Case Report

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Dentistry Section

# Malignant Peripheral Nerve Sheath Tumor -A Rare Malignancy in Mandible

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## ABSTRACT

Malignant Peripheral Nerve Sheath Tumor (MPNST) is biologically an aggressive tumor that is usually found in the extremities, trunk and infrequently found in the head and neck area particularly in the jaws, arising from the cells allied with nerve sheath. Mandibular MPNST may either arise from a preexisting neurofibroma or develop de novo. Because of the greater variability from case to case in overall appearance both clinically and histologically, a case of MPNST of the mandible in a 25-year-old female patient is reported. The lesion was excised and immunohistological studies (S-100 & Neuron specific enolase) were conducted to confirm the neural origin.

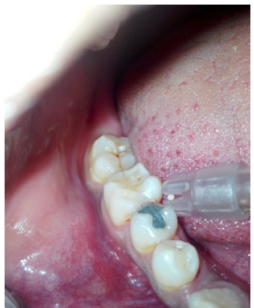
**Keywords:** Immunohistochemistry, Neurofibrosarcoma, Neuron specific enolase

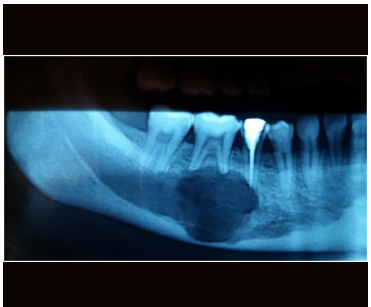
## CASE REPORT

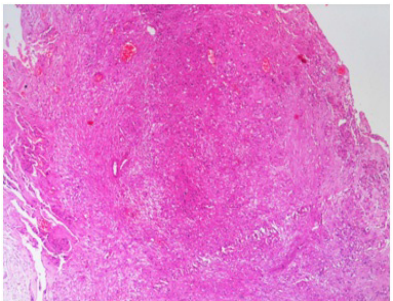
A 25-year-old female patient reported to the Department of Oral Medicine and Radiology with a chief complaint of pain in her lower right back tooth region since three months. Pain was sharp, sudden in onset and intermittent in nature. Past dental history revealed that the patient had visited a dentist five years back for the same complaint of pain in the same region for which she had undergone root canal treatment. There was no marked abnormality in facial symmetry on extra oral examination and lymph nodes were not palpable. On intraoral examination, there was no marked swelling intraorally [Table/Fig-1]. On palpation, there was mobility of teeth associated with tenderness. Percussion revealed mild tenderness over root canal treated tooth (right second premolar). A panoramic radiograph showed ill-defined, unilocular radiolucency with irregularly ragged borders measuring about 3×3cms in size extending from the apices of the right premolar and first molar teeth to the lower border of the mandible along with the root resorption of these teeth [Table/Fig-2]. The inferior alveolar canal could not be traced. The lower border of mandible was thinned out, but intact. Based on both clinical and radiological findings the provisional diagnosis was made as radicular cyst (arising from second premolar), whereas the differential diagnosis was made as primary intraosseous squamous cell carcinoma, desmoplastic fibroma, fibrosarcoma and metastatic tumor of the jaw bone. Later the patient was advised a minor surgical procedure during which extraction of involved tooth and incisional biopsy from the lesional site were performed after all the necessary routine hematological investigations. The Hematoxylin and Eosin stained soft tissue section showed loose connective tissue with hypo and hypercellular areas and proliferation of atypical spindle-shaped cells in streaming fascicles [Table/Fig-3,4]. Under 40X view pleomorphic cells with wavy, comma-shaped nuclei and hyperchromatic nuclei were seen [Table/Fig-5, 6]. Mast cells and many blood capillaries were also evident in the connective tissue [Table/Fig-6]. Foci of necrosis were also seen. Based on these findings histopathological diagnosis was made as "Neurofibrosarcoma". Final diagnosis of neural origin is often difficult especially in the absence of neurofibromatosis and without any history of neurofibroma. Immunohistological (IHC) studies were conducted to confirm the neural origin by S-100 and Neuron Specific Enolase (NSE). The section was strongly positive for S-100 and NSE [Table/Fig-7,8]. Wide excision of the lesion with negative margins was done. The excision biopsy also suggested MPNST. There was no recurrence till date and the patient is still under follow-up with frequent dental visits.

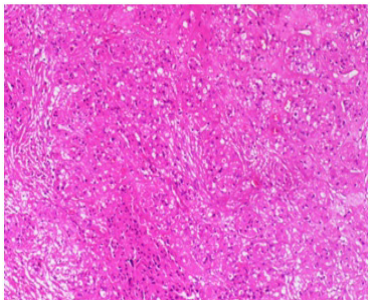
## DISCUSSION

Malignant peripheral nerve sheath tumors (MPNSTs) are a variety of tumors that originates from or recapitulates the phenotype of peripheral nerves cells, such as schwann cells, perineural fibroblasts, or fibroblasts [1]. MPNSTs accounts for about 5% of all soft tissue sarcomas and arises in nearly one –fourth to one –half of Neurofibromatosis 1(NF1) [1]. Rarely, MPNSTs develop post-radiotherapy [2]. Usually the MPNSTs are found involving the extremities and the trunk, unlike benign Schwannomas, are infrequently found in the head and neck area [1].

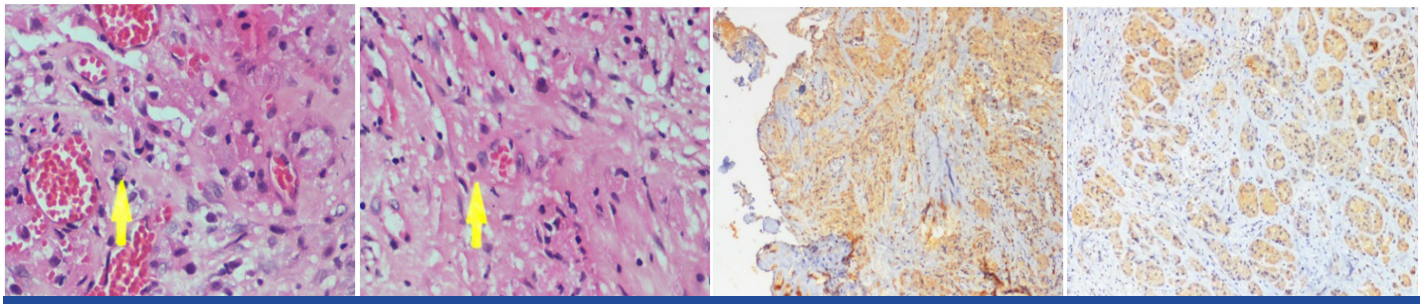








**[Table/Fig-1]:** Clinically there was no marked or diffuse intra oral swelling in relation to 44, 45 tooth regions. **[Table/Fig-2]:** Panoramic radiograph suggesting an ill defined unilocular radiolucency with irregular ragged borders along with resorption of the premolar and molar tooth roots. **[Table/Fig-3]:** The Hematoxylin & Eosin stained soft tissue section showing the connective tissue with hypo and hypercellular areas. (4X view of light microscope). **[Table/Fig-4]:** The Hematoxylin & Eosin stained section shows proliferation of atypical spindle shaped cells in the form of streaming fascicles. (10X view of light microscope).



**[Table/Fig-5]:** In 40X view of light microscope shows pleomorphic cells with hyperchromatic and atypical nuclear features like comma shaped nuclei, vesicular nuclei, and few blood capillaries. **[Table/Fig-6]:** 40X view showing neoplastic cells with wavy & comma shaped nuclei and mast cells. **[Table/Fig-7]:** 10X view showing immunohistochemical positive staining for S-100. **[Table/Fig-8]:** 10X view showing immunohistochemical positive staining for NSE.

The primary malignancy of peripheral nerve origin is preferably called a MPNSTs, also known as spindle cell tumor, neurilemmoma, neurogenic sarcoma and neurofibrosarcoma [3,4]. Mostly these MPNSTs occur in the age group of 20–50 years, with an equal male and female sex ratio [5,6]. The lesion presents mostly on the proximal areas of the extremities and the trunk and only 10% to 15% of cases occur in the head and neck region [3]. Oral tumors may occur anywhere, but the most common sites are the mandible, lips and buccal mucosa [3,5].

Microscopic evaluation of MPNST shows fascicles of atypical spindle-shaped neoplastic cells, which often resemble the cells of fibrosarcoma [3]. However, these neoplastic cells are more irregular with wavy or comma-shaped nuclei. In addition to streaming fascicles, less cellular myxoid areas also may be present. A confirmed diagnosis to state such lesions to be of neural origin is often difficult, especially in the absence of neurofibromatosis. Positive immunostaining for S-100 protein, NSE is a helpful diagnostic aid, but this is found in only about 50% of all cases [1, 7]. In the present reported case along with all classical histological features of MPNST positive staining of the tumor cells with NSE marker and S100 were noticed.

MPNSTs express as a group of highly heterogeneous human malignancies frequently with various multiple histological origins, peculiar differentiation patterns and diverse immunohistochemical presentations. Epithelioid or other heterogeneous components can be experiential in 15% of MPNSTs, the latter include other differentiations like rhabdomyoblasts, cartilaginous, osseous and rarely smooth muscle, glandular or lipoarcomatous components have been reported. It is rare that there are two or more heterogeneous components in a single MPNST [8]. However these types of differentiation patterns were not found in the present case.

The present case report is unique because of its unusual clinical presentation and rare anatomical site. There was no associated swelling, the regional teeth were mobile with resorbed roots, was non tender. The occurrence of this lesion in a young female is also a rarity, especially in the absence of Neurofibromatosis Syndrome Type 1(NF1) [2].

Recent literature has recorded few such cases in young females with MPNST in mandible [9,10]

## CONCLUSION

MPNST 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 mandibular molar and premolar teeth and without severe pain and paresthesia. Since this malignancy can mimic the clinical features of any benign lesion, dentists must be careful in giving the final diagnosis and treatment.

**Ethical approval:** Yes.

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