Malignant Peripheral Nerve Sheath Tumor in the Maxilla: Report of a Rare Case

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Abstract

Malignant peripheral nerve sheath tumor (MPNST) is a rare malignant tumor that develops either from a preexisting neurofibroma or *de novo.* The cell of origin is believed to be the Schwann cell and possibly other nerve sheath cells. In this report, we describe a rare case of MPNST that arise from the socket of second left maxillary molar that has been already extracted in a young man. He was referred to a dentist's office with a tumor-like mass of soft tissue on his left maxillary gingiva. Biopsy and histopathologic examination was performed and based on histologic and immuno-histochemical findings, the diagnosis of MPNST was made. MPNST is a rare malignant tumor in the oral cavity. Dentists must be careful and conscious because this rare malignancy can occur in gingiva and can mimic the clinical feature of any benign gingival enlargements.

Key Words: Malignant peripheral nerve sheath tumor, maxilla, neurofibroma.

Introduction

The principal malignancy of peripheral nerve origin is preferably called a malignant peripheral nerve sheath tumor (MPNST). These tumors account for 5% of all soft tissue sarcomas, with about half of such cases occurring in patients with neurofibromatosis type I. The lesion is most common on the proximal portions of the extremities and the trunk: only 10% to 15% of cases occur in the head and neck (1). The cell of origin is believed to be the Schwann cell and possibly other nerve sheath cells (2). Malignant peripheral nerve sheath tumors are most common in young adults. The mean age in patients with neurofibromatosis (29 to 36 years) is about one decade younger than in those without this condition (40 to 46 years). The tumor is an enlarging mass that sometimes exhibits rapid growth. Associated pain or a nerve deficit is common.

Oral tumors may occur anywhere, but the most common sites are the mandible, lips, and buccal mucosa (1). In this report, we describe a young male patient with an enlarging soft tissue mass in his second left maxillary molar socket that resemble any benign soft tissue mass in the absence neurofibromatosis type I with diagnosis of MPNST based on clinical, histo-pathological and immunohistochemical examination.

Case Report

A 24-year-old man referred to a dental office at December 2009. The chief complaint of the patient was an enlarging soft tissue mass that proliferated from his second left maxillary molar socket into the

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oral cavity (Fig. 1). His third and second left maxillary molar was extracted already by a general dentist because of mobility of these teeth. The patient's medical and family history showed no document of a syndrome or serious general disease such as neurofibromatosis type I.

CT Scans of this region revealed a soft tissue mass that pushed into the left maxillary sinus cavity that approximately obliterated by this lesion (Fig. 2). Patient's maxillary and mandibular teeth were healthy without carries and calculus. Biopsy and histopathologic examination was performed. Microscopic evaluation of H & E stained sections revealed proliferation of spindle cells with comma and wavy shaped nuclei, pleomorphism and hyperchromasia of nuclei as well as abnormal mitosis and abundant blood vessels (Fig. 3). Immunohistochemical investigation revealed strong positivity for S-100 protein (Fig. 4) and diffuse positivity for vimentin. Tumoral cells showed negativity for HMB45, CD68, desmin and CD31. Based on these findings, diagnosis of MPNST was made and melanoma, malignant fibrous histiocytoma, leiomyosarcoma and angiosarcoma eliminated from the diagnostic list of diseases.

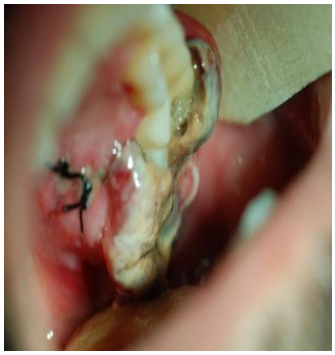

Figure 1. Intraoral view: the huge soft tissue mass that proliferated from the socket of second molar

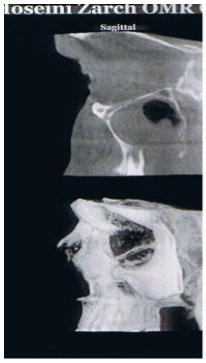

Figure 2. CT scan of left maxillary sinus shows tumoral mass that pushed into the sinus cavity

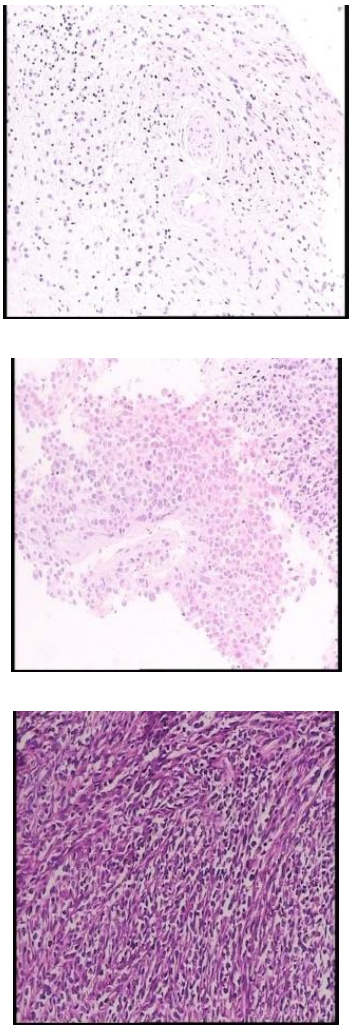

Figure 3. Upper image: Section of tumor. Fascicles of neural cells are seen. Middle and lower images: Epithelioid and sarcomatous shape of tumoral cells is visible (H & E staining, Magnification: x100)

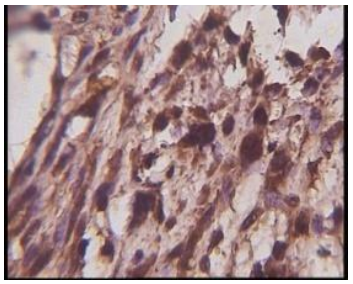

Figure 4. Strong positivity for S-100 protein (Magnification: x400)

Discussion

The principal malignancy of peripheral nerve origin is malignant peripheral nerve sheath tumor. These tumors account for 5% of all soft tissue sarcomas, with about half of such cases occurring in patients with neurofibromatosis type I (1). The cell of origin is believed to be the Schwann cell and possibly other nerve sheath cells (2). Malignant peripheral nerve sheath tumors are undoubtedly uncommon particularly in the mouth or jaws (3). The lesion is most common on the proximal portions of the extremities and the trunk: only 10% to 15% of cases occur in the head and neck (1), common in young adults. The mean age in patients with neurofibromatosis (29 to 36 years) is about one decade younger than in those without this condition (40 to 46 years). The tumor is an enlarging mass that sometimes exhibits rapid growth. Associated pain or a nerve deficit is common. Oral tumors may occur anywhere, but the most common sites are the mandible, lips, and buccal mucosa (1,3,4). Microscopic evaluation of MPNST shows fascicles of atypical spindle-shaped cells, which often resemble the cells of fibrosarcoma (1). However, these cells are frequently more irregular in shape with wavy or comma-shaped nuclei. In addition to streaming fascicles, less cellular myxoid areas also may be present. A definitive diagnosis of neural origin is often difficult, especially in the absence of neurofibromatosis. Positive immunostaining for S-100 protein is a helpful clue, but this is found in only about 50% of all cases (1-3). Traditionally, MPNST has been among the most challenging soft tissue tumor diagnoses to make because of the lack of standard histologic criteria (4). The capacity of MPNSTs to undergo focal mesenchymal (or even epithelial) differentiation is well known. Epithelial areas may be histologically benign; however, mesenchymal differentiation is sarcomatous in nature and histologically malignant (4,5).

Here, we describe a rare case of MPNST that is unique not only because of its location (in the socket of maxillary second molar), but also because of its unusual clinical presentation as a huge soft tissue that caused mobility of three maxillary left molars without pain or paresthesia as well as it's occurrence in a young patient (24-year-old) that is unusual especially in the absence of neurofibromatosis syndrome type 1. As previously mentioned, biopsy and histopathologic examination was performed and based on microscopic features of this tumor, malignancies with epithelioid cells, neural origin, vascular origin and spindle cells came into the diagnostic list. Because of its neuro-mesenchymal architecture, immunohistochemical staining for S-100 protein, vimentin recommended. Also, because of epithelioid appearance of tumoral cells that resembles to malignant melanoma HMB45 immunohistochemical staining (6) and also because of angiogenesis of tumors, immunohistochemical staining for CD31 for rull out of angiosarcoma and desmin for leiomyosarcoma were performed respectively. Microscopic findings revealed positivity for S-100, protein, and vimentin but negativity for CD68, HMB45, CD31 and desmin. Based on histopathologic findings and very aggressive clinical behavior of tumor that was pushed into the maxillary left sinus cavity in a short time, diagnosis of MPNST for this tumor was made and to our knowledge this case report is the first case of MPNST in the socket of extracted tooth in Iran and English articles.

The treatment of malignant peripheral nerve sheath tumors consists primarily of radical surgical excision, possibly along with adjuvant radiation therapy and chemotherapy. The prognosis is poor, especially in patients with neurofibromatosis (1,3) and is significantly worse than patients without NF1 (7). Based on a study, the most important prognostic factors for this disease are presentation with either primary or recurrent disease, tumor size, and the site of origin (8). For this case radical surgery was performed and patient underwent a regular follow up program.

Conclusion

MPNST is a rare malignancy in the oral cavity especially in sockets of teeth. This malignancy is very uncommon in young patients without NF1 syndrome and based on this case report, its clinical behavior can be very different as loosening of maxillary molar teeth and pushing into the maxillary sinus cavity without pain and paresthesia. Though, dentists must be careful and conscious while are making diagnosis of apparently simple and inoffensive soft tissue enlargement in the oral cavity, since this malignancy can mimic the clinical features of any benign lesions.

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