Bilateral Malignant Peripheral Nerve Sheath Tumor Associated with a Massive Osteoma of the Mandible

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Introduction: Malignant peripheral nerve sheath tumor (MPNST) is a highly aggressive soft tissue sarcoma, which is uncommon in the maxillofacial area. **Case Report:** We present a rare case of bilateral MPNST with an osteoma in the Mandible. Clinically, massive bilateral swelling in the mandibular angle with rapid growth for 9 months was seen. **Results:** Histological evaluation showed the proliferation of malignant spindle cells arranged in a fascicular pattern with infiltrating margins. Immunohistochemical evaluation was positive for S100 protein and vimentin and negative for CD 68, Desmin, and CD31. Resection the soft tissue lesions with debulking osteoma did not show any recurrence in five years follow-up. **Conclusion:** Surgical excision may be an essential treatment option to eradicate MPNST.

Keywords: Malignant Peripheral Nerve Sheath Tumor; Mandible; Neurofibromatosis Type 1; Osteoma

Introduction

Malignant peripheral nerve sheath tumor (MPNST) is a highly aggressive soft tissue sarcoma with a high rate of local recurrence and distant metastasis, which is commonly found in lower extremities and only 10% to 20% of all lesions occur in head and neck region, thus creating it a rare entity (1, 2). MPNSTs are capable of arising from pre-existing benign neurofibromas in patients with neurofibromatosis type 1 (NF1) or schwannoma (3).

The incidence of MPNST originating in NF is 4.6%, which is superior to 0.001% in the general population (4).

Solitary neurofibromas without an association with NF1 have a low recurrence rate after excision, and nonmalignant transformation potential is infrequent in these lesions (5). Lung is the most common metastatic site of MPNST (6).

To the best of our knowledge, we report the first case of bilateral MPNST associate with a massive osteoma in the Mandible. Previously, a few articles had been reported unilateral MPNSTs in the Mandible (2, 7). The management of this lesion and a brief review of the literature are described.

Case Report

A 21-year-old male presented to the department of Craniomaxillofacial surgery with a chief complaint of bilateral mandibular swelling. A moderately tender, firm, non-fluctuant mass in the vestibular sulcus was clinically observed. According to the patient's history, the swelling appeared bilaterally in mandible gradually increased its size over 9 months (Figure1). There was a history of moderate intermittent pain, difficulty in speech, and mastication. Fine needle aspiration biopsy did not show any significant materials.

History of presenting illness revealed that physicians visited him in his local area, the southwest of Iran, and they performed an incisional biopsy. The initial histopathology report demonstrated a neurofibromatosis lesion. The clinical evaluation did not express the signs and symptoms of von Recklinghausen's neurofibromatosis. Panoramic view illustrated bilateral opaque lesions in the angle of the Mandible; the involvement of the right side lesion was more significant than the left (Figure 2). The CT scan confirmed bilateral hard and soft tissue lesions with a diameter of 12×7×5cm in the right and 4×5×5cm on the left side, eroding the underlying border of mandible and destructed the medulla without significant lymph node enlargement (Figure 3). A chest x-ray did not show any metastatic lesion in the lung. The whole-body bone scan for ruling outdistance metastatic lesions was negative. In the second incisional biopsy, Hematoxylin and Eosin sections showed the proliferation of malignant spindle cells arranged in a fascicular pattern with infiltrating margins. The tumoral cells had comma and wavy nuclei, mild polymorphism,

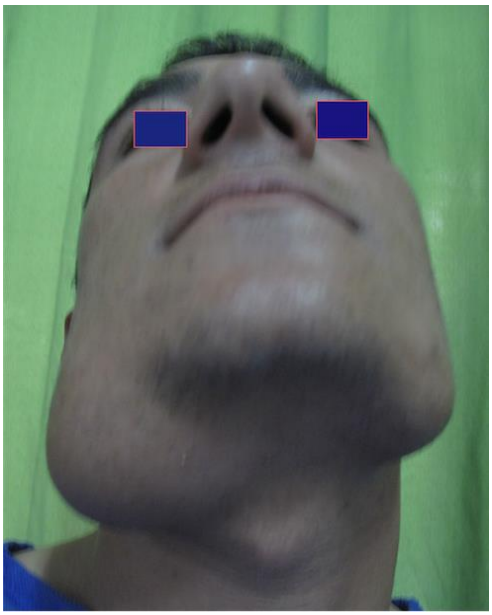

Figure 1. Pre-operation patient's view shows a bilateral swelling of the Mandible

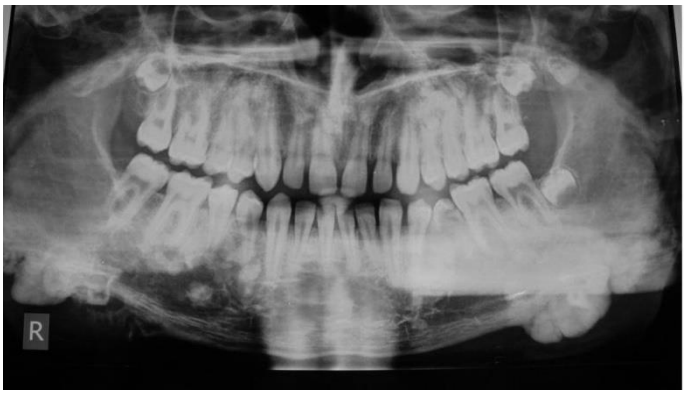

Figure 2. Pre-operation OPG view demonstrated opacity in the angle of the Mandible

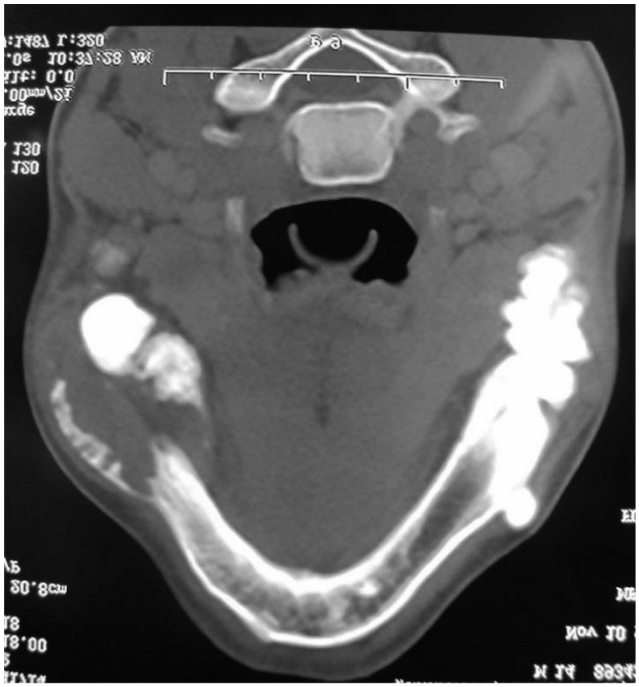

Figure 3. Axial view shows a mix lesion (opaque-lucent) in the right side

hyperchromatism, and a few mitoses. Immunohistochemical evaluation was positive for S100 protein and vimentin and negative for CD68, Desmin, and CD31 (Figure 4). These findings were in favor of low-grade MPNST.

In the hard tissue lesions, microscopic examination of the specimen disclosed a hard mass consisting entirely of dense lamellar compact bone.

Extraoral incisions were made bilaterally, i.e., 4 cm underneath of the mandibular border. Soft tissue lesions, MPNST, were excised entirely, and osteoma lesions were debulked. Clinically, there was no evidence of any recurrence during five years follow-up (Figure 5).

Discussion

MPNST is the inventive term defined by WHO as a sub-type of soft tissue sarcomas which is a malignant proliferation of any cell of the nerve sheath, i.e., Schwann cell, perineural fibroblast or endoneurial fibroblast. Two more than 50% of these tumors are associated with neurofibromatosis type 1 (8, 9).

Occasionally, the involvement of cranial nerves has been reported in MPNSTs, occurring de nova or by the malignant transformation from benign nerve sheath tumor, containing schwannomas and neurofibromas. The transformation of benign vestibular schwannoma to MPNST has been reported formerly in seven cases (10). Loss of chromosomal arm 17q sequence, including complete inactivation of neurofibromatosis-1 gene, is supposed to be a multi-step and multi-gene process in the etiology of MPNST development (10). However, MPNST is rare in the head and neck region. Based on previous studies, nasopharynx, paranasal neck, sinus, nasal cavity, orbit, oral cavity, para pharyngeal region, neck, thyroid gland, and larynx are the most commonly affected site in head and neck (11).

The rate of incidence in MPNST is 0.001% in the general population. Principally, it occurs in adults:

Only 10% to 20% of MPNSTs are identified in the first 2 decades of life. Even though the nearly equal incidence has been

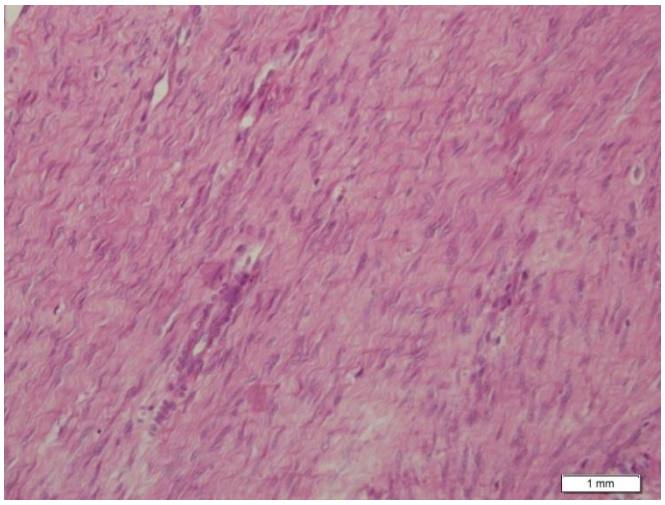

Figure 4. Hematoxylin and Eosin sections showed proliferation of malignant spindle cells arranged in fascicular pattern with infiltrating margins. The tumoral cells had comma and wavy nuclei, mild polymorphism, hyperchromatism and a few mitosis

reported between males and females, males suffered further than women from this disease because of the earlier development of local recurrences and metastases or a higher metastatic spread rate (9). The diagnosis of MPNST is challenging. A combination of gross, histopathologic, and immunohistochemical findings is required (12).

Immunohistochemistry participates a significant role in the diagnosis of MPNSTs. Rule out fibrosarcoma, synovial sarcoma, fibrous histocytoma, adenoid cystic carcinoma, neurogenic sarcoma, and chondrosarcoma would be possible with this method (13, 14). Responsiveness to the S100 protein and vimentin with focal positivity to CD68 and negativity to keratin demonstrated in Immunohistochemically study of the tumor cells (2).

Gross inspection of MPNSTs reveals a fusiform, fleshy, lobulated, or nodous, unencapsulated infiltrating mass with areas of degeneration and secondary hemorrhage. Even if the histomorphology of MPNST is various, this tumor type consists of spindle cells with nuclei hyperchromasia and numerous mitoses. Tumor cells are organized in sweeping fascicles, interlaced fascicles, or herringbone pattern. The marble-like pattern is often examined with an irregular arrangement of dense cells, fascicles, and hypodense cells. Occasionally tumor cells are arranged in storiform, whorled, paliform, or perithelioma patterns. Sometimes, irregular arrangement of round or short spindle tumor cells is detected. Uncommonly, epithelialization of adenoid and isolation of cartilage and bone were reported with tumor cells (2, 9, 15).

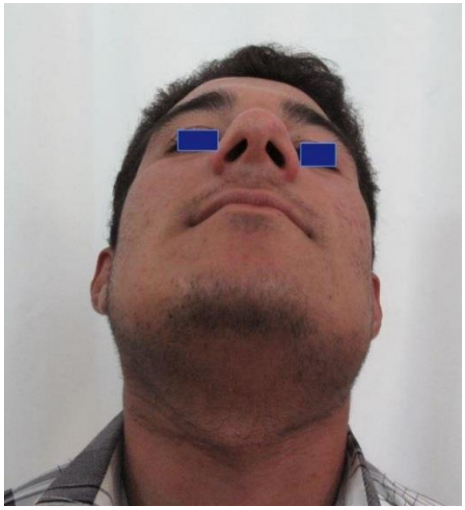

Figure 5. Patient's view five years after surgery

Treatment of MPNST is controversial. The strongest predictor of survival rate is complete surgical removal. Radical tumor excision with as wide a margin of healthy tissue even surrounding neurovascular, subcutaneous, and bony tissues due to their local invasion, hematogenous and rare lymph node metastasis is reasonable to achieve free histologic margins. However, routine neck dissection is not recommended (9, 15). Based on the risk of local and distant recurrence, residual disease after initial surgery, tumor size, and grade, adjuvant therapies have been suggested (15). Though chemotherapy , high dose of doxorubicin , and often radiotherapy are done as adjuvant/neoadjuvant treatments, their role is debatable (2).

Hematogenous metastasis takes place in at least half the cases (2, 16). The overall prognosis of MPNST is miserable. the literature survival rates are described to depend on patients' age, tumor size, location, the status of surgical margins, stage, grade, connection with NF1, and a history of past radiation therapy (9, 15). Generally, the survival rate was reported 40% to 70% (2). Consequently, MPNST has a distinctly worse prognosis than another soft tissue sarcoma (17).

Conclusion

MPNSTs are an uncommon extremely aggressive lesion in the Mandible, which could be challenging to treat despite considerable progress in treatment modalities. Surgical excision may be an essential treatment option to eradicate the lesion. However, local and distant recurrence lesions are expected.

Conflict of Interest: 'None declared'.

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