Case Report

Malignant peripheral nerve sheath tumor: A rarity

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Abstract

Malignant peripheral nerve sheath tumor (MPNST) of the mandible is an uncommon tumor that develops either from a preexisting neurofibroma or *de novo*. MPNSTs are sarcomas that originate from peripheral nerves or from cells associated with the nerve sheath, such as Schwann cells, perineural cells or from fibroblasts. Because MPNSTs can arise from multiple cell types, the overall appearance can vary greatly from one case to the next. A case of MPNST of the right side of the mandible in a 23-year-old female is reported.

Key words: Fibrosarcoma, malignant peripheral nerve sheath tumor, Schwann cells, spindle cells

INTRODUCTION

The principal malignancy of peripheral nerve origin is preferably called a malignant peripheral nerve sheath tumor.[1] MPNST is also termed as spindle cell malignancy, neurilemmoma, neurogenic sarcoma, neurofibrosarcoma.[2-3] It is an extremely rare malignancy that is encountered in patients with neurofibromatosis type I with an incidence of 2–5%, while in the general population it has an incidence of 0.001%.[4-6] MPNSTs are mainly located in the buttocks, thighs, brachial plexus and para-spinal region, and are rare in the head and neck region.[7,8]

CASE REPORT

A female patient of age 23 years reported to the Department of Oral Medicine and Radiology with a chief complaint of pain and swelling in the lower right side of the jaw since 1 month. Past history revealed that the patient had visited a dentist 6 months back with a complaint of pain, swelling and mobility of teeth in the same region. A panoramic radiograph was advised, and it showed a radiolucent lesion extending from the midline to the distal aspect of 46. The patient was advised a minor surgical procedure during which extractions and incisional biopsy were performed. The histopathological report revealed that it was a case of fibrosarcoma. The patient complained that there was recurrence of the swelling and pain in the same region since 1 month for which she was referred to our hospital. The pain was sharp, intermittent and localized in nature, aggravating while eating food and getting relieved on taking medication. The recurrent swelling was initially small in size and had attained the present size within 1 month.

On general physical examination, the patient was moderately built and nourished with vital signs within the normal limits. On examination, an extra-oral swelling was seen on the right side of the mandible that was extending from the midline to the right ramus of the mandible, superior-inferiorly from the corner of the mouth to 5 cm below the inferior border of the mandible, crossing the midline of size measuring 5 cm × 8 cm; margins of the swelling were well defined. On palpation, the swelling was hard in consistency, fixed to the underlying structures and tender. I/O examination showed missing 31, 32, 41, 42, 43, 44, 45 and 46 and a diffuse swelling in the right of the alveolus measuring about 6 cm × 2 cm, extending anteriorly from 31 to 47 posteriorly with obliteration of the buccal vestibule. On palpation, there was buccal cortical plate expansion that was nontender and firm. [Figure 1] Based on the history,

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past history and clinical examination, recurrent fibrosarcoma was given on the right side.

Further investigations were carried out. Panoramic, occlusal, cone beam computed tomography (CBCT), computed tomography (CT) and chest radiograph were advised. The panoramic radiograph revealed irregular shaped radiolucency, ragged borders on the right side of the mandible extending from the distal aspect of 32 to the mesial socket of 48, superoinferiorly from the crest of the ridge to the inferior border of the mandible, destruction of mental foramen and thinning of the inferior border of the mandible and missing 31, 32 and 41–47. The occlusal radiograph revealed pathologic fracture on the right side. To know the extent of the lesion, CBCT was advised. The CBCT revealed few bone islands with complete destruction of both buccal and lingual cortical plates. The CT scan revealed a large lobulated enhancing soft tissue density mass lesion measuring 5 cm × 3.7 cm × 6.3 cm seen on the body and ramus of the mandible, right buccal mucosa and gingivobuccal sulcus extending up to retromolar region. Few subcentimeter lymph nodes on the right side at level two were seen, with the largest measuring 6 mm × 7 mm [Figure 2]. To know the metastasis, a chest radiograph was advised, which was reported to be normal.

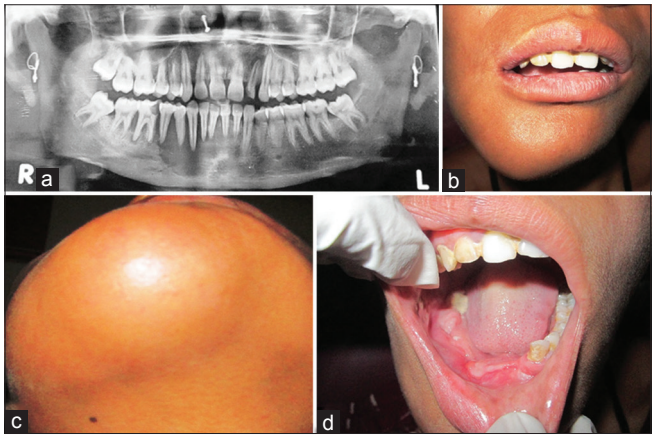

Figure 1: a) panoramic radiograph revealed irregular ragged borders on the right side of mandible, b) extra oral swelling on the right side of mandible, c) extra oral swelling involving submental region on right side, d) intra orally a diffuse swelling seen on the right side of the alveolus

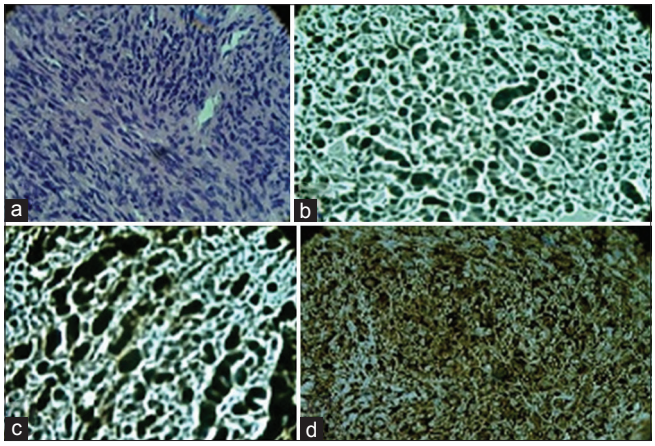

Figure 3: a) photomicrograph of H and E section showing spindle cells, b) NSE immunohistochemistry was positive, c) S-100 stain, d) vimentin stain

The patient was subjected to incisional biopsy. Histopathology showed highly cellular connective tissue stroma composed of a mixed population of cells, predominantly spindle cells, arranged in streaming fascicles. These findings were suggestive of spindle cell sarcoma. To confirm the diagnosis, an immunohistochemistry (IHC) investigation was performed. The tissue was strongly reactive for vimentin, [Figure 3] S-100 and nuclear specific enolase [Figure 3] and negative for cytokeratin and CD 34; hence, favoring the diagnosis for MPNST. The patient was referred to a regional oncology institute for further evaluation and management. Excision of the lesion was performed where hemimandibulectomy was performed and the specimen was sent for IHC investigation [Figure 4].

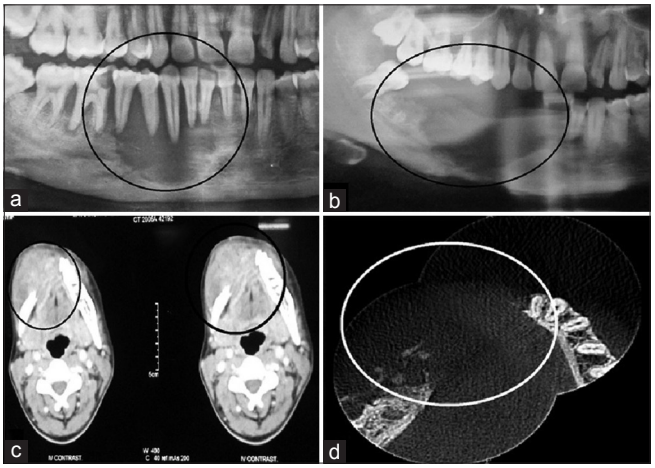

Figure 2: a) Panoramic radiograph revealing floating teeth appearance of 43.44 and 45, b) post operative panoramic radiograph revealing irregular, ragged borders on the lower right side of mandible, c) CT scan revealing soft tissue density mass on the right side of mandible, d) CBCT revealed few bone islands with complete destruction of both buccal and lingual cortical plates

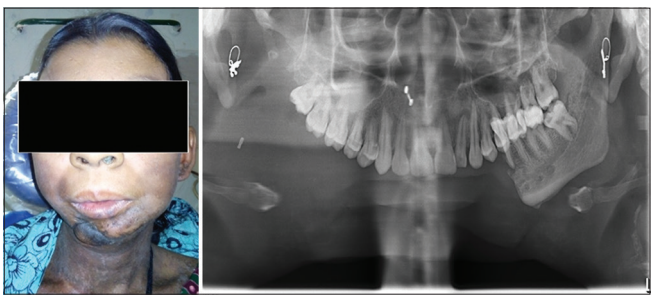

Figure 4: Post operative clinical photograph after hemimandibulectomy and radiation therapy with post operative panoramic radiograph

The excision biopsy report confirmed the diagnosis of MPNST. Compiling the information obtained by the radiological histopathological and IHC investigations, a final diagnosis of MPNST was arrived at.

DISCUSSION

A sarcoma is defined as a MPNST when at least one of the following criteria is met:

- It arises from a peripheral nerve
- It arises from a preexisting benign nerve sheath tumor (neurofibroma)
- It demonstrates Schwann cell differentiation on histologic examination.[9]

MPNST is an uncommon sarcoma with an incidence of 1:100,000/year, compromising 5–10% of all soft tissue sarcomas (STS). MPNST originates from or recapitulates the phenotype of peripheral nerves cells, such as Schwann cells or perineural cells. Although the histogenesis of MPNST remains unclear, there is a higher incidence in patients with prior radiation exposure and neurofibromatosis type 1 (NF1), who have a lifetime risk of 10% of developing MPNST. MPNSTs typically arise in the extremities (40%), followed by the trunk/retroperitoneal (38%) and head and neck regions (21%). Most MPNSTs are biologically high-grade sarcomas that tend to recur (40–65%) and metastasize (40–80%). MPNSTs usually metastasize hematogenous, most commonly to the lungs.[10]

Most of the MPNSTs occur in the age group of 20–50 years, with an equal predilection to males and females.[8,9] Usually, MPNSTs are found in the extremities and the trunk and, unlike benign Schwannomas, are seldom found in the head and neck area.[11] Oral tumors may occur anywhere, but the most common sites are the mandible, lips, buccal mucosa, paranasal sinus, nasal cavity, orbit, cranial nerves, larynx, parapharngeal or pterygomaxillary space, minor salivary glands and the thyroid gland.[1,8] The present case comprises an extremely unique presentation of this malignancy involving the mandible on the right side in a 23-year-old female. Clinically, they present as an enlarging mass, often associated with pain and nerve deficit.[12]

CT is useful in assessing the tumor extension and eventual metastasis (the more frequent are bone and lung metastasis). Magnetic resonance imaging (MRI) can reveal the nerve of origin, and it is more accurate to evaluate the topographical relationship of the tumor with the neighboring structures, especially the vascular and muscular structures and fat planes involvement. In particular, MRI distinguishes the lesion from the fat tissue better than CT, whose dislocation and thinning of the fat tissue thickness has a critical importance in localizing the space of origin of a neck lesion.[4] Fine needle aspiration, or FNA, is a biopsy method employed to obtain individual cells for cytologic review. It can be performed with a very small needle, which is more easily tolerated by the patient and is often useful to establish the presence of malignant cells. A second type of biopsy is a core needle or tru-cut needle biopsy, which uses a larger hollow-bored needle gauge to obtain a more substantial tissue sample. This type of sample offers inspection of both individual cells as well as the architectural arrangement of those cells within a given part of the tumor mass.[9] Approximately 80–85% of MPNSTs are spindle cell tumors with fasciculating patterns that contain histologic features similar to those of a fibrosarcoma. They are often high-grade, demonstrating four or more mitotic figures per high-powered field. The remaining 15% of MPNSTs is composed of tumors that exhibit variable differentiation, allowing them to be sub-classified as distinct entities. A MPNST with rhabdomyoblastic differentiation is characterized by both neural and skeletal muscle differentiation. Within this category is the malignant triton tumor, which refers specifically to a MPNST occurring in association with rhabdomyosarcoma. Other examples of MPNSTs with differentiation include glandular malignant Schwannoma, epithelioid malignant Schwannoma and superficial epithelioid MPNST.[9] In 50–70% of the MPNSTs, scattered tumor cells express S-100 protein. S-100 positivity is not seen in other spindle cell sarcomas. Also, some cases of MPNST stain positive for neuron-specific enolase.

In our case of MPNST, the tissue was strongly and diffusely positive for Vimentin, S-100 and neuron-specific enolase.[8]

The mainstay of treatment is surgical resection. The goal of the operation is to achieve complete surgical excision of the tumor with negative (wide) margins. This offers the best outcome with respect to both local recurrence and distant metastases.[9] Radiation therapy can be employed in preoperative, intraoperative and postoperative settings. Treatment with adjuvant radiotherapy has yielded a statistically significant reduction in the rates of local disease recurrence. Studies have shown the average 5-year survival rate for these patients ranges from 16% to 52%.

CONCLUSION

MPNSTs are very infrequent diseases of the head and neck region and in young patients without neurofibromatosis I syndrome. In our case, MPNST was seen in a young female patient involving the mandible

without neurofibromatosis I syndrome, which is said to be rare as stated in the literature. A combination of clinical, pathological and immunohistochemistry helps in diagnosing these tumors. Although a multimodality therapy, including surgical resection and adjuvant radiotherapy, is available, the prognosis remains dismal. Modern clinical studies and the development of effective targeted chemotherapy are needed to gain control of the disease.

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