CASE REPORT

MASSIVE LEIOMYOSARCOMA OF THE MAXILLA

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Leiomyosarcoma is a malignant smooth-muscle tumour that is exceedingly rare in the head and neck region. Presenting signs and symptoms of leiomyosarcoma in the orofacial region are generally non-specific. The tumour is commonly encountered as a slow growing, discreet firm, and non-ulcerated painless mass. We presented a case of massive leiomyosarcoma of the midface which might be the first leiomyosarcoma of midface reported in Pakistan so far to our knowledge. We also discuss the diagnosis and treatment of leiomyosarcoma in this aspect.

Keyword: Leiomyosarcoma, tumour, malignant, orofacial

INTRODUCTION

Leiomyosarcoma is a malignant tumour of mesenchymal origin exhibiting smooth muscle differentiation. Only 3–10% of superficial leiomyosarcomas arise in the head and neck. This reflects the paucity of smooth muscle found in this region. Smooth muscle is derived from primitive mesenchyme and is found mainly in blood vessels, erector pile musculature of skin, circumvallate papilla, primitive mesenchyme and myoepithelial cells of salivary glands.

Reported sites of occurrence are larynx, hypopharynx, tongue, trachea, floor of the mouth, gingiva, soft palate, parapharyngeal space, lips and thyroid.[1] The commonest intraoral site appears to be the tongue.[2] Youngest reported case is 1 year old and the oldest 88 years of age.[4] Higher incidence is supposed to occur among the middle age or elderly.[1] Presenting sings and symptoms are generally non-specific, usually painless. Discolouration, ulceration, umblication and overlying skin changes can be found.[2] Epistaxis, dysphagia, hoarseness, fever, stridor and cough have also been reported.[4]

Microscopically, cells are spindle shaped with blunt ended nuclei, growing in interlacing cords.[5] Nuclear palisading and myofibrils are recognisable. Anaplastic features of large, bizarre, pyknotic nuclei and mitotic figures are seen in varying degrees.[5] Other histologic variants such as leiomyosarcoma with giant cells[8], pleomorphic leiomyosarcoma[9], inflammatory leomyosarcoma[10], granular cell leiomyosarcoma[11], epithelioid leiomyosarcoma[4] and desmoplastic leiomyosarcoma[3] have been identified. Immuno-histochemical identification is essential for an accurate histologic diagnosis. To avoid misdiagnosis it is advisable to use a broad panel of antibodies including actins (SMA and HHF-35), desmin, vimentin, cytokeratins and S-100 proteins.[2]

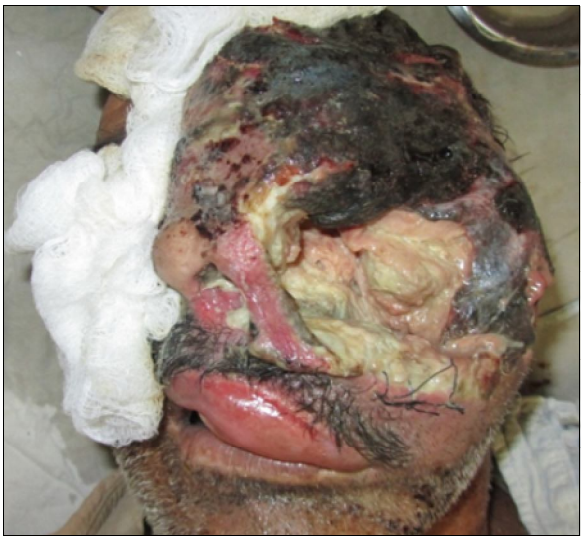
Recommended treatment for leiomyosarcoma is primarily surgical consisting of wide local excision. Adjuvant external beam therapy is indicated in some cases, no clear survival benefit has been reported with the use of chemotherapy.[2]

CASE REPORT

A 65-year-old man presented with a 8-month history of huge swelling involving the midface. According to the patient he was alright 8 months back when he noticed a small swelling on left side of his midface. He ruptured it with hand but it again reappeared. He remained under the treatment of some local hakeems but the growth keep on increasing in size rapidly. After 8 months patient presented to the outpatient department of Mayo Hospital with the huge swelling arising from the midface (Figure-1). The mass with the dimension of approx. 35cm into 25cm was fungating with large area of necrosis involving most of the left side of face sparing only lower third of the face extending from superior orbital margin to the body of mandible. Anteroposteriorly it extended from the nose to the preauricular region. There was no palpable cervical lymphadenopathy. On intra oral examination there was a hard swelling on the palate crossing the midline. At the time of presentation the patient was malnourished and dehydrated. He was immediately admitted to the maxillofacial ward and IV support was given. Incisional biopsy and baseline investigations were done immediately. Histopathology evaluation suggested the lesion to be the leiomyosarcoma. Actin was positive in the neoplastic spindle cells with cytokeratin, desmin, S-100 and CD-34 were negative.

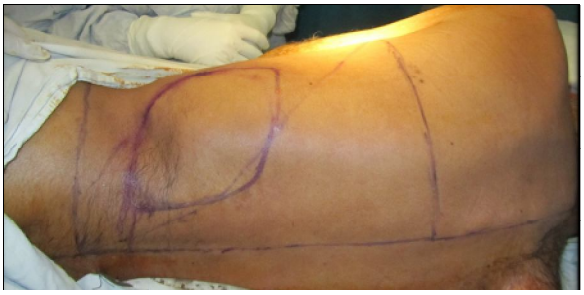
Contrast enhanced computed tomographic (CT) examinations demonstrated an ulcerated soft tissue density enhancing mass on the left cheek with extension into left maxillary sinus, nose, oral cavity, orbit and forehead. No cervical lymphadenpathy was present on CT examination.

The patient subsequently underwent a workup for metastasis, which resulted negative. Excision with 2cm safe margins and subtotal maxillectomy was performed. Orbital exenteration was also done. The defect was reconstructed with lattismus dorsi myocutaneous pedicled flap. There was no complication during the surgery and the recovery was uneventful. The patient is now referred to the oncology department for further management.

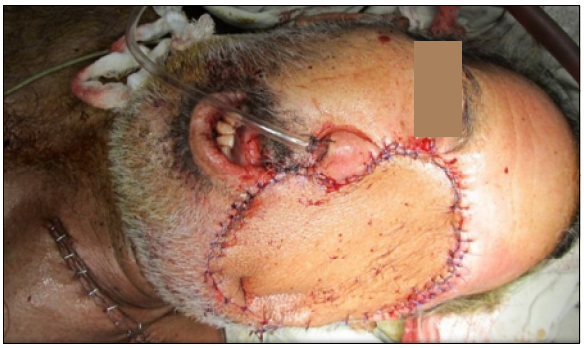


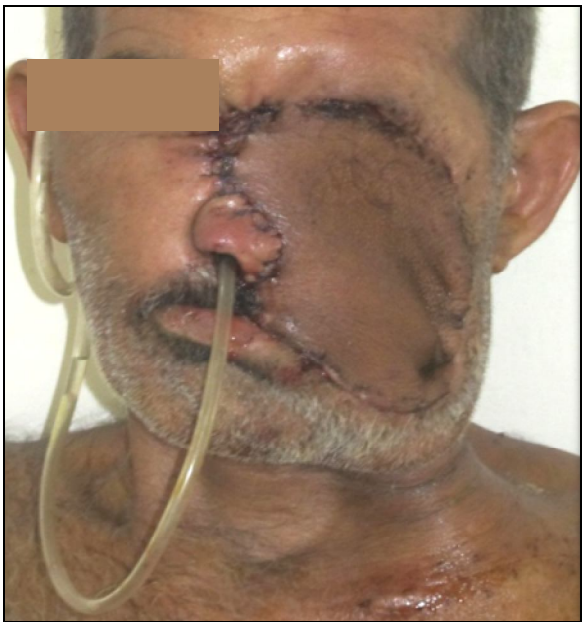
Preoperative view of the patient

Dimensions of the defect after resection of the tumour



Marking for Lattismus dorsi myocutaneous pedicled flap





Postoperative view after three weeks

DISCUSSION

LMS are classically divided into those that originate from the retroperitoneal/intra-abdominal structures, superficial soft tissues and blood vessels.[10] These tumours only rarely present in the oral cavity, and this is thought to be due to the general absence of smooth muscle in the region, except in the blood vessels, circumvallate papillae of the tongue and the occasional primitive mesenchymal tissue.[9] The exact source of smooth muscle in individual tumours remains speculative.

Oral LMS occurs more frequently in males (M:F=1.3:1). All tumours occurred in adults with a peak incidence in the 6th decade. This is similar to that reported earlier[8,11–13] although some tumours have been diagnosed in infants[8,14].

The tumours most frequently presented as a painless mass as in this patient the mass was painless. This finding is similar to those reported in the literature. Interestingly, Montgomery *et al*[15] reported that while 80% of leiomyomas were painful, only two out of 36 LMS (6%) were painful or tender. The lack

of any distinguishing clinical features and the rarity of these lesions often results in them being mistaken for the more common lesions affecting the oral cavity, and correct diagnosis is made only following definitive histological examination.

Our patient presented with the lesion of the maxilla which is the most common site in the literature followed by the mandible, tongue, cheek and floor of mouth in descending frequency. The cause of this apparent variation is unknown and is quite different from benign leiomyomas, which occur more frequently in the lips, tongue, cheeks and palate[9]. Almost 65% of the reported tumours involved the maxilla or mandible, and this predilection to involve the jaw bones has been noted in previous reports.[8–13]

The diagnosis is often made on light microscopy and confirmed by immunohistochemistry.[8–11,13,14] Most lesions demonstrate light microscopic features of interlacing bundles of spindle-shaped smooth muscle cells, with eosinophilic cytoplasm and oval nucleus. The presence of mitosis, cellular and nuclear atypia, and necrosis are often necessary to diagnose a malignant lesion. Immunohistochemical studies frequently reveal the presence of positive staining for the myogenic markers smooth muscle-specific actin, desmin and HHF-35. Most lesions in addition are negative to S100 protein and CAM5.2, although in a very few cases these might be positive, but a diagnosis of LMS is only made if the tumour positively stains for the myogenic markers.[8] In this report patient histopathology report demonstrated light microscopic features of LMS, and showed positive staining for the myogenic marker smooth muscle actin and negative for cytokeratin, S-100 and CD-34.

Although histological grade of the tumour, which encompasses mitotic index, amount of necrosis and degree of differentiation, has been considered a prognostic factor for soft-tissue sarcomas[9], this information has been rarely reported in relation to oral leiomyosarcomas.

Surgical excision seems to offer the best outcome provided there is complete removal of the tumour.[8,10,13,15] In this case, patient had complete excision with reconstruction of the defect with lattismus dorsi pedicled myocutaneous flap. After healing of the surgical wounds patient was referred to the oncology department for further management as this case was presented in the tumour board where it was decided that patient would undergo surgery and then followed by adjuant chemo/radiotherapy. Leiomyosarcomas are generally considered radio-resistant. Some reports have suggested a beneficial effect in terms of decreased recurrence, increased survival and the possibility of less radical surgery[11,14], but these differ from the uniformly poor response to radiotherapy used either as a primary or adjuvant method of treatment in oral leiomyosarcomas.[8,15] Chemotherapy is usually used as a palliative modality for inoperable lesions and metastatic disease.[8,13]

The reported incidence of local recurrence for primary oral LMS is 34% and differs from tumours originating from the superficial soft tissues (50%)[11], and retroperitoneum and blood vessels (50%).[6] The frequency of distant metastasis in primary oral LMS is 35%. This is lower than that found in retroperitoneal (57%) and vascular (50%) LMS, but similar to that associated with LMS of the superficial soft tissues (40%).[4] Lungs are the most frequent site of metastasis, although oral LMS seemed to differ from LMS elsewhere in that they metastasized to the regional lymph nodes more frequently (15%).[8]

Although numerous prognostic factors have been identified for LMS arising in other sites, including size, site, grade and TNM stage[12,14], it is generally acknowledged that there are no reliable prognostic factors in the case of primary oral LMS.[8,13] The TNM classification of soft-tissue tumours is not directly applicable to oral LMS, especially in terms of the T stage, and histological grade of tumour has only rarely been reported in oral LMS. It is important to try and identify factors of prognostic significance in this patient group.

The estimated 5 year survival is 55%. Tumours demonstrating bony involvement (maxilla/mandible) and metastasis are associated with poorer prognosis. Increasing age and male gender showed a trend towards worse prognosis, Interestingly, neither increasing size of tumour nor recurrence is associated with poor survival, unlike tumours occurring in other sites.[4,9]

In summary, primary oral LMS is a rare tumour in the oral cavity and is often mistaken for other more common neoplasms arising in the mouth. The diagnosis is established by histological examination and often following immunohistochemical confirmation. Surgical excision seems to be the preferred method of treatment. Local recurrence and metastasis were not uncommon, and site of the tumour was a predictor of metastasis. Bony involvement and metastasis were associated with poor prognosis. A more detailed and standardized reporting of demographic, clinical, pathological and treatment factors, and critical evaluation of the outcome will be the best way forward to obtain more robust indicators of prognostic significance.

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