been reports of predisposing factors such as history of recurrent trauma, surgery, radiation, fractures, osteonecrosis, Paget's disease, non-ossifying fibroma and fibrous dysplasia.^[10] In our case the patient had a history of multiple biopsies of the area informed as fibrous dysplasia.

Clinically, the most common symptom is the presence of a mass of progressive and rapid growth that may be associated with nonspecific symptoms such as pain and paresthesia of the compromised site. Imaging studies also are non-specific, showing the computed tomography and magnetic resonance the presence of a lobulated soft tissue mass.^[10] The clinical and imaging presentation of our patient was consistent with that reported in the literature.

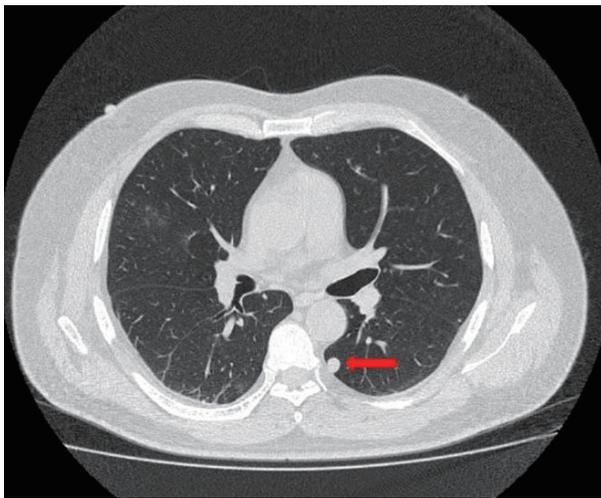


Figure 5. CT showing metastatic nodule in the left lung (red arrow)

The definitive diagnosis is by histopathological examination of samples obtained by incisional biopsy. Less invasive techniques such as fine needle aspiration have low sensitivity ranging from 60% to 80% versus 94% of the incisional biopsy.^[12] Histologically they present a heterogeneous cell pattern with pleomorphism and aberrant giant cells with nonspecific immunohistochemical markers due to the lack of cellular differentiation, being only positive for vimentin.^[5,13] In our case the immunohistochemistry was positive for vimentin, CD99 and BCL2 and negative for p53, keratin AE1/AE3, keratin 5/6, S100, CD 31, CD 34, actin, desmin, myogenin and TLE1.

Cervical metastases are rare, occurring in 3%, 2% to 18% of cases. Metastases have been reported more frequently in the lung.^[14,19] Our patient had bilateral cervical lymphadenopathy of inflammatory aspect and multiple bilateral pulmonary nodules suggestive of metastases.

The management of the sarcomas are not always applicable to the head and neck, due to complex anatomy and the difficult of obtain wide surgical margins.^[16] The treatment of choice is surgery.^[15] Adjuvant radiation therapy is recommended after surgery with positive margins and regional lymph node dissection for nodal metastases. This, however, do not improve the survival rates because succumb to distant metastases.^[17] Chemotherapy is used for palliation or as adjuvant therapy in cases of aggressive behavior or potential metastases for maximizing local control and treating the potential of micrometastatic disease.^[17,18]

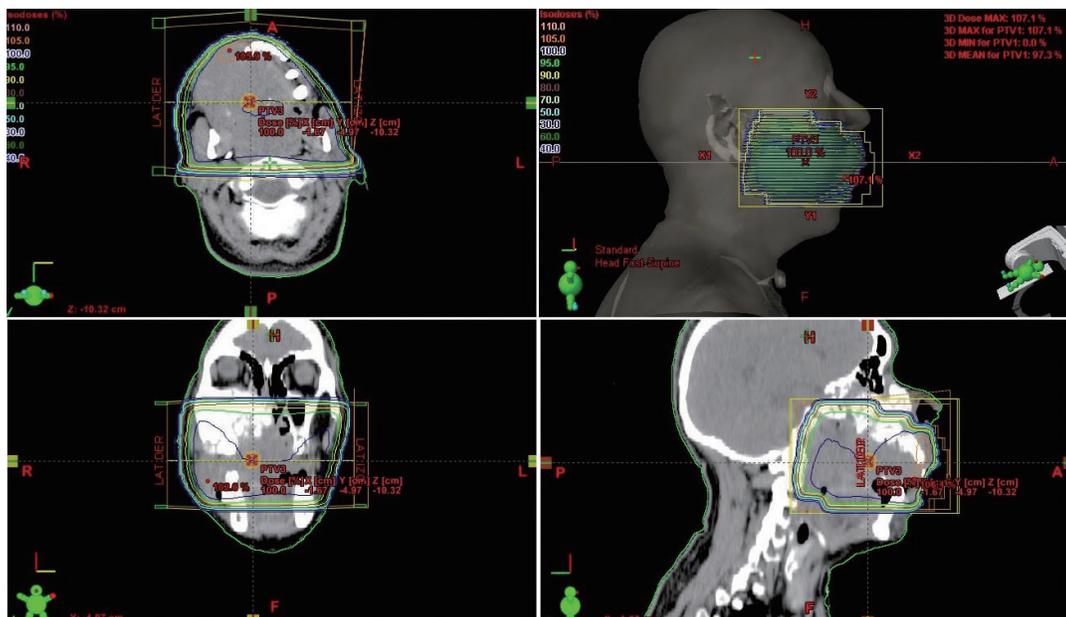


Figure 6. 3-dimensional conformal radiation plan

Effective targeted treatment is unavailable for most sarcomas and doxorubicin and ifosfamide-which have been used to treat soft-tissue sarcoma for more than 30 years-still have an important role. Whether doxorubicin alone or the combination of doxorubicin and ifosfamide should be used routinely is still controversial.^[20] Combination therapy is critical to success, being the 5-year survival of 50%-60%.^[1]

4. CONCLUSION

Undifferentiated high-grade pleomorphic sarcoma of oral cavity is a rare neoplasm of mesenchymal cells in head and

neck. Usually it presents as a rapidly growing mass in the oral cavity that may be associated with local pain and paresthesia. Because of nonspecific clinical and imaging features it is essential the histological analysis for its diagnosis, emphasizing the presence of pleomorphic cells without cell differentiation with positive immunohistochemistry for vimentin. The treatment of choice is surgery associated with adjuvant radiotherapy.

CONFLICTS OF INTEREST DISCLOSURE

Authors declare have no interest conflict.

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