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Case Report

Surgery Section

# Uterine Leiomyosarcoma with Cutaneous and Fibular Bone Metastasis: A Rare Case Report

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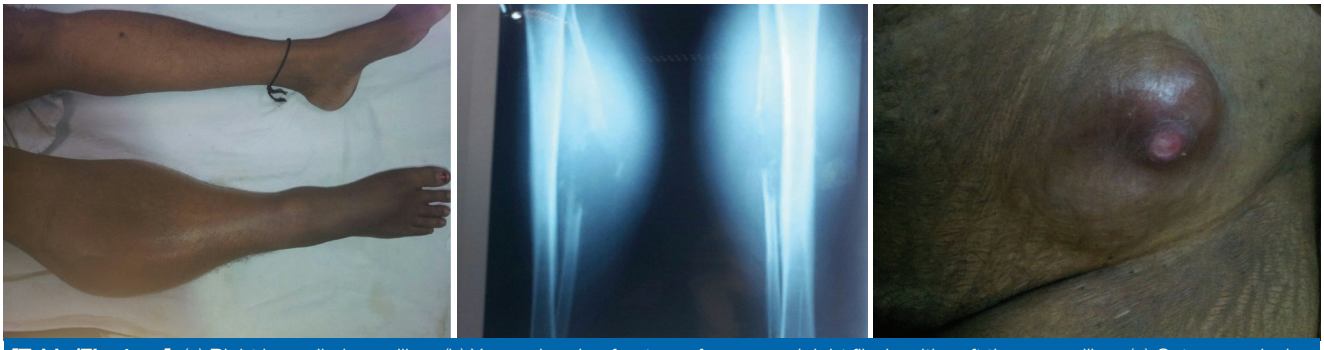
## ABSTRACT

Uterine sarcomas accounts for about 3% of all uterine cancer. Cutaneous and bony metastasis is extremely rare in uterine leiomyosarcomas. Bony metastasis to fibula from endometrial carcinomas is very rare and very few case reports have been documented with axial skeletal and cutaneous metastasis from uterine leiomyosarcoma. We hereby reports a rare case of metastasis to the fibula and soft tissue, which occurred in less than six months after uterine leiomyosarcoma diagnosis and its treatment, demonstrating the ability of this tumour to metastasize to both bone and soft tissue.

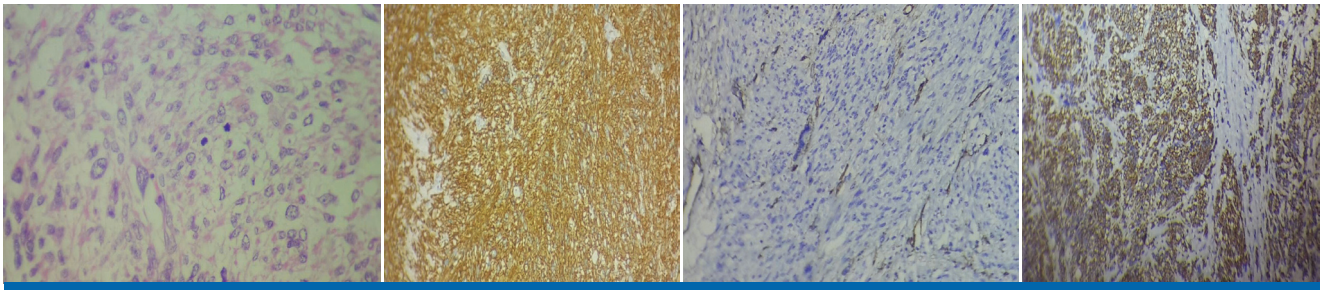
**Keywords:** Amputation, Mesenchymal tumour, Salpingo-oophorectomy

## CASE REPORT

A 42-year-old female patient was referred to us from the Department of Gynaecology with chief complaints of pain in the right lower limb for past four months with ulcerated lesion in the ipsilateral iliac fossa with a previous history of total abdominal hysterectomy and bilateral salpingo-oophorectomy six months back for a uterine mass. On the basis of histopathology, diagnosis of uterine leiomyosarcoma was made, which was confirmed with immunohistochemistry. She received one cycle of MAID regimen after this therapy developed those above mentioned symptoms. Her pain was not relieved on medications including opiods. On detailed examination 15x10 cm swelling present in upper fibular region with warmth and bony crepitations, severe tenderness was present [Table/Fig-1a]. Distal neurovascular examination was normal. X-Ray of the right leg done showed fracture of the fibula in upper 1/3$^{rd}$ with increased soft tissue density [Table/Fig-1b]. A 5x5 cm swelling present in the right iliac fossa fixed to skin [Table/Fig-1c]. Other systemic examinations were within normal limit. Core needle biopsy from the lesion showed features of spindle cell tumour and biopsy of groin lesion showed features of metastasis from spindle cell tumours [Table/Fig-2a]. X-ray chest was normal and performance status of patient was good [KPS-0]. Accordingly patient underwent for above knee amputation of the right lower limb with wide local excision of the groin lesion as a palliative procedure. She withstood the procedure well and recovered within seven days. She totally relieved of the pain and is off analgesics. The final histopathology was suggestive of leiomyosarcoma of the fibula and same histological features in the skin lesion with all margins negative. Immunohistochemistry showed SMA, H-CALDSMON positivity, [Table/Fig-2b-d] focal positivity for desmin and negative for CK, EMA, S-100. The final conclusion was uterine leiomyosarcoma with fibula and cutaneous metastasis and she was planned for palliative chemotherapy. She received six cycle of chemotherapy and three monthly follow-up was done with clinical examination and X-ray chest till two years and she is still in follow-up without any recurrence.



**[Table/Fig-1a-c]:** (a) Right lower limb swelling; (b) X- ray showing fracture of upper end right fibula with soft tissue swelling; (c) Cutaneous lesion of anterior abdominal wall in the left iliac region.



**[Table/Fig-2a-d]:** (a) Microscopic picture showing spindle cell with pleomorphism; (b) Immunohistochemical staining for smooth muscle antigen; (c) Absence of immunohistochemical staining for CD 34; (d) Immunohistochemical staining for H-caldesmon.

## DISCUSSION

Uterine sarcomas have varieties of histological subtypes which includes leiomyosarcoma, which is the most common subtype followed by endometrial stromal tumours, undifferentiated sarcomas, pure sarcomas, mixed epithelial and mesenchymal tumours. Uterine leiomyosarcomas account for approximately 7% of all new cases of soft tissue sarcoma and 1% of all uterine malignancies with an annual incidence of 0.64 per 100,000 women [1]. Majority of cases are de novo, but few cases have been reported with a history of prior irradiation [2]. Lung, thyroid, liver, brain, and bone are the most common sites of metastasis via haematogenous route [3,4]. Less common metastatic sites include the breast [5] and soft tissue. Uterine leiomyosarcomas have high metastatic potential with five year survival rates of 0-73%. Reported 5-year survival of leiomyosarcomas varies with stage of the disease. For stage I disease reported 5-year survival is 53% and whereas, it is less than 10% for stage IV. Uterine leiomyosarcomas have a poor prognosis because of its high local recurrence and distant metastasis [6]. Important prognostic factors as related to survival as described in literature are stage, age, and tumour size and parity status of the patient. Higher parity (up to three deliveries) had a negative influence on survival in cases of uterine sarcoma [7].

Cutaneous metastasis is very rare in and defined as any malignant lesion arising from the skin not in continuity with the primary tumour [8]. These lesions account for 0.7% to 9% of all metastases [9]. Cutaneous metastasis in uterine leiomyosarcomas is rare. To the best of our knowledge very few cases have been reported in the literature worldwide which includes two cases of scalp and one case of facial metastasis from proven uterine leiomyosarcoma [10-13]. Gardiner reported first case of cutaneous metastasis in an autopsy study which showed widespread metastasis involving multiple organs like lungs, liver, pancreas, bone and scalp [10]. Another case disseminated intra-abdominal metastases and multiple cutaneous lesions reported later in 2004 [11]. In 2010 cutaneous metastasis to scalp and back was published [12] and another case of histologically confirmed scalp metastasis from primary ULMS was published [13]. In pubmed search we found one case of primary uterine leiomyosarcoma with orbital metastasis other with skull bone metastasis [14,15]. Two more cases of skeletal metastasis were reported in literature but the primary was rectum and intestine [16]. Bone metastasis from uterine sarcomas is more common than leiomyosarcomas. Three cases of uterine sarcomas with skeletal metastasis – two of them in the thoracic spine have been reported. One case appeared 18 years and the other 7 years, after initial treatment [17] and other report of cranial metastasis to the parietal bone, which occurred 1 year after diagnosis [18].

Simple hysterectomy with or without salpingo-oophorectomy is the surgical gold standard. Dissection of pelvic and para-aortic lymph nodes are not recommended since lymph node involvement is rare. Adnexal removal and lymphadenectomy have not proved its role in the management of uterine leiomyosarcomas and still a matter of debate [19]. Reported incidence of lymph node metastasis is 0–47% in some studies and survival has not been significantly affected by the extent of lymphadenectomy [20]. The role of adjuvant therapies is also controversial. Radiotherapy seems to improve local control but not overall survival. Adjuvant chemotherapy does not decrease the risk of distant metastatic or improves the survival. The response rates of different chemotherapeutic regimens in recurrent uterine sarcomas have been reported up to 57%. However, it is reasonable to offer palliative chemotherapy to patients with advanced uterine sarcoma. The effects of hormone therapy in cases of recurrent uterine sarcoma have been assessed in only a few studies [21].

## CONCLUSION

Uterine leiomyosarcoma is a rare disease. Once diagnosed, systematic evaluation should be done to evaluate the stage and rule out any metastasis. Staging evaluation includes clinical examination, local examination in form of ultrasound and contrast enhanced CT-abdomen, pelvis and chest multidisciplinary team discussion should be done for stage wise management. In view of recurrent or metastatic disease biopsy should be done to identify any suspicious skin lesion and keep high index of suspicion for metastatic potential of leiomyosarcoma to the skin and bones. Due to the rarity of these cutaneous and bony metastases and the lack of studies in this area, there is no specific guideline as to the optimal extent of an oncological safe margin.

## CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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## ABBREVIATIONS

KPS, Karnofsky Performance Status; CK, Cytokeratin; EMA, Epithelial Membrane Antigen; SMA, Smooth Muscle Antigen; MAID, Mesna Adriyamycin Ifosamide Dacarbazine; TAH+BSO, Total Abdominal Hysterectomy and Bilateral Salpingo-oophorectomy.

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