CASE REPORT

Malignant peripheral nerve sheath tumor of the maxilla

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Summary

We present a case of a malignant peripheral nerve sheath tumor [MPNST] of the maxilla corresponding to a 12 year old male who had a painful, ulcerated rapid growing tumor. Histology revealed the presence of a tumor consisting of fusiform cells with abundant mitosis, with negative cytokeratins, actin, desmin, myoglobin and factor VIII. In contrast, the protein S-100 was positive. MPNST was not associated with signs of Neurofibromatosis Type 1. The patient was surgically intervened and received radiotherapy, then died within 10 months posterior to surgical intervention after suffering recurrence.

KEYWORDS
Malignant peripheral nerve sheath tumor; Neurosarcoma; Maxilla

Introduction

Malignant peripheral nerve sheath tumors [MPNST] are very rare malignant neoplasias. They have been described so much to be in association with type 1 Neurofibromatosis¹ and isolated forms also.² It deals with a malignant lesion originated from the neural tissue that shows significant histological likeness to fibrosarcoma. Differential diagnosis is facilitated when the patient presents a previous history of fibromatosis. MPNTS is characterized by the presence of very irregularly shaped cells with nuclei that exhibit a “wavy, buckled appearance”. Tissue organization shows great variability, with hypocellular myxoid areas and areas of major cellularity.¹ The histologic changes of MPNST include spindle cells with comma-shaped nuclei, tactoid bodies, nuclear palisading, hyaline bands, and schwannoma-like and curlicue foci.³

Cases of MPNST have been reported mostly in the soft tissues,⁴ less common in the bones⁵ and very rare in the maxilla.¹,⁶ The aim of this article is to present a case of MPNST in the maxilla showing its clinico-pathologic characteristics and evolution.

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Case report

A twelve year old male patient with a history of having pneumonia 10 years previously and occasional cephalaeas since the age of 3. Visited our consultation service of Stomatology at the University General Hospital of Valencia for presenting pain in the superior maxilla for the past 2 months.

Upon intraoral examination a well delimited and rapid growing tumor was appreciated in the upper jaw, on the right side (Fig. 1), of 2×3 cm in size and with mobility of the superior right canine and both premolars of the same side.

Orthopantomography (Fig. 2), computerized axial tomography (CAT) and blood tests were requested and incisional biopsy of the tumor was performed. An osteolytic zone in the apical area of the canine and the premolars were appreciated in the orthopantomography. Maxillofacial CAT scan revealed a lytic lesion in the right part of the maxilla, with destruction of the bone of the hard palate. It was very poorly delimited (Fig. 3). Blood analysis was normal in the three hematologic series, so as the hemostatic variables. Sedimentary rate (SED) was 59 mm in the first hour (normal reference values of: 1–13). Alkaline phosphatase and calcium levels were normal.

Histopathologic studies revealed a solid tumor, extensively necrotized, consisting of fusiform cells and epithelial cells with marked pleomorphism, bizarre and multinucleated giant cells, observing abundant mitosis (6 mitosis/10HPF) (Figs. 4 and 5). The diagnosis was sarcoma of a high grade malignancy, extensively necrotized. Immunohistochemical studies showed tumor cells negative for cytokeratines, actin, desmin, myoglobin and Factor VIII. It was positive to Vimentin and Protein S-100.
All of which lead to the diagnosis of malignant peripheral nerve sheath tumor of the maxilla.

With this diagnosis the patient was surgically intervened performing a hemimaxillectomy and resectioning borders free of tumor. The patient later received radiotherapeutic treatment. In spite of everything the patient suffered a relapse on the same original area, died 10 months after surgical intervention.

Discussion

In spite of being uncommon, malignant peripheral nerve sheath tumors have been described in different locations of the body, occasionally associated or not with Neurofibromatosis type 1. Thus, isolated cases in the orbit,7 neck,8 parapharyngeal region,2 colon,9 pancreas,10 duodenum,11 thyroid gland,12 vulva13 and uterus14 have been reported. On occasions the existence of the tumor is related to the presence of Neurofibromatosis or von Recklinghausen’s disease, with described cases located in the bladder,15 thorax,16 orbit,17 mediastinum18 and prostate.16

Two series of MPNSTs of the head and neck have been presented. Loree et al.20 informed of 17 cases in 9 men and 8 women, whose mean survival in 5 years was 52%, seven of them was associated to Neurofibromatosis type 1. Vege et al.21 presented 27 patients with the average age of 42 years, with lesions in which 44.6% of the cases were in the neck. 18.5% of the patients had distant metastases and survival within 5 years was at 33%.

Primary MPNTs in the bones are exceptionally rare.5 In this concept isolated cases in the spinal column22 and femur5 have been reported. Neville et al.1 presented two cases of primary MPNSTs in the maxilla, both of which were associated to a neurofibromatosis type 1. The first patient was a 27 year old woman with a painful tumor lesion in the superior maxilla. Radiology revealed an osteolytic area which includes the alveolar bone and the right maxillary sinus. The second case corresponded to another woman, however of 74 years old, that also consulted the clinic for presenting a painful tumoral mass, although this time in the mandible.

Our case, different from the two cases presented by Neville et al.1 was a 12 year old male, but with a clinical case very similar to the first case of those described by the previous authors, since it dealt with a superior maxillary tumor on the right side, of rapid growth, with an ulcerated surface and with significant osteolysis evidenced in both orthopantomography and the CAT scan. Histopathologic studies revealed fusiform cells with epitheloid habit, marked nuclear pleomorphism and abundant mitosis, for which the tumor was catalogued as a sarcoma. The negativity for epithelial, muscular and vascular markers, so as the positivity for protein S-100 established the final diagnosis of MPNST.

After immunohistochemical studies of 12 MPNST cases Mawrin et al.23 suggested that alterations in the protein P53 play an important role in the control of the cellular reproductive cycle and the production of tumors, both in sporadic cases of MPNST and in those associated with neurofibromatosis.

The recommended treatment of this type of sarcoma is the surgical extirpation including wide margins and complementary radiotherapy.24 However, the prognosis is poor.1,21 In this perspective, Wenabo et al.24 demonstrated that the mean survival of his 28 cases with MPNSTs was 44 months. If a patient, in conjunction with MPNSTs has a neurofibromatosis type 1, the prognosis is better; although Loree et al.20 think otherwise.

Just as what happened to both maxillary bone cases presented by Neville et al.1 the evolution of our patient was poor. Dying within ten months in spite of the performed treatments.

References


