Case Report

# Rare case report - Recurrent Spindle Cell tumor of the thigh

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## ABSTRACT

This case is to discuss the rarest of a rare recurrent spindle cell tumor of the proximal thigh which is one of the soft tissue neoplasms that can occur in various locations of our body including the trunk and extremities. Soft tissue masses are commonly present in surgical practices and range from benign lipomas to high-grade soft tissue sarcomas. A subset of soft tissue sarcomas includes a spindle cell sarcoma, which is a rare connective tissue tumor that originates in the layers of tissue found below the skin. The aim of this case report is to describe the presentation of spindle cell tumors, the investigations required, and the optimal management of the disease. Surgical resection i.e., wide local excision is the mainstay of treatment for spindle cell sarcomas but treatment can also include a combination of surgery, chemotherapy, and radiation.

**Key words:** Recurrent tumor, Spindle cell tumor, Wide local excision

Spindle cell tumors are a rare group of malignant tumors arising from the mesenchymal tissue, which make up <1% of all adult malignancies [1]. Since most of them are painless, mobile, and have slow growth, both physicians and patients confuse them with benign tumors, and appropriate investigations such as biopsies are missed [2]. The World Health Organization classifies the group of soft-tissue neoplasm into >100 different histological subtypes based on the presumptive tissue of origin and their architectural pattern. A huge group comprises undifferentiated soft-tissue sarcomas [3]. Sarcoma usually presents as a painless mass and rarely presents with distant metastasis, especially in the lungs, liver, brain, etc. [4]. Spindle cell sarcoma is one of the rare varieties of undifferentiated soft-tissue sarcomas [5].

Due to a rare entity, only a few cases have been described in medical literature [5]. Here, we report the case of a 31-year-old male with spindle cell sarcoma of the proximal thigh, which recurred locally after surgery.

## CASE PRESENTATION

A 31-year-old man came with complaints of recurrent swelling over the left proximal thigh after a previous surgery which was done 2 months back at a different center. There was no positive family history in his 2nd or 3rd-degree relatives.

The patient had moderate general condition, nutritionally well-built with normal vitals having a pulse of 80 bpm and blood pressure of 130/70 mmHg. On clinical examination, the patient had a large lobulated swelling around 6 × 5 cm, 5 × 4cm, and 3 × 3 cm over the lateral aspect of the left proximal thigh (Fig. 1). The swelling was not associated with pain or any constitutional symptoms, and the inguinal nodes were not palpable clinically along with no history suggestive of distant metastasis, his opposite thigh was normal.

Magnetic resonance imaging (MRI) of the thigh was suggestive of 3 heterogeneously enhancing lesions in a subcutaneous plane not involving underlying muscle or fascia (Fig. 2a and b). An ultrasound (USG)-guided trucut biopsy was done which was suggestive of soft tissue neoplasm-"spindle cell tumor." Staging was done based on an HRCT chest, USG abdomen, and other blood investigations.

A multidisciplinary approach involving medical oncologists, radiologists, and surgeons was adopted. He underwent surgery after the proper staging of the disease and wide local excision of the tumor (Fig. 3a). One plane below the tumor and 2 cm of adequate margin with primary flap closure named 0 to Z flap (Fig. 3b) with drain placed.

Post-operatively, the patient was discharged on day 4 with a drain and had regular follow-ups in the outpatient department. He had a sarcoma associated with the flap, so it was aspirated and regular dressing was done. Histopathology, the entity was diagnosed as spindle cell sarcoma with all naga five margins of tumor, and immunohistochemistry was suggestive of malignant peripheral nerve sheath tumor (Ki67-40%, CD34 -ve, SMA -ve, Desmin -ve, S-100 -ve, SOX10 -ve).

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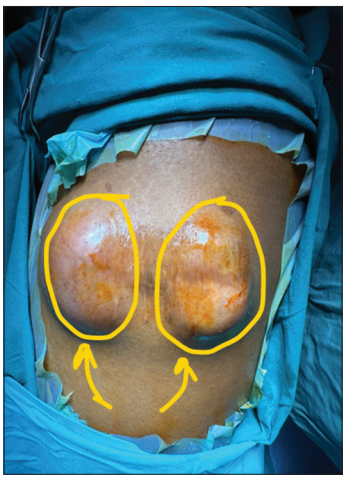

**Figure 1: Picture depicting lobulated masses over left proximal thigh and also showing previous surgery scar mark**

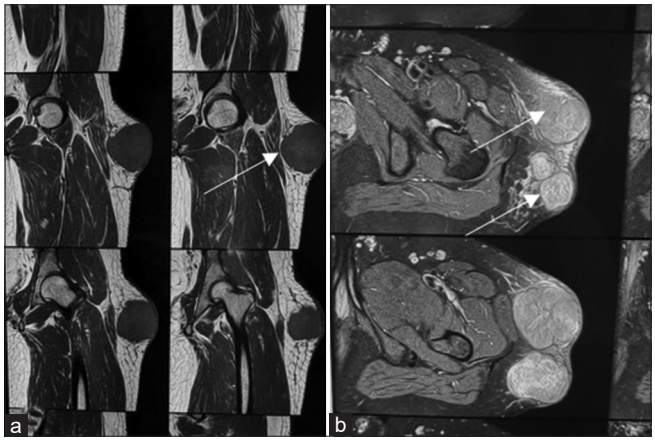


**Figure 2: (a) Magnetic resonance imaging of left thigh detecting a heterogenous mass which is well above the muscle plane and in the subcutaneous tissue (b) Another section depicting a multilobulated heterogenous mass well above the adjustment muscle plane and fascia**

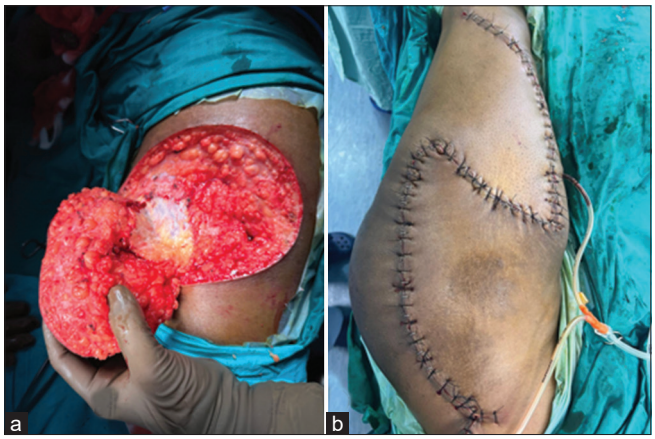


**Figure 3: (a) Intraoperative Image depicting wide local excision well above the muscle plane getting margin-free excision (b) Final image depicting a 0-Z flap closure of the large defect after wide local excision**

## DISCUSSION

Soft tissue sarcomas are challenging due to their aggressive nature although the potential for metastasis is low. This case emphasizes the importance of early and accurate diagnosis, complete surgical resection with clear margins, and the role of adjuvant radiotherapy in managing high-grade tumors. The patient's favorable outcome highlights the effectiveness of a multidisciplinary approach to treatment.

The evaluation of a patient with a suspected soft-tissue sarcoma includes history, imaging, and biopsy [6]. MRI is the preferred modality for the evaluation of soft-tissue masses of the extremities, trunk, head, and neck [7]. All patients diagnosed with sarcoma should also have a computed tomography scan of the chest at the time of diagnosis and in follow-up [6].

Spindle cell sarcomas affect people of almost any age and sex [8]. Two separate studies by Feng *et al*. and Smith *et al*. showed the median age at presentation of 57 years [9, 10]. In contrary to these studies, the age at presentation in this case is quite early (31 years). In the same study by Smith *et al*., the median tumor size found was 9.87 cm [10]. This patient had 3 lobulated tumors of different sizes at the first presentation while it grew up to the sizes of 6 × 5 cm, 2 × 3 × 3 cm, and 4 × 5 × 6 cm when they recurred later after the first surgery. This presentation is quite rare as this was a recurrence and advanced to a large size.

Due to their rarity, delay or misdiagnosis is common for sarcomas, especially in settings with limited facilities [11,12]. In this case, the mass was initially identified as lipofibroma in previous surgery. Later, repeat biopsies/FNAC revealed a spindle cell tumor. The rarity of the case and the little experience in diagnosing rare cases might have led to this error in histological diagnosis [13].

This patient had a recurrence within 1.5–2 months after surgery, which also is quite rare. A regular follow-up is therefore advised after the excision of sarcoma, especially in the first 2 years. Soft tissue tumors are best managed by surgical resection with or without radiotherapy. Chemotherapy is mainly reserved for patients who present with metastases or with deep tissue invasion [14,15].

In this case, the lack of proper communication between the hospitals and doctors involved might be one of the reasons for his loss of follow-up. Besides his financial constraints and ignorance, inadequate counseling about the diagnosis may have led to the discontinuity of follow up, which is very common in developing countries. Due to limited resources in hospitals, it is common in developing countries to refer patients to other centers for further care. Hence, one of the main focuses regarding this patient would be to ensure adequate follow-up. To avoid such circumstances in the future, it is better if the healthcare team takes steps and arranges continuity of care for the patient so that any future patients do not leave without completing their course of treatment. The use of telemedicine is proven to be highly effective in ensuring follow-up in developing countries. In diseases like soft tissue tumors, it is necessary to maintain compliance with the treatment so as to avoid such relapse and recurrence.

## CONCLUSION

Hence, concluding this case report on a note that the management of soft-tissue sarcomas is not always simple and straight forward

and it should be best done in a center with appropriate expertise in multiple fields. It has to be a multidisciplinary approach with which this kind of rare cases get cured with a good success rate. In the absence of appropriate expertise or resources, patients do not get adequate treatment on time. This may lead to grave consequences, such as metastasis or recurrence causing significant problems of the patients. Regular follow-up after treatment with history, physical examination and chest imaging is of utmost importance. Soft tissue tumors are always unpredictable neoplasms that need proper attention and aggressive management with a multidisciplinary treatment and regular follow-up of these patients is the key. Multi-centric studies using this approach will help in establishing standard management guidelines for these patients.

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