Leiomyosarcoma of the Mediastinum Depicted on CT Images: A Rare Origin Case and Literature Review

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AUTHOR CONTRIBUTIONS

First author: wrote and revised; second author: revised; Corresponding author conceptualization, review and editing

BACKGROUND

Primary mediastinal leiomyosarcoma remains an exceptionally rare malignancy (<15 cases in 20 years), posing significant diagnostic challenges through nonspecific symptoms and imaging features. Reporting this case is vital to enhance recognition of its clinical and imaging patterns, ultimately aiding earlier pathological confirmation and management of this aggressive neoplasm.

CONSENT

Informed written consent was obtained from the patient for the publication of this case report and accompanying images. Every effort has been made to ensure anonymity.

CONFLICT OF INTEREST

The authors declare no competing interests.

HUMAN AND ANIMAL RIGHTS

This study was conducted in compliance with ethical standards and did not involve any experiments on animals.

ABSTRACT

Introduction: Leiomyosarcoma (LMS), a malignant smooth muscle tumor, accounts for 10%–20% of soft tissue sarcomas. Primary mediastinal LMS is rare, with only 13 cases reported in the last two decades. This study aims to delineate its clinical and radiological characteristics through a novel case presentation and literature review. Methods: We report the case of a 57-year-old male presenting with progressive dyspnea and back pain. Diagnostic workup included contrast-enhanced chest CT, histopathological analysis of CT-guided biopsy specimens, and immunohistochemical staining (positive for Vim, Des, SMA, and Calponin; negative for S100 and CD34). A systematic PubMed review of mediastinal LMS cases was performed to summarize imaging and pathological features.

Results: Imaging revealed a 16.1 × 10.9 cm heterogeneous posterior mediastinal mass with necrotic foci, vascular encasement, and right pleural effusion. Histopathology showed spindle-shaped tumor cells with a Ki-67 index of 40%, confirming LMS. Literature synthesis (n=14 cases) demonstrated that 71.4% of tumors exceeded 5 cm, with a predilection for the anterior mediastinum (71.4%) and heterogeneous enhancement (100%).

Discussion: Mediastinal LMS radiologically mimics other malignancies but exhibits distinctive pathological markers (SMA/Desmin+). Surgical resection remains the primary treatment; our patient received neoadjuvant chemotherapy (liposomal doxorubicin/dacarbazine) and achieved a partial response. This case highlights the importance of multidisciplinary collaboration (imaging-pathology correlation) for the timely diagnosis of this aggressive neoplasm.

CASE REPORT

CASE REPORT

Basic Information

A 57-year-old man presented to the Emergency Department of West China Hospital with a 20-day history of progressive dyspnea and back pain of unknown etiology. Contrast-enhanced chest CT demonstrated a mediastinal mass with moderate right pleural effusion. Physical examination showed an alert and oriented patient with normal pupillary light reflexes. Auscultation revealed asymmetrically diminished breath sounds over the right hemithorax without rales. Neurological examination documented intact muscle strength in all extremities, and no peripheral edema was observed.

Laboratory Inspection

Laboratory tests revealed thrombocytosis (platelet count 378×10⁹/L; reference range: 100-300×10⁹/L), prolonged prothrombin time (13.5 s; normal range: 9.6-12.8 s), hyperfibrinogenemia (5.67 g/L; reference range: 2.0-4.0 g/L), and elevated D-dimer (4.06 mg/L FEU; cutoff: <0.55 mg/L). Liver and renal function tests were within normal ranges.

Imaging Examination

Contrast-enhanced chest CT demonstrated a large (16.1 × 10.9 cm) heterogeneous soft tissue mass in the posterior mediastinum, extending from the subcarinal region to the esophageal hiatus. The lesion exhibited mixed attenuation (unenhanced CT ~40HU) with heterogeneous enhancement (post-contrast ~80HU), containing multiple hypodense necrotic foci and tortuous feeding vessels□partially originating from the descending aorta. The mass caused significant compression of the esophagus, pulmonary veins, and bilateral atria, with ill-defined margins adjacent to the descending aorta. Secondary findings included partial narrowing of right bronchial branches with associated atelectasis, scattered pulmonary fibrotic changes, and a 5 mm nodule in the left lower lobe. No lymphadenopathy, cardiomegaly, or pericardial effusion was identified. Moderate right pleural effusion was present (Figure 1).

Pathological Examination and Diagnosis

A needle biopsy was performed. Histopathological examination demonstrated proliferating spindle-shaped atypical cells without significant necrosis. Immunohistochemical staining revealed the following profile□Vim(+)□Des (+) □SMA(+)□Calponin(+)□S100(-)□CD34(-)□stat(□)□PCK (-)□TLE1(-).The Ki-67 proliferative index was approximately 40%. The morphological and immunohistochemical findings supported a diagnosis of spindle cell sarcoma, most consistent with leiomyosarcoma. Clinicopathological correlation was recommended.

Chemotherapy and Surgical Findings

After thorough discussion, the patient underwent chemotherapy with liposomal doxorubicin and dacarbazine. The patient experienced nausea, vomiting, and decreased appetite, which improved with supportive care. A follow-up CT one month later showed a reduction in tumor size.

DISCUSSION

Leiomyosarcoma (LMS) is a highly malignant tumor originating from smooth muscle tissue and classified as a subtype of soft tissue sarcoma. It is characterized by aggressive biological behavior with a high tendency for local recurrence and distant metastasis. LMS accounts for approximately 10%–20% of all soft tissue sarcomas. It predominantly affects middle-aged and older adults, with a peak incidence between 40 and 60 years of age. There is a slight female predominance, particularly in cases arising from the uterus [1].

LMS can occur in any tissue containing smooth muscle, including the uterus, gastrointestinal tract, vascular walls, retroperitoneum, and deep soft tissues of the extremities. Leiomyosarcoma of the mediastinum is rare and represents a small proportion of primary malignant tumors of the mediastinum [2].

A PubMed search identified 13 reported cases of primary mediastinal LMS over the past two decades [3-15] (Table 1). Imaging data were available in 12 of the 13 cases with patients aged 41–85 years (6 females, 7 males). Among 14 total cases including the current one, 4 presented with chest/back pain, 5 with dyspnea, 5 were asymptomatic and detected incidentally, and 1 had hoarseness.

Imaging Features:

Among the 14 reported cases, including the present one, all lesions were located in the mediastinum. The anatomical distribution demonstrated predilection for the anterior mediastinum (n=10), followed by posterior (n=3; including our case) and middle mediastinum (n=1). Tumor dimensions ranged from 1.5 to 18 cm in maximal axial diameter (median 7.5 cm), with 71.4% (10/14) exceeding 5 cm in long-axis measurement, with poorly defined margins and exerted mass effect on adjacent structures. Contrast-enhanced scans typically showed heterogeneous enhancement.

Current evidence from this case and literature review indicates that mediastinal LMS can develop in all mediastinal compartments (anterior/posterior/middle).They often appear adjacent to or originate from major vessels (such as the superior vena cava or aorta) or the esophagus. The tumors are usually solid, irregular or lobulated in shape, and large in size (commonly >5 cm), with soft tissue density. The margins may be well- or ill-defined. Some cases show invasive growth patterns. Central necrosis or cystic degeneration is common, appearing as low-density areas within the mass, suggestive of tumor necrosis. Calcifications are rare and generally absent.

Contrast enhancement is typically mild to moderate and heterogeneous. In the arterial phase, some areas may show

marked enhancement, indicating a rich blood supply. In the venous or delayed phase, the enhancement tends to become more uniform or persistent, which may suggest a vascular smooth muscle origin. If the tumor arises from a vascular wall, deformity, narrowing, or occlusion of the vessel lumen may be observed.

Associated imaging findings may demonstrate compression or invasion of adjacent structures, such as displacement or deformation of the trachea, pericardium, or esophagus. In cases involving the superior vena cava (SVC), signs of SVC syndrome (e.g., facial edema, jugular vein distension) may be present. In advanced stages, pulmonary nodules or distant metastases may also be detected. In this case, because of large mediastinal mass, the adjacent pulmonary veins and both atria were compressed and narrowed, and he partial right lung atelectasis were showed also.

MRI typically shows iso- to hypointense signals on T1 and heterogeneous hyperintensity on T2, especially in necrotic areas. DWI reveals high signal in solid components, and post-contrast images show heterogeneous enhancement with non-enhancing necrosis. Tumors may compress or encase major vessels.

FDG-PET/CT shows high, heterogeneous uptake (SUVmax 5–15+), corresponding to viable tumor areas, with low uptake in necrotic or cystic regions [8]. It assists in detecting metastases, recurrence, and staging, but uptake is non-specific and requires pathological confirmation.

All 14 cases were confirmed as leiomyosarcoma via percutaneous or thoracoscopic biopsy, or postoperative histopathology and immunohistochemistry, with final diagnosis based on characteristic pathological features.

Histological Features

LMS consists of spindle-shaped cells in interlacing or fascicular patterns, with elongated nuclei and coarse chromatin. Nuclear atypia, pleomorphism, and mitotic figures, including atypical forms, are common and aid in grading. Tumor necrosis, often central and extensive, is frequent in aggressive cases. LMS is graded as low, intermediate, or high, with poor differentiation linked to worse prognosis.

Immunohistochemical Features: SMA(+), Desmin(+), H-caldesmon(+) — confirming smooth muscle origin. Ki-67 >10% — indicates high proliferative activity.S-100(–) — excludes neural tumors.CD117/DOG-1(–) — rules out gastrointestinal stromal tumors. Cytokeratin(–) — excludes epithelial tumors like thymomas or carcinomas.

Treatment Principles

The mainstay of treatment for LMS is surgical resection. Chemotherapy and radiotherapy may be used as adjuncts in selected cases. All cases reported in this article underwent surgical excision of the tumor.

CONCLUSIONS

This case highlights a rare instance of primary mediastinal leiomyosarcoma. Due to its nonspecific clinical and radiological features, diagnosis is often delayed until the tumor is large and compressing adjacent structures. However, imaging can offer valuable clues regarding malignancy, vascular origin, and invasion. Pathological confirmation remains essential for diagnosis and treatment planning. Enhanced awareness of its imaging and pathological characteristics may aid in timely diagnosis and management.

TEACHING POINT

Primary mediastinal leiomyosarcoma is rare but should be considered when CT reveals a large (>5 cm), lobulated, heterogeneously enhancing mass with necrosis and tortuous vessels, often compressing or invading adjacent structures. Diagnosis requires biopsy (spindle cells, SMA/Desmin/H-caldesmon(+), high Ki-67). Surgery is primary treatment; chemoradiation is adjunctive.

QUESTIONS

Question 1: On contrast-enhanced chest CT, which pattern is most characteristic of mediastinal leiomyosarcoma?

A□Homogeneous soft-tissue mass with smooth margins and fine calcifications

B□ Bulky lobulated mass showing marked heterogeneous enhancement with central non-enhancing necrotic areas

C□Well-defined fat-attenuation mass with thin internal septa

D□Thin-walled air-filled cyst abutting the trachea

Explanation: LMS is typically a large irregular mass that enhances unevenly because of viable tumor, interspersed necrosis, and hemorrhage

Question 2: What is the typical 18F-FDG PET/CT appearance of a high-grade mediastinal leiomyosarcoma?

A□Mild uptake (SUVmax < 3) throughout the mass

B□No uptake because of poor vascularity

C□Intense peripheral FDG uptake with a photopenic central zone corresponding to necrosis

D□Diffuse bone-marrow FDG uptake without uptake in the mass

Explanation: High-grade sarcomas are FDG-avid; central necrosis often lacks uptake, producing a “rim-and-hole” look

Question 3: Compression of which structure is most likely to cause dysphagia in patients with posterior mediastinal LMS?

A□Left main bronchus

B□Superior vena cava

C□Esophagus

D□Thoracic duct

Explanation: The esophagus runs immediately posterior to the left atrium in the posterior mediastinum; a large LMS in this compartment frequently compresses it, leading to swallowing difficulty.

Question 4: Which immunohistochemical profile confirms smooth-muscle lineage in this tumour?

A□Cytokeratin, TTF-1 and napsin-A positivity

B□S-100 and GFAP positivity

C□SMA, Desmin and H-caldesmon positivity with a high Ki-67 index

D□CD34 and STAT6 nuclear positivity

Explanation: Co-expression of smooth-muscle markers (SMA, Desmin, H-caldesmon) plus high proliferative activity (Ki-67) is diagnostic for leiomyosarcoma

Question 5: What is the recommended first-line treatment once a mediastinal leiomyosarcoma has been confirmed?

A□Observation only

B□Complete surgical resection

C□Neoadjuvant chemoradiotherapy without surgery

D□Systemic chemotherapy alone

Explanation: Radical en-bloc resection offers the highest chance of local control and long-term survival. Chemotherapy and/or radiotherapy are generally reserved for unresectable, residual, or recurrent disease rather than being used as definitive first-line therapy.

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FIGURES

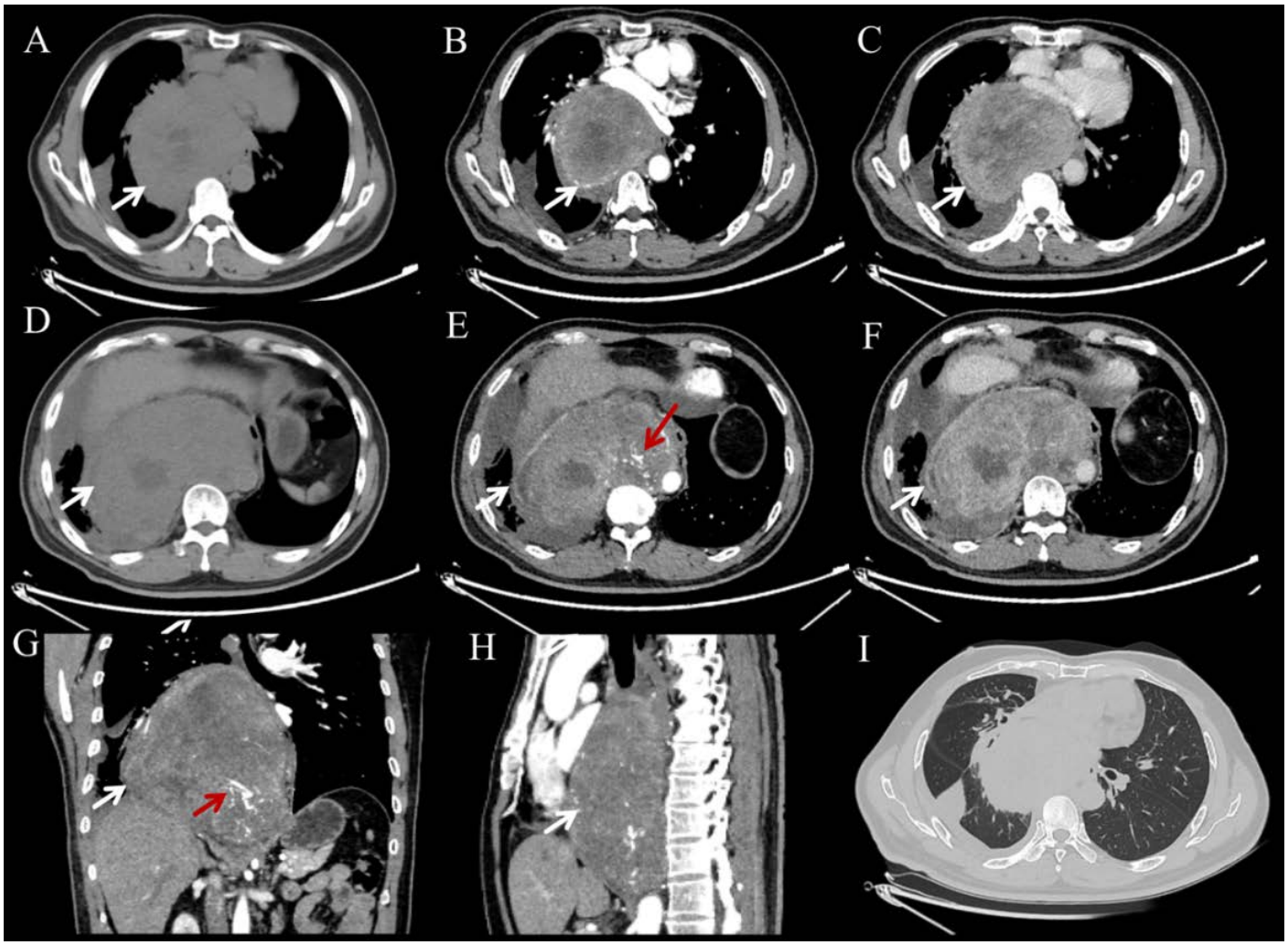

Figure 1: A: Non-contrast chest CT demonstrates a soft tissue mass located in the posterior mediastinum (white arrow indicates); B-C (Enhanced arterial and venous phases): The contrast-enhanced CT reveals a posterior mediastinal mass compressing the adjacent left atrium and pulmonary venous trunk (white arrow indicates); D: Non-contrast chest CT demonstrates a soft tissue mass located in the posterior mediastinum (white arrow indicates); E–H (Figures E, G and H show the arterial phase, while Figure F shows the venous phase): Contrast-enhanced chest CT (axial, coronal, and sagittal views) reveals a heterogeneously enhancing soft tissue mass in the posterior mediastinum (white arrow indicates), adjacent structures are compressed, and necrotic areas are observed within the lesion. Multiple small intralesional vascular structures are observed (red arrow indicates). The esophagus appears compressed and narrowed, and the interface between the lesion and the descending aorta is indistinct. I: Narrowing of a segmental bronchus in the right lung, accompanied by compression-induced consolidation and partial atelectasis.

Table 1: Leiomyosarcoma of the mediastinum

Study	Age (years)	Gender	Size (cm)	Location	Clinical features	Diagnosis method	Therapy	Outcome	Follow-up duration
Zhao H□et al. (2024)	59	Female	4.6	Posterior mediastinal paravertebral	chest and back pain	biopsy	Surgery, radio-therapy	No recurrence	4 months
Ishikawa M□et al. (2022)	79	Male	11	anterior mediastinum	an abnormal shadow on chest X-ray.	surgery	surgery	Mediastinal nodal recurrence was suspected	18months
Mancini M□et al. (2022)	79	Female	8	left mediastinum	cough, exertional dyspnea, sternal pain	biopsy	surgery	NA	NA
Collaud S□et al. □2022□	70	Female	11	middle mediastinum	cough and increasing dyspnea.	surgery	surgery	No recurrence	10months
Ishikawa A□et al. (2021)	85	Female	1.5	anterior mediastinum	Chest CT revealed a mediastinal mass	surgery	surgery	no recurrence or metastasis.	NA
Wu X□et al. (2019)	56	Male	NA	left middle mediastinum.	persisting chest tightness	surgery	surgery	NA	NA
Xue X□et al. (2018)	61	Male	5.2	anterior mediastinum	a mediastinal space-occupying lesion	Surgery	surgery	NA	NA
Iijima Y□et al. (2018)	77	Male	4.8	middle mediastinum	extrapleural sign in upper mediastinum on chest X-ray	surgery	surgery	Recurrent solid right pulmonary metastases (x2), both resected	NA
Vaziri M.□et al. (2012)	55	Female	18	right mediastinal	right-sided chest pain radiating to the back	Surgery	Surgery, radio-therapy, chemo-therapy	No recurrence	10months
Iwata T□et al.(2012)	66	Male	7	left upper mediastinal	hoarseness	biopsy	radio-therapy□Surgery	A solitary left pleural metastasis	12months
D'Aiuto M□et al. (2005)	62	Male	6	posterior mediastinum	occasional finding during cardiovascular examinations	Surgery	surgery	No recurrence	6months
Hirano H□et al. (2003)	60	Female	11.9	anterior mediastinum	dyspnea with exertion, cough and fever	Surgery	surgery	Tumor relapse	6months
Eroğlu A□et al.(2002)	41	Male	10	anterior mediastinum	dyspnea and chest pain	Surgery	Surgery, radio-therapy	No recurrence	8months

KEYWORDS

leiomyosarcoma: mediastinum: computed tomography: image feature

ABBREVIATIONS

LMS = LEIOMYOSARCOMA
CT = COMPUTED TOMOGRAPHY
SVC = SUPERIOR VENA CAVA
MRI = MAGNETIC RESONANCE IMAGING
FDG-PET/CT = FLUORODEOXYGLUCOSE-POSITRON EMISSION TOMOGRAPHY/COMPUTED TOMOGRAPHY
SUVMAX = STANDARDIZED UPTAKE VALUE MAXIMUM
VIM = VIMENTIN
DES = DESMIN
SMA = SMOOTH MUSCLE ACTIN
PCK = PAN CYTOKERATIN
TLE1 = TRANSDUCIN-LIKE ENHANCER PROTEIN 1

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