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**Case Report**

# Leiomyosarcoma of mandible-a rare case report with review of literature

**Nupur Rastogi[1]*, Kaushal Gautam[2], Sunidhi Rohatgi[2], Anushree Khandelwal[2], Priya Rajawat[2]**

[1]Diagnostic Centre, Sudha Medical College, Kota, Rajasthan, India
[2]Sudha Medical College, Kota, Rajasthan, India

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***Correspondence:**
Dr. Nupur Rastogi,
E-mail: nupurkota123@gmail.com



## ABSTRACT

Sarcomas are rare group of malignant mesenchymal tumors, about 1% of all malignancies in adults, leiomyosarcoma constitute 3-10% of all sarcomas in the head and neck region, of them only 0.64% are in oral and maxillofacial region, hence qualifying to be an extremely rare malignancy due to rarity of smooth muscle in that area. Till date approximately 65 cases from 1950-2011 were identified and 20 cases from 2012 till 2024 (as per google search) have been described. We present a case of 51-year-old female who presented with polypoidal growth in right lower alveolus without any lymph node involvement. On histomorphological features and immunohistochemistry, diagnosis of leiomyosarcoma was confirmed.

**Keywords:** Right lower alveolus, Leiomyosarcoma, Primary

## INTRODUCTION

Sarcomas are rare group of malignant mesenchymal tumors, accounting for about 1 % of all malignancies in adults. Leiomyosarcomas constitute 3-10 % of all sarcomas in the head and neck region, they are mostly seen in gastrointestinal tract, bladder, retroperitoneal space.[1] Leiomyosarcoma are only 0.64 % cases in oral and maxillofacial region, extremely rare in mouth and jaw due to rarity of smooth muscles in that area i.e. in the vessel wall, erector pilli muscles, circumvallate papillae, myoepithelial cells of salivary glands.[2-4]

From 1950 -2011, only 65 cases of leiomyosarcoma located in the mandible were described.[5] From 2011 to 2024 till date 20 cases were described. It most frequently occurs in elderly with sex ratio of 11:9 male:female. Etiological factors include predisposing disease (chronic lymphedema, disease accompanied with immunosuppression, viral disease). Environmental factors (radiation, trauma, foreign body, chemical compounds, herbicides, pesticides) and genetic disorders (neurofibromatosis, Gardner's syndrome, Li-Fraumeni syndrome).[6] In the present case, female aged 51 years presented with mass in oral cavity of 2 months duration, painless. On H&E and immunohistochemistry, the tumor was diagnosed to be primary leiomyosarcoma of right lower alveolus.

## CASE REPORT

A 51 years female presented with growth in right lower alveolus region. A biopsy was taken which was reported as Well differentiated squamous cell carcinoma elsewhere. Patient was sent for PET CT scan before doing extensive surgery. It showed a large FDG avid mass in right lower gingiva-buccal sulcus with large erosion of mandible in the region of incisor, canine, premolar, measuring approximately 30×40×36 mm. Minimal extension to right side floor of mouth was seen. There was no involvement of upper gingivobuccal sulcus, buccinator, masseter, upper alveolus, RMT, maxilla, carotid vessels. Approximately

8×10 mm, 8 right level I B lymph node were seen. No other FDG avid foci was seen elsewhere in scanned region. Right partial mandibulectomy with resection of right and left cervical nodes was done and the specimens were sent for histopathological examination.

Gross examination of Right partial mandibulectomy specimen with 5 teeth attached, showed a greyish white polypoidal growth in right lower alveolus measuring 5.5×4.5×3.2 cm (Figure 1,2). Cut surface was firm, greyish white, with whorled appearance. On the right side 25 lymph nodes and on left side 9 lymph nodes were identified and subjected for fixation and processing.

The microscopic examination on H&E-stained slides from polypoidal mass showed stratified squamous epithelium with underlying tissue about 0.5 cm from basal layer showing proliferation of spindle shaped cells with plump cigar shaped nucleus, showing moderate degree of pleomorphism, eosinophilic cytoplasm, arranged in fascicular pattern, multinucleated bizarre tumor giant cells. Mitotic activity was 5-6/HPF. Pushing borders was seen at the periphery of tumor. Necrosis was seen (Figure 3,4,5).

All margins were free of tumor. Underlying bone showed presence of tumor. All lymph nodes showed features of Sinus Histiocytosis. Based on histomorphological features a diagnosis of pleomorphic sarcoma, leiomyosarcoma was suggested. To confirm the histological diagnosis, Immunohistochemistry was done which showed positivity for SMA (Figure 6) and Vimentin (Figure 7). The tumor showed negativity for Desmin (Figure 8) S-100 (Figure 9) CD-34, HMB-45, CK, H-caldesmin.

Hence, histomorphological and immunohistochemistry confirmed the diagnosis of primary leiomyosarcoma of mandible.

**Figure: Cut surface showing greyish white surface with whorled appearance.**

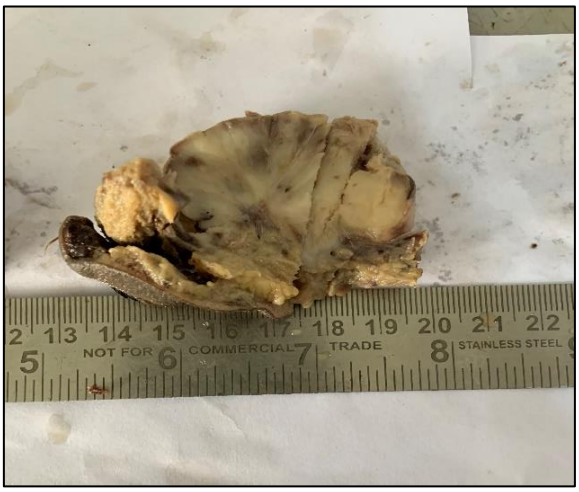


**Figure 2: Cut surface of tumor.**

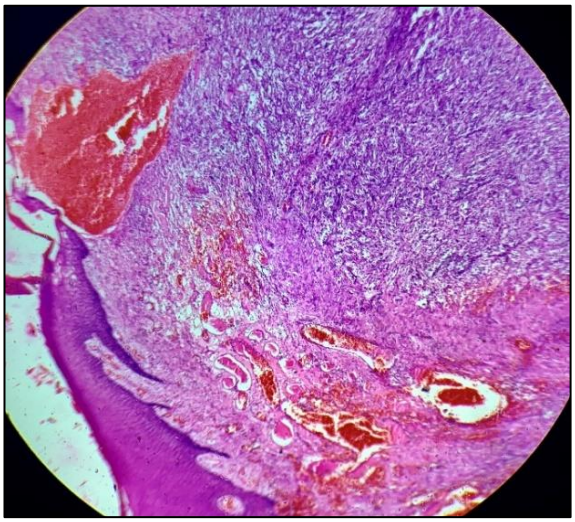

**Figure 3: H & E, 10X stratified squamous epithelium with underlying tissue showing proliferation of tumor cells.**

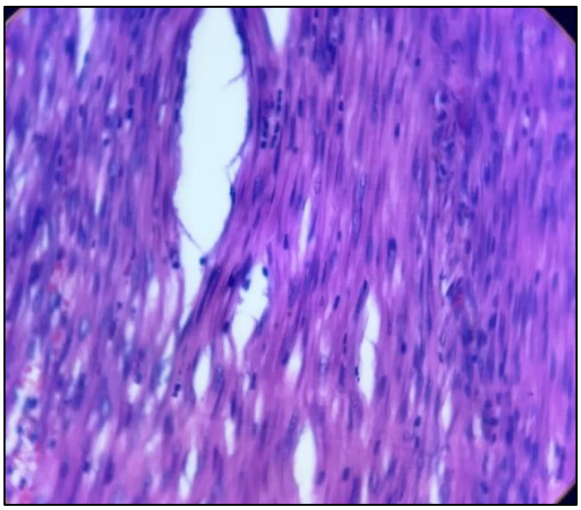

**Figure 4: H & E, 40X Fascicles of smooth muscle with cigar shaped nucleus.**

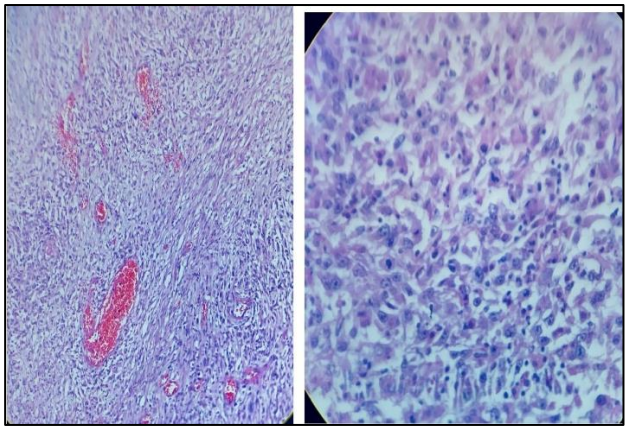

**Figure 5: H & E, 40X, showing vague fascicular pattern, mitotic figures.**

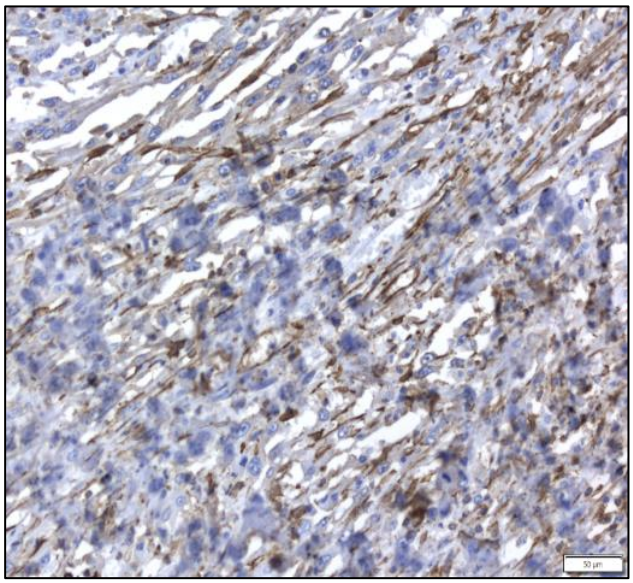

**Figure 6: 10X, SMA positive.**

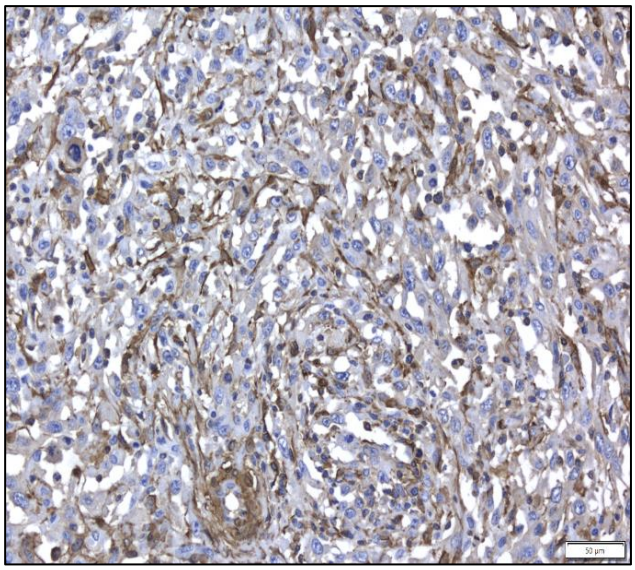

**Figure 7: 10X, Vimentin positive.**

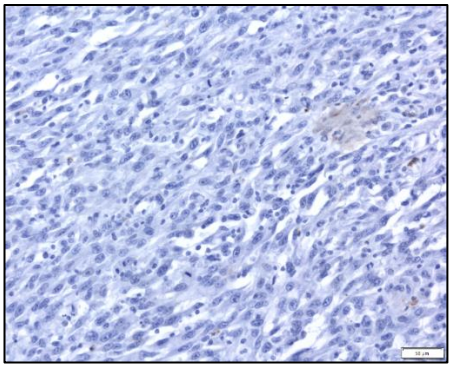

**Figure 8: 10X, Desmin negative.**

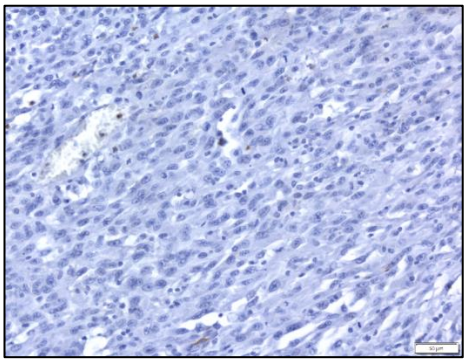

**Figure 9: 10 X, S-100 negative.**

## DISCUSSION

Leiomyosarcoma is a malignant neoplasm originating in smooth muscle that occurs most frequently in the uterus and gastro intestinal tract. Fernandez et al, reported that although leiomyosarcoma accounts for approximately 7 % of all soft tissue sarcomas, it is extremely rare in the field of oral and maxillofacial region. According to literature, only 0.64 % of cases were reported in the oral area due to rarity of smooth muscles in oral cavity. The first case of primary LMS of the mandible was reported by Carmody et al, in 1944.[7] Generally primary LMS of the jaw are considered to arise from smooth muscle cells of the tunica media of vasculature, erectores pilorum, circumvallate papillae or from pluripotent mesenchymal cells.[8] Age group varies from 16-66 years, our case is of 51 years female which falls in this age group.[9]

Intraoral LMS is an extremely uncommon neoplasm, about 85 cases of LMS of mandible from 1950 till 2024 have been reported in the literature. The intraoral locations of these tumours are- cheek, mandible, gingiva, maxilla, floor of mouth, tongue, soft and hard palate mucosa.[10] Jaw bones appear to be a site of predilection for oral LMS and approximately 50 % of these tumours arise in the jaw, predominantly mandible.[11] Mandibular canal may be the site of origin in some cases of mandibular involvement.[12] Clinically sarcomas of the head and neck present with non-specific signs and symptoms, and in majority of cases these tumors manifest as a painless mass.[13,14] Distant metastases can occur in 28 % of patients at diagnosis or

during follow-up and a more frequent in high grade sarcomas. The most frequently involved site for distant metastases is lung followed by bone, CNS and liver.[15]

In the present case from right lower alveolus, microscopic findings showed spindle shaped cells with cigar shaped nucleus in fascicular pattern, bizarre tumor giant cells, pleomorphism, necrosis was seen. Immunohistochemical markers were positive for SMA, Vimentin and negative for Desmin, CD-34, HMB-45, CK, H-caldesmon. Diagnosis of pleomorphic sarcoma (leiomyosarcoma) was made combining the histopathological and immunohistochemical findings.

Based on the microscopic findings Differential diagnosis were made. Metastatic Leiomyosarcoma to oral cavity from other primary sites of which lung is the most common.[16] Oral Leiomyosarcomas tend to metastasize to cervical nodes, lung and liver, unlike LMS in other soft tissue which rarely show nodal involvement.[17]

Malignant spindle cell tumor including spindle cell carcinoma, melanoma, Leiomyosarcoma, adult spindle cell rhabdomyosarcoma (has more myxoid stroma), myofibroma, MPNST (shows fascicles of atypical spindle shaped cells with wavy nuclei.) Spindle cell carcinoma is high grade malignancy arising from superficial epithelial layer. Malignant melanoma was excluded as the tumor was not a mucosal lesion.

Myofibroma of the jaw is more common in first decades of life and has biphasic appearance in histopathology. Immunohistochemistry is confirmatory for diagnosis as LMS which is positive for SMA and Desmin. Negative for S 100, CK, myogenin, Vimentin. P53 is associated with high rate of recurrence and shorter survival. Prognosis of LMS is usually poor.[8] Wide surgical excision with radical neck dissection for Lymph node metastases is the mainstay of treatment. Because it has high rate of recurrence and metastases, long term regular follow up is necessary.

## CONCLUSION

As leiomyosarcoma of oral cavity is an extremely rare entity, accurate diagnosis is helpful in proper treatment (chemotherapy, radiotherapy) as the prognosis is poor. Diagnosis is made by histomorphological features and Immunohistochemistry. This case is presented as it is important to keep differential diagnosis of Leiomyosarcoma in any malignant lesion with spindle cell lesion.

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