

# Bilateral Renal Metastases In Oral Leiomyosarcoma: A Case Report

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## Citation

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## Abstract

A primary leiomyosarcoma in the oral cavity with bilateral renal metastasis in a 22-year-old female is reported. The tumour started during pregnancy and progressed to form a large unresectable mass post delivery. CT scan showed an extensive soft tissue lesion in the maxilla extending to the floor of the middle cranial fossa, oral cavity, parapharyngeal space and facial soft tissue. FNA and biopsy from maxilla showed a spindle cells lesion, which expressed smooth muscle actin and desmin in immunohistochemistry and was diagnosed as leiomyosarcoma. The tumour did not express estrogen and progesterone receptors, which have been reported in some uterine and extrauterine leiomyosarcomas. CT scan abdomen showed bilaterally heterogeneously enhancing masses in right and left kidneys. CT guided FNA showed clusters of malignant spindle cells. The patient is being treated with chemotherapy consisting of Adriamycin 60mg (Day1) and Ifosfamide 2g (Day1 to Day3) to be given in 6 courses at an interval of 21 days. The case is interesting due its presentation in association with pregnancy and the presence of bilateral renal metastases. Renal metastases from sarcomas are very rare and have not been reported in primary leiomyosarcoma of oral cavity, which in turn is a rare tumor.

## INTRODUCTION

Leiomyosarcoma account for 5-10% of soft tissue sarcomas. They are principally tumors of adult life and are more common in women than in men. Primary oral leiomyosarcoma is a rare entity with unusual bone location because of paucity of smooth muscle in that site. Isolated renal metastasis in nonrenal sarcomas is extremely rare. We report a histologically proven case of leiomyosarcoma of maxilla with bilateral renal metastasis, which were visualized in CT and diagnosed by CT guided fine needle aspiration cytology (FNAC).

## CASE REPORT

**Clinical History:** We report the case of a 22-year-old female who presented to our outpatient in Dec 2006, with a huge maxillary swelling with intraoral and temporal extension (fig 1a). She had first noticed this swelling one-year back when she was pregnant. She gave a history of progressive increase in the size of the lesion during her pregnancy and also following the delivery. She presented to us four months following her delivery. The swelling was massive involving the whole of the right face. It was congested, painful and there was associated mucus and blood discharge in the oral extension. Lymph nodes were not palpable. Clinical systemic examination revealed no other signs. The

biochemical and haematological parameters were within normal limits.

## RADIOIMAGING STUDIES

**CT scan face:** showed a large soft tissue attenuation lesion with epicentre at the right infratemporal fossa and masseteric space extending superiorly up to the floor of the middle cranial fossa, inferiorly up to the level of upper border of right submandibular gland, medially up to the parapharyngeal space, anteriorly it had destroyed the posterior wall of right maxillary sinus, laterally it had involved the masseter and facial soft tissue and posteriorly it had involved the posterior styloid compartment of right parapharyngeal space (fig.1b).. The impression was that of a nerve sheath tumour or a neoplastic soft tissue lesion.

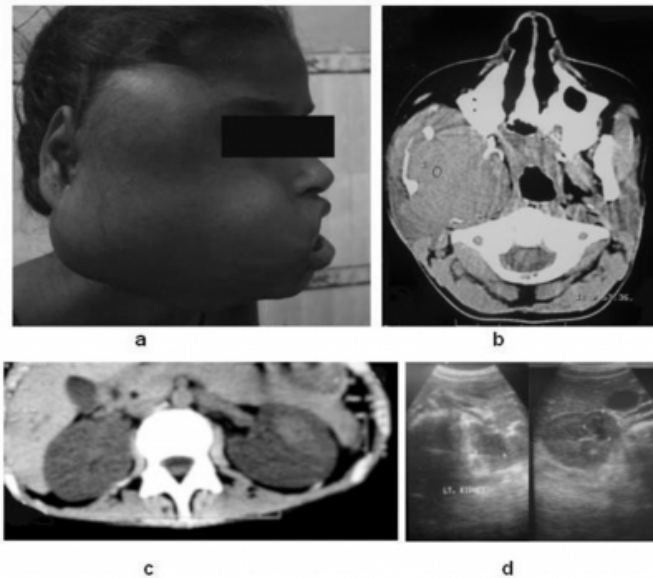
**Chest X-Ray:** showed normal architecture. No mass lesions were visualized.

**Ultrasonography of the Abdomen** revealed bilateral hypoechoic renal masses (fig.1c).

**CT scan Abdomen:** showed ill-defined hyperdense lobulated masses in both kidneys (fig1d).

**Figure 1**

Fig 1: (a) Photograph of patient showing extensive lesion on right side of face (b) CT scan face: large extensive soft tissue attenuation lesion with epicentre at the right infratemporal fossa and masseteric space (c) Ultrasound kidney: bilateral hypoechoic renal masses (d) CT abdomen: showing bilateral heterogenous renal masses



## **PATHOLOGICAL EVALUATION**

FNAC of the oral lesion: showed cellular smears with clusters of short spindle cells with oval nuclei and moderate amount of cytoplasm. The cells showed moderate degree of nuclear pleomorphism (fig2a). Few cells in mitoses were not evident. The impression was that of a spindle cell sarcoma and an incisional biopsy was advised.

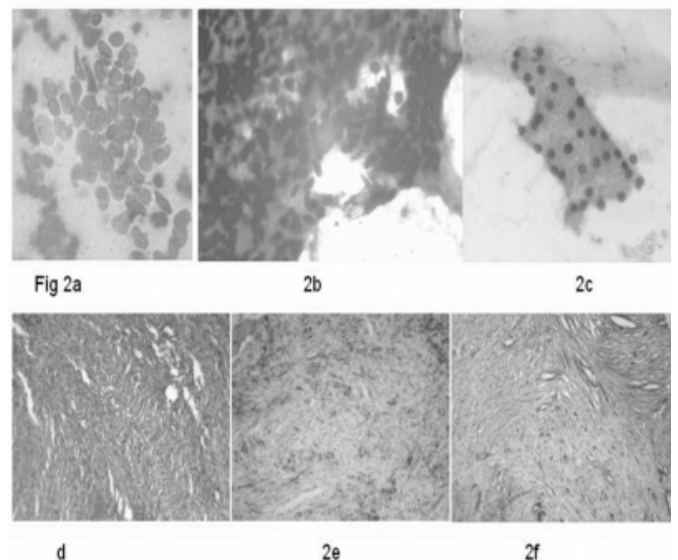
Incisional biopsy of the oral lesion: Histopathological evaluation of incisional biopsy taken from the oral protrusion of the tumor revealed a malignant neoplasm consisting of intersecting fascicles of spindle cells cut at right angles to each other (fig 2d). Tumour cells displayed vesicular, oval to elongated nuclei with occasional prominent nucleoli and eosinophilic cytoplasm. Mitotic activity was present. The histopathological picture was consistent with a spindle cell sarcoma. On Immunohistochemical expression of smooth muscle actin (fig 2f) and desmin (fig 2e) was diffusely positive in most tumor cells confirming the diagnosis of Leiomyosarcoma. Neurofilament expression was negative. In view of the evolution and growth of tumor in pregnancy we evaluated the lesion for presence of estrogen and progesterone receptors. Both these receptors were not expressed. All primary antibodies were obtained from Dakopatts Denmark and secondary LSAB-2 kit from Dakopatts Denmark was

used for detection with Diaminbenzidine as substrate.

CT guided aspirate from bilateral renal lesions revealed few cellular clusters of bipolar spindle cells with high nucleocytoplasmic ratio and moderate degree of nuclear pleomorphism (fig 2b). Clusters of normal renal tubular cells were also seen (fig 2c). These findings were consistent with the diagnosis of metastatic leiomyosarcoma in the kidneys.

**Figure 2**

Fig 2: (a) Cluster of tumour cells in FNA from maxilla (Giemsa x 525 x digital magnification) (b) Spindled tumour cells in FNA from renal lesion and (c) a cluster of benign renal tubular cells (H& E x 525 x digital magnification) (d) Biopsy showing intersecting fascicles of spindled cells (H & E x 125 x digital magnification) (e) Tumour cells expressing desmin and (f) smooth muscle actin (LSAB x 125 x digital magnification)



Therapy: Surgical resection was not possible due to the extensive local spread. In view of the fact that the primary tumour was not resectable and renal metastases were present, the patient was put on chemotherapy. She is currently receiving Adriamycin 60mg (Day1) and Ifosfamide 2g(Day1-Day3) in 6 courses to be repeated at 21days.

## **DISCUSSION**

Leiomyosarcoma (LMS) of the oral cavity is a very rare tumor that is associated with aggressive clinical behavior and low survival. They are best treated surgically, early and aggressively.<sup>1</sup>

The incidence of LMS among sarcomas of oral & maxillofacial region was found to be 6.25% by Yamaguchi et al<sup>2</sup> in a study of 32 cases of oral and maxillofacial

sarcomas. Oral LMS have been found to have a peak incidence in the 3<sup>rd</sup> decade and 6<sup>th</sup>-7<sup>th</sup> decade with no gender predilection. Females have a higher incidence in the third decade whereas males have an even age distribution<sup>3</sup>. Estrogen receptor positivity has also been reported in some leiomyosarcomas raising the possibility of hormonal responsiveness. In an Immunohistochemical study of estrogen receptor (ER) and progesterone receptor (PR) expression in uterine and extrauterine leiomyosarcomas, it was noted that most uterine LMS co-express ER and PR, and most extrauterine LMS do not stain for these antigens. However, a subset of extrauterine LMS is ER and/or PR immunoreactive<sup>4</sup>. We have evaluated the expression of estrogen and progesterone receptors in our case but found no nuclear positive staining for receptors. Further in our case the tumour developed and progressed during pregnancy. The occurrence of cancer in pregnant women is relatively infrequent. Sarcomas, which have been diagnosed during pregnancy, include osteosarcoma, rhabdomyosarcoma, liposarcoma, Ewing's Sarcoma, chondrosarcoma<sup>5</sup>. Though there are reports of uterine LMS and vulvar LMS diagnosed in pregnancy, however we did not come across any oral leiomyosarcomas occurring during pregnancy in the literature.

Leiomyosarcomas are rare in the maxilla due to minimal smooth muscle in the region. In several studies of oral LMS, 50% -70% of the cases arose from the jaws<sup>(1,3,6)</sup>. Most of the cases appeared to be associated with neurovascular structures of the facial skeleton as evidenced by the imaging studies<sup>3</sup>. In our case, though the clinical examination and imaging results indicated a maxillary origin of the tumour, but the exact origin could not be ascertained due to advanced local disease and unresectability of the tumour at the time of presentation.

Distant metastasis seen in 39% cases of oral LMS, occurred mostly to the lungs and some cases showed cervical lymph node metastasis<sup>1</sup>. The case presented above had no metastasis to the lung as evidenced by chest X-ray and had no enlarged lymph nodes. A case of maxillary LMS with spinal metastasis has been reported in the literature<sup>7</sup>. We could find no report in literature of renal metastasis from oral LMS.

Although metastases to the kidney have been reported to occur in 7-20% of patients with cancer at autopsy<sup>8</sup>, the diagnosis of metastases to the kidney in patients without evidence of a disseminated nonrenal malignancy is rare. In a

review of 100 consecutive patients with nonrenal malignancies diagnosed with renal masses at presentation or follow-up, a total of 19 patients were found with metastases to the kidney which included those with primary tumors of the lung, lymphoma, esophagus, head and neck, breast, colon, pancreas, extremity sarcoma, testis and myelodysplasia<sup>9</sup>. Other primary neoplasms which may metastasize to the kidney include carcinoma of the stomach, melanoma and leukaemia. Metastases to the kidneys are frequently small, asymptomatic and detected only at autopsy. Renal metastases may present as multiple or bilateral small renal masses or, less commonly, as a solitary lesion. Our case presented with bilateral renal masses in the presence of a nonrenal malignancy, which was very suggestive of metastasis to the kidney. We further confirmed the presence of spindle cell neoplasm in CT guided FNAC from the renal lesions.

The diagnosis of soft tissue leiomyosarcoma is based on the morphologic features mainly a spindle cell neoplasm with intersecting bundles, blunt ended nuclei, varying degrees of anaplasia and mitoses with the support of IHC, chiefly Smooth muscle actin and desmin<sup>10</sup>. The case presented, fulfilled these criteria confirming the diagnosis of leiomyosarcoma.

Surgical resection, hemimandibulectomy or hemimaxillectomy, partial maxillectomy, are the preferred initial treatments for cases of leiomyosarcoma primary in the jawbones. Regional lymph node resection is recommended if there is clinical evidence of lymphadenopathy. In the present case as surgical resection was not possible chemotherapy was instituted. The patient has received three cycles of anthracycline based chemotherapy and is planned for further chemotherapy. Some regression in the size of the lesion has been observed.

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