# Incidental Finding of a Peripheral Nerve Sheath Sarcoma

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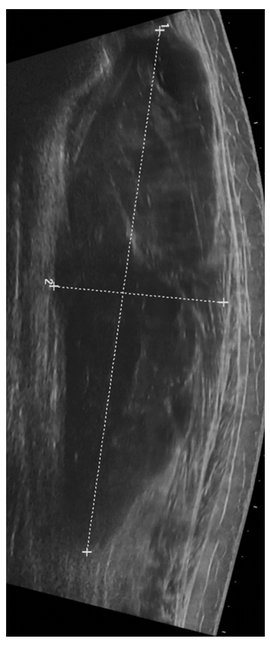
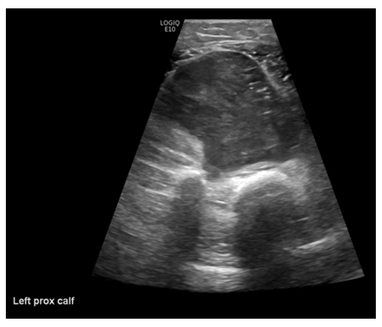
## Patient history

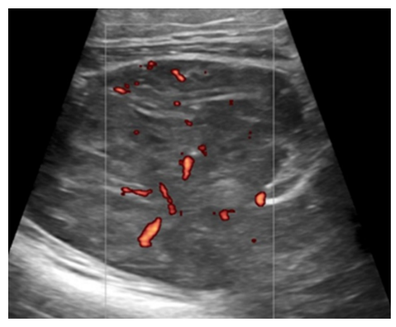
A 30 year old female patient, with a six month history of left calf pain and foot drop, presented with  intense calf pain and swelling. No history of trauma. Initial referral for MRI scan of her foot and ankle six months ago reported 'suggested myopathic changes in several muscles'. No follow-up treatment was given. Patient experienced increasing difficulty in walking and developed localised severe calf pain. She was referred by her GP for an ultrasound of her leg veins to exclude a DVT.

## Ultrasound findings

On ultrasound examination there was no evidence of a DVT, however, in the popliteal fossa a well defined, heterogeneous, intramuscular mass was visualised extending inferiorly in the proximal posterior calf with abnormal vascularity measuring 20 cm in length with a diameter of 6 x 6 cm. Longitudinal, transverse and panoramic views were acquired, and power doppler assessed vascularity of the mass. Images were reviewed immediately by a Radiologist and referred for urgent MRI calf.



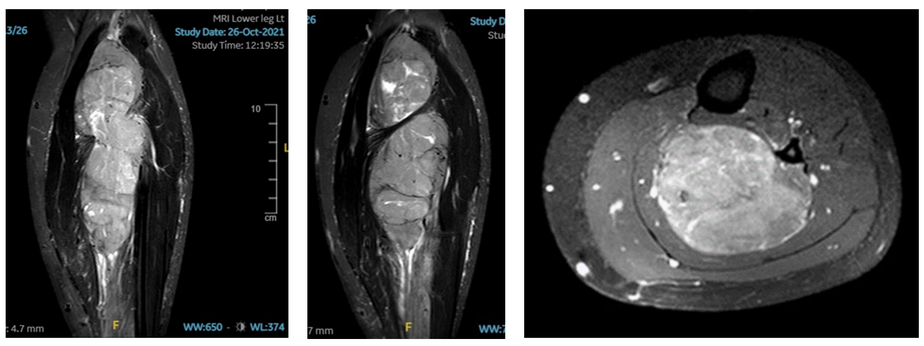


Ultrasound of the proximal calf mass

## MRI findings

The MRI report stated 'there is a well-defined elongated intermuscular soft tissue mass lesion in the calf closely related to the neurovascular bundle, suspicious for a tumour of neurogenic origin possibly a malignant peripheral nerve sheath tumour or given its close relationship to neurovascular bundle possibly a leiomyosarcoma.  The previous signal changes on the MRI foot could be due to denervation/ compression of the posterior tibial nerve.'




MRI images of the calf

## Patient outcome

Patient was referred to the specialist Sarcoma team, underwent amputation of the distal lower limb and is undergoing chemotherapy. She has been hospitalised with sepsis twice since surgery.

She has now been referred to Stanmore Hospital, a specialist centre in the care and treatment of patients who suffer from sarcomas. Treatment includes bespoke endo-prostheses, bone transportation and bone grafting. Histology of the tumour confirmed the diagnosis of a malignant peripheral nerve sheath tumour.

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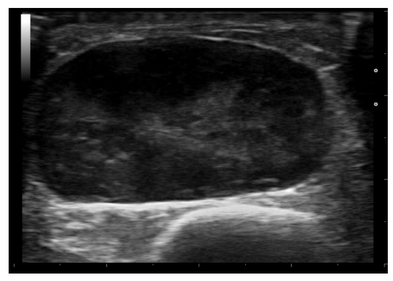
## Soft Tissue Sarcoma STS

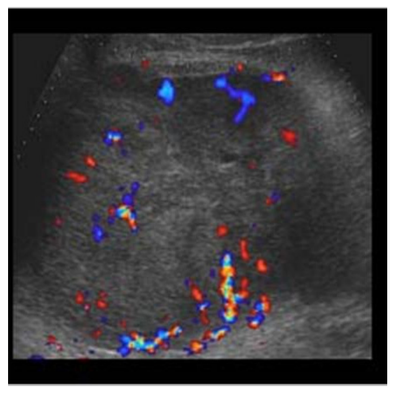
Soft tissue sarcoma (STS) are a group of neoplasms of mesenchymal origin. They account for 88% of total sarcomas and represent 1% of all malignant tumours. They usually present as asymptomatic masses, often delaying diagnosis. Metastases are present in 20% cases when the tumours are identified.

The average age of STS diagnosis is 50-60 years. The risk increases with previous radiotherapy, ionizing radiation, chronic lymphoedema (lymphangiosarcoma), exposure to vinyl chloride, arsenic, dioxins and Thorotrast (hepatic angiosarcoma). It is also associated with some inherited syndromes. It has been observed as an association between viral infections, such as Epstein Barr virus in patients with AIDS and leiomyosarcoma. More than 50% of soft tissue sarcomas arise in any of the mesodermal tissues of extremities (only 14% in upper limbs). The most frequent form of presentation is usually a painless, fast-growing tumor of increased consistency.

## Malignant nerve sheath tumour

Malignant peripheral nerve sheath tumours (MPNST), also known as neurofibrosarcomas, can occur anywhere throughout the body mainly in adults. They are rare malignant mesenchymal lesions that account for 5% to 10% of all soft tissue sarcoma. They are extremely aggressive with high local recurrence rate and poor survival. Early diagnosis increases the chance of successful treatment. Resection surgery with a margin is the main therapy for MPNST, radiation and systemic chemotherapy are also widely used despite their uncertain effect.



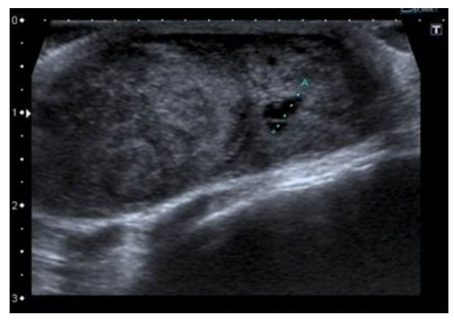


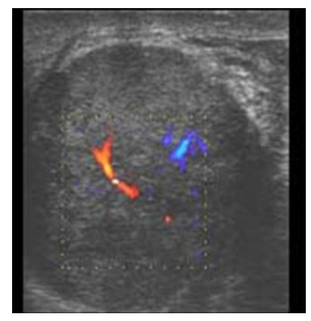
Examples of MNST on ultrasound

## Leiomyosarcoma

Leiomyosarcomas (LMS) are malignant soft tissue sarcomas arising from smooth muscle cells (involuntary muscles). LMS account for approximately 16% of all non-visceral soft tissue sarcomas. In comparison with other sarcoma subtypes, it represents a relatively aggressive tumor with a high risk of metastasis.

The prognosis mainly depends on tumor size, the histological grade and tumor persistence at the surgical margins. High-grade sarcomas are related to higher rates of local treatment failure and higher metastatic potential.





Examples of LTS on ultrasound

## References

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