

# Malignant Peripheral Nerve Sheath Tumor of Buccal Mucosa: An Oncological Surprise

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Received September 08, 2014; Revised October 10, 2014; Accepted October 20, 2014

**Abstract** Malignant peripheral nerve sheath tumors are aggressive sarcomas with poor prognosis. It is usually encountered in lower extremities. Only a few cases have been reported in head and neck region with buccal mucosa being an unusual site.

**Keywords:** malignant peripheral nerve sheath tumor, radiation, surgery

**Cite This Article:** Ram Abhinav Kannan, Kirthi Koushik, Usha Muniyappa, Ritika Harjani, and Arvind Murthy, "Malignant Peripheral Nerve Sheath Tumor of Buccal Mucosa: An Oncological Surprise." *American Journal of Medical Case Reports*, vol. 2, no. 10 (2014): 222-224. doi: 10.12691/ajmcr-2-10-6.

## 1. Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are spindle-cell sarcomas which are aggressive tumors accounting for 5–10% of all soft tissue sarcomas. These tumours are usually found in the lower extremities, with retroperitoneum being next most common site [1]. Only 8–16% of MPNSTs develop in the head and neck region. The oral cavity is an unusual site for this tumor [2]. In this paper, we report a case of MPNST arising from the buccal mucosa along with available review of literature.

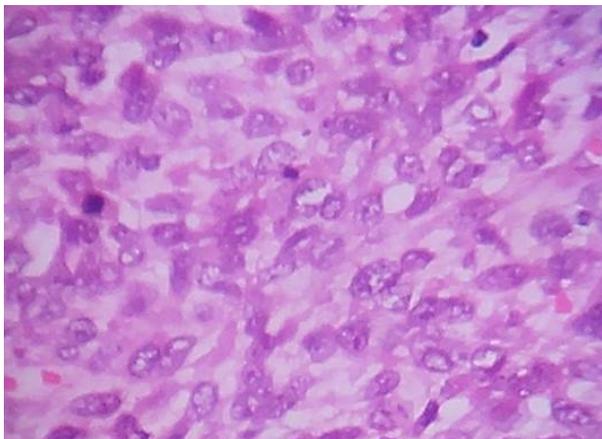


**Figure 1.** Heterogeneously enhancing lesion in left buccal mucosa, medially abutting the tongue and posteriorly abutting the submandibular gland and masseter

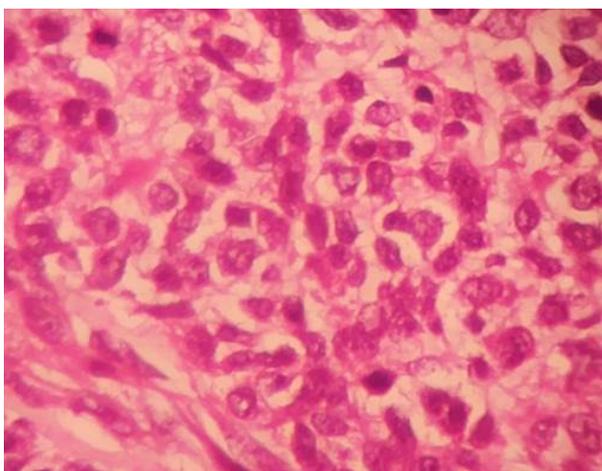
## 2. Case Report

A forty year old female presented with complaints of swelling in left side of her mouth that gradually progressed over 3 months. She also gave history of mass in the same region two years back, for which she underwent left mandibular resection along with plate reconstruction, which was diagnosed as ameloblastic fibroma on post-operative histological examination. Examination of oral cavity revealed a 3X3 cm mass in the left buccal mucosa with extension to left lateral border of tongue. Computed tomography scan (CT scan) with intravenous contrast revealed a heterogeneously enhancing lesion in the left buccal mucosa measuring 3.7 X 3.3 X 3.1 cm (Figure 1). The lesion was abutting the tongue medially with mild asymmetrical enhancement of the tongue and posteriorly, the lesion was abutting the submandibular gland and masseter. A lymph node measuring 1 X 1 cm was noted in the left submandibular region. Biopsy of the tissue mass showed tumor cells which are predominantly round to oval with few spindle shaped cells having large vesicular nuclei and moderate to scanty eosinophilic cytoplasm with numerous mitotic figures, suggestive of sarcoma or a poorly differentiated carcinoma (Figure 2, Figure 3). Immunohistochemistry revealed tumor cells strongly positive for vimentin, S-100 and are negative for pan-CK, CD 34, desmin and smooth muscle actin (Figure 4). The histopathological examination and immunohistochemistry was consistent with the diagnosis of malignant peripheral nerve sheath tumor. CT scan thorax and ultrasound of the abdomen was negative for distant metastasis. There was no history suggestive of neurofibromatosis. Patient underwent extended hemi-mandibulectomy and ipsilateral neck node dissection under general anaesthesia. The resected tumor measured 4.5 X 4 X 2.5 cm with posterior, medial and inferior mucosal margins being involved by the tumor.

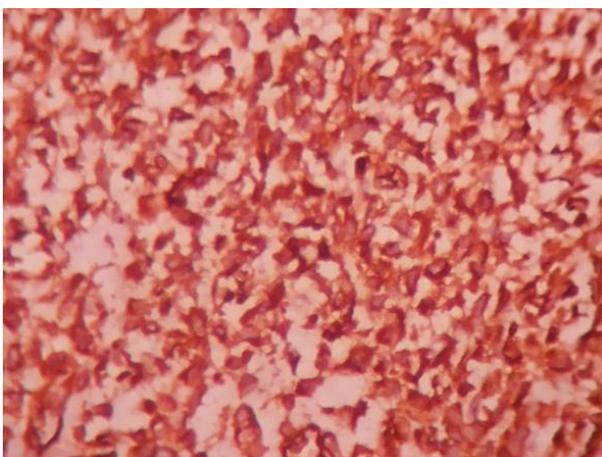
Dissected lymph nodes were negative for malignancy. The histopathological examination was consistent with the pre-treatment diagnosis of malignant peripheral nerve sheath tumor. In view of positive surgical margins, patient received post-operative local radiation to a dose of 66Gy in 33 fractions using three dimensional conformal radiation therapy.



**Figure 2.** Spindle shaped tumor cells with elongated vesicular to hyperchromatic nuclei



**Figure 3.** Spindle shaped tumor cells with mitotic figures



**Figure 4.** Diffuse strong positivity for S-100 protein

### 3. Discussion

Neurogenic tumors include malignant and benign variants; benign group constitute neurofibroma, schwannoma, neuroma

and perineuroma; malignant group constitutes malignant peripheral nerve sheath tumors [3,4]. Previously, various terminologies like neurogenic sarcoma, neurilemmosarcoma, malignant fibrosarcoma and malignant neurilemmoma had been used, but World Health Organization (WHO) has recently adopted the term ‘malignant peripheral nerve sheath tumor’. MPNST are sarcomas with one of the following features: (1) arising from a peripheral nerve (2) arising from a pre-existing benign nerve sheath tumor (3) which demonstrate schwann cell differentiation on histologic examination. These tumors share a common neural origin but present microscopic and clinical heterogeneity. Their occurrence in the oral cavity is extremely rare with most common sites in it being mandible, lips and buccal mucosa [2].

About 40-50% of MPNSTs are associated with a family history of neurofibromatosis-1 (NF-1) [1]. It is generally accepted that MPNSTs occur in about 2–5% of NF1 patients compared with an incidence of 0.001% in the general population [5]. The tumour commonly occurs in the age group between 20-50 years with an equal male and female predilection [6].

Microscopically the tumour consists of spindle cells with a high mitotic rate and indistinct cytoplasmic borders arranged in bundles or fascicles. The diagnosis of MPNST is difficult and elusive in the soft tissue disease due to lack of standardized criteria. It is not always possible to demonstrate the origin from a nerve, especially when it arises from a small peripheral branch. Immunohistochemistry plays an important part in the diagnosis, with tumor cells showing specific positivity for S-100 [7,8,9,10].

Common routes of spread are through direct extension, hematogenous extension and by perineural spread. Lymph node metastasis is rare. Treatment is predominantly surgical. Complete surgical excision of the tumor with negative margins offers the best outcome with respect to both local recurrence and distant metastases. Complete resectability rates are determined primarily by anatomic location of the tumor [5,11,12].

Radiation therapy has been tried in preoperative, intraoperative, and postoperative settings. Addition of postoperative radiation therapy has yielded a statistically significant reduction in the rates of local disease recurrence, but not improvement in overall survival [13]. The value of chemotherapy has not been firmly established and is usually considered for patients with large tumor size (> 5 cm), unresectability or metastatic disease.

MPNSTs are locally invasive lesions, frequently leading to multiple recurrences and eventual metastasis. Its local recurrence rate ranges from 40% to 42%. Most common metastatic sites are lungs, followed in decreasing order of frequency by soft tissue and bone [14,15]. Prognosis is poor and survival is found to be influenced by tumor location, size and association with NF-1.

Typical MPNST has been reported to have a 5-year survival rate of 34–39% [15]. The malignant transformation of a neurofibroma has an extremely poor prognosis with prevalent recurrences and distant metastasis [5,11,12]. Favourable prognostic factors include tumor size < 5 cm, lack of local recurrence, low histologic grade and extremity location [16]. Low intensity p53 staining and positive S-100 staining is also found to be associated with better outcomes [17].

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