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# Giant Atypical Spindle Cell Lipomatous Tumor of Retroperitoneal Origin Presenting with Lung Metastasis and Extension to the Thigh: A Case Report with Literature Review

Paschyanti Kasat[1] Shivali Kashikar[1] Pratapsingh Parihar[1] Vadlamudi Nagendra[1] Pratiksha Sachani[1]

[1] Department of Radiodiagnosis, Jawaharlal Nehru Medical College (JNMC), DMIMS, Wardha, Maharashtra, India

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**Address for correspondence** Paschyanti Kasat, MBBS, Department of Radiodiagnosis, Jawaharlal Nehru Medical College (JNMC), DMIMS, Sawangi Meghe, Wardha, Maharashtra 442001, India (e-mail: paschyantikasat@gmail.com).

## Abstract

Retroperitoneal liposarcomas are rare, and their variants, atypical spindle cell lipomatous tumor (ASCLT), are rarer. Though ASCLTs are benign, they have high recurrence despite complete surgical excision. We present a rare case of a 22-year-old male presented with insidious-onset, and gradually progressive swelling over left inguinofemoral, and iliac fossa region. Also, the patients had a history of weight loss, pricking-type pain radiating to left lower limb, and breathlessness on exertion. Contrast-enhanced computed tomography (CECT) abdomen revealed a large well-defined heterogeneously enhancing soft mass located in pelvis, and involving left-sided iliopsoas muscle suggestive of retroperitoneal soft tissue sarcoma and CECT thorax showed metastasis. The CT-guided core biopsy led to the definitive diagnosis of low-grade ASCLT. Subsequently, the patient was initiated on concurrent neoadjuvant chemoradiotherapy followed by complete surgical excision. The findings in this report make it critical to enhance our understanding of this rare tumor, with surgery being the best treatment option.

**Keywords**

- atypical spindle cell lipomatous tumor
- liposarcoma
- retroperitoneal
- soft tissue sarcoma
- spindle cell tumor

## Introduction

Retroperitoneal sarcomas (RPS), rare malignant tumors (< 1%), originating from mesenchymal cells, are generally situated in muscular, adipose, and connective tissues. In the retroperitoneum, around 33% malignant tumors are sarcomas, and approximately 15% of the soft tissue sarcomas (STS) have retroperitoneal (RP) origin.[1] Liposarcoma (LS), accounting for around 20% of malignant mesenchymal tumors, and 40% of RPS, is the predominant STS in adults.[2] Characterized by prominent spindle cell component, spindle cell liposarcoma (SCLS) is a rare variant of well-differentiated LS.[3] In a recent classification, the World Health Organization has renamed SCLS as atypical spindle cell lipomatous tumors (ASCLT) and classified them as benign tumors.[4] Owing to rarity, their prevalence is unknown.[3,5–7]

Patients with sarcoma are generally asymptomatic, and have a painless mass (15–18 cm) at presentation.[8] Rarely, the patient presents with distant metastasis, particularly in the lungs.[5] Computed tomography (CT) of the abdomen and pelvis as well as thorax is sufficient to reach the diagnosis, and assess the distant metastasis, respectively. Though literature suggests extension of RP LS to thigh,[9,10] extension of ASCLT to thigh has not been reported yet. Herein, we present a case of giant metastatic ASCLT originating from the RP region in a young male.

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## Case Description

A 22-year-old male presented with swelling over left inguinofemoral region (6 × 5 cm), and left iliac fossa region (5 × 4 cm) over the last 45-, and 15 days, respectively. Both the swellings had insidious-onset, and gradually progressed to the current size. The patient had a history of weight loss (4 kg in the last 30 days), and pricking-type pain radiating to left lower limb. He also had a history of on-and-off fever, and cough with expectoration over the last 15 days, and breathlessness on exertion over last 30 days. Also, 30 days back, he complained of right-sided chest pain, and breathlessness that was managed conservatively. There was no history of trauma and comorbidities.

General and systemic examinations were normal, except for the presence of left-sided axillary lymphadenopathy, and decreased right-sided breath sounds. Local examination revealed swelling over left inguinofemoral region, 6 × 5 cm in size, firm in consistency, nonmobile, nontender, with well-defined margins, no local rise in temperature, and no skin changes. Chest radiography revealed right lower zone consolidation (►**Fig. 1A**), and multiple variable size nodular opacities in bilateral middle and lower zones suggestive of metastatic lesions, with right pleural-based large metastatic lesions observed in periphery. Abdominal ultrasonography illustrated well-defined heterogenous predominantly hyperechoic lesion with minimal vascularity on Doppler (►**Fig. 1B**). Contrast-enhanced computed tomography (CECT, ►**Fig. 2**) abdomen revealed a large well-defined heterogeneously enhancing soft mass (23 × 13.5 cm) with internal necrosis located in pelvis, and involving left-sided psoas and iliacus muscle suggesting RP origin and exerting mass effect. There was gross hepatomegaly (21 cm) with borderline splenomegaly. There were multiple enhancing left inguinal lymph nodes (largest 2.4 × 1.3 cm), anterior to the lesion. All these findings were suggestive of RP STS. On CECT thorax, there were multiple heterogeneously enhancing variable-sized soft tissue lesions in bilateral lung parenchyma involving mediastinum and visceral pleura suggestive of metastasis. Additionally, filling defects were observed in right branch of main pulmonary artery with extension to subsegmental branches of right middle and lower lobe suggestive of pulmonary thrombosis.

CT-guided core biopsy of the tumor was performed, and the microscopic examination demonstrated moderately cellular lesion composed of uniform, long, slender, atypical spindle-shaped fibroblasts arranged in bands, and fascicles separated by varying amount of fibrous stroma (►**Fig. 3**). The cells had moderate cytoplasm and slender nuclei. Additionally, the cells revealed moderate pleomorphism and mitotic activity with absence of necrosis. The findings led to the definitive diagnosis of low-grade ASCLT, and the patient was initiated on methylprednisolone (500 mg intravenous [IV] on days 1–3), cyclophosphamide (300 mg IV on days 1–3), mesna (100 mg/mL IV on days 1–3), and vincristine (2 mg IV, day 4). Subsequently, the chemotherapy regimen was modified to adriamycin (60 mg/m$^2$ IV on days 1–4), ifosfamide (7500 mg/m$^2$ IV on days 1–3), and mesna (100 mg/ml IV on days 1–4). This was coupled with concomitant 50 Gy radiotherapy (in 25 fractions). Following completion of the concurrent chemoradiotherapy and shrinkage of the tumor size, the patient underwent complete surgical excision of the tumor. The postoperative recovery was uneventful, and the patient was discharged in stable condition with advice to follow-up at 6 months.

## Discussion

RPS have an overall expected yearly incidence of 0.5 to 1 per 100,000 individuals.[8] LS is the most frequent type of STS, and includes four major histological types, including atypical lipomatous tumor/well-differentiated LS, myxoid/round cell LS, dedifferentiated LS, and pleomorphic LS.[11] Additionally, the well-differentiated LS has five variants, including sclerosing, lipoma-like, inflammatory, liposarcoma with meningothelial whorls, and spindle cell.[1]

Reported initially by Weiss and Enzinger (1896), SCLS arises from a spectrum of tumors, including fibrosarcoma,

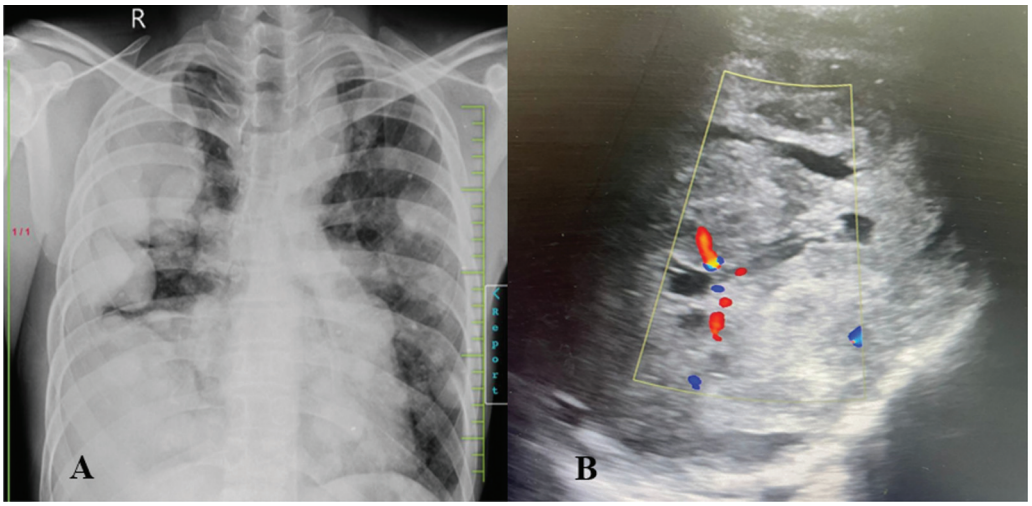


**Fig. 1** Chest radiography illustrating right lower zone consolidation with multiple variable size nodular opacities in bilateral middle and lower zones suggestive of metastatic lesions, with right pleural-based large metastatic lesions observed in periphery (**A**). Abdominal ultrasonography illustrating well-defined heterogenous predominantly hyperechoic lesion with minimal vascularity noted on Doppler (**B**).

 

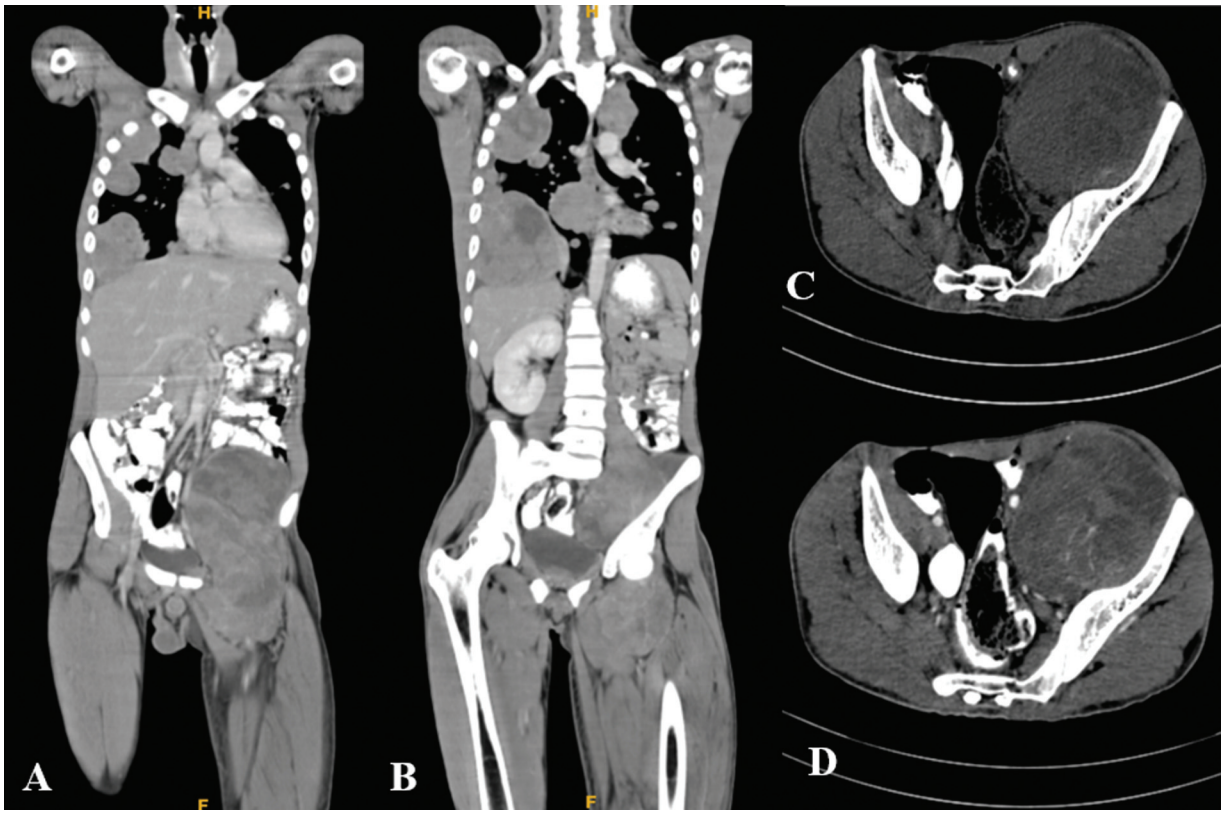


**Fig. 2** Contrast-enhanced computed tomography (CECT) abdomen: Coronal view (**A**); sagittal view (**B**); axial view, noncontrast (**C**); and axial view contrast, arterial phase (**D**). CECT abdomen illustrating a large, well-defined heterogeneously enhancing soft tissue density lesion (23 × 13 cm) with internal necrotic areas within the pelvis on left side involving left psoas and iliacus muscles, most likely retroperitoneal in origin. Anterosuperiorly invading iliopsoas muscles, abutting left rectus abdominis anteriorly with loss of fat planes and is displacing bowel loops laterally.

intraabdominal desmoid tumor, and gastrointestinal stromal tumor. Though SCLS does not have predilection for any sex and age, it mainly affects young adults aged 20 to 40 years, and occasionally diagnosed in children.[6] Likewise, our patient was 22 years old, and belonged to the most frequently affected age group.

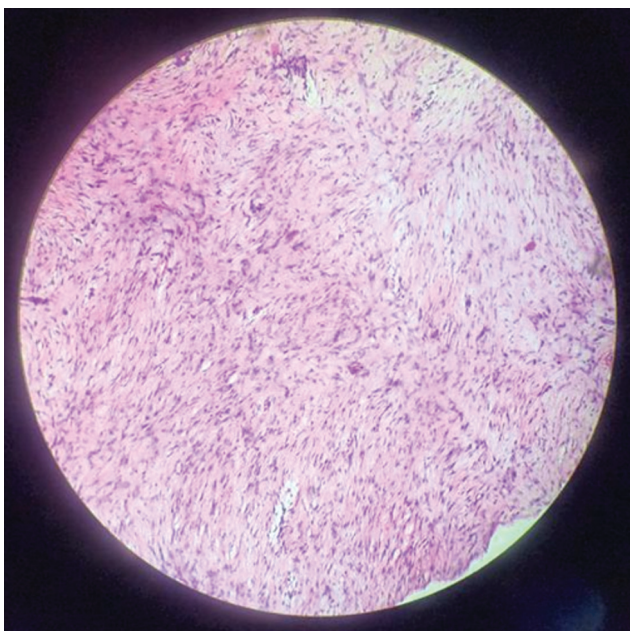

**Fig. 3** Microscopic examination of biopsy sample illustrating uniform, long, slender, spindle-shaped fibroblasts arranged in bands and fascicles separated by varying amount of fibrous stroma.

History, imaging, and biopsy are required to reach the diagnosis. As sarcomas arise from RP space, they have the propensity to grow enormously without producing symptoms resulting in late presentation.[7] Apart from asymptomatic abdominal mass, observed in 80% patients, a patient may present with nonspecific symptoms, related to mass effect or local invasion, including gastrointestinal obstruction, pain, abdominal fullness, and weight loss,[1,7] thus easily ignored leading to late diagnosis. Additionally, neurologic and musculoskeletal symptoms are referred to the lower limbs.[1] Likewise, our patient presented with asymptomatic, gradually progressive abdominopelvic mass. Also, the patient complained of weight loss, and pain radiating to left lower limb.

Magnetic resonance imaging is preferred for the evaluation of STS involving head, neck, trunk, and extremities. However, in a resource constraint setting, like ours, CECT very well illustrates the extent of STS and is helpful in reaching the diagnosis. Imaging also helps in obtaining the biopsy, and reaching the definitive diagnosis, as observed in our patient. Additionally, patients with STS should be evaluated with CT chest at the time of diagnosis and during follow-up.[5] Distant metastasis is infrequent, observed in 5% patients, while the

 

local disease is mostly associated with mortality.[9] In addition to abdominopelvic swelling, our patient had respiratory complaints, and thus CECT chest was performed, resulting in diagnosis of lung metastasis.

Complete surgical resection with R0 margin is desired, and feasible in 80 to 88% patients. To achieve complete resection of the tumor, 57 to 77% patients require resection of adjacent structures.[9] However, recurrence of sarcoma following primary surgical resection is a constant issue. Around 69 to 80% patients with first recurrence tend to have multiple local recurrences.[12] The chances of local recurrence are reduced, and survival is improved in patients with low-grade tumors and macroscopic clearance.[9] Neoadjuvant therapies (chemotherapy, radiotherapy, or their combination) are safe in well-selected patients, and may be considered only after a careful review. In our patient, owing to giant size of ASCLT, neoadjuvant chemoradiotherapy was initiated with an aim to shrink the tumor size. Subsequently, the patient underwent complete surgical excision of the tumor, and advised a regular follow-up at 6 months, to look for any recurrence. However, the survival advantage associated with adjuvant or neoadjuvant therapies is still unclear.

Literature related to the extension of RP and pelvic STS to the thigh is scarce. In 2001, Lewis et al described the extension of STS to the thigh in 11 patients through various routes, including obturator foramen ($n = 5$), inguinal ligament or spermatic cord ($n = 3$), sciatic notch ($n = 2$), and multiple routes ($n = 1$).[10] Subsequently, in 2011, Salemis et al reported a patient with RP LS extending to the thigh along the course of the iliopsoas muscle.[9] Likewise, in our patient, the SCLS extended from RP region to thigh along the iliopsoas muscle. However, unlike our patient, available literature does not mention about variants of LS, thus making the findings of our patient unique.

## Conclusion

In conclusion, due to rare nature, literature pertaining to ASCLT is still scarce. Their management requires tertiary care setting with multitherapy expertise. Though complete resection is the gold standard of care, recurrence remains a persistent issue. Role of adjuvant and neoadjuvant therapies requires further research, and CT chest should be performed at the time of diagnosis as well as during follow-up.

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Prior Presentation of Manuscript
None.

Authors' Contributions
P.K., S.K., P.P., V.N., and P.S. conceptualized the study and critically reviewed the study; P.K., S.K., and P.P. designed the study; S.K., P.P. supervised the study and provided material; P.K., V.N., and P.S. helped in data collection and/or processing, literature search, writing, and analysis and/or interpretation.

Declaration of the Patient Consent
Consent to write and report this case was obtained from the patient.

Conflict of Interest
None declared.

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