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Case Report

Primary Leiomyosarcoma of the Superior Vena Cava: A Rare Case Report

Andrey Ryabov[1], PhD; Stansilav Cygelnikov[1], PhD; Oleg Pikin[1], PhD; Oleg Aleksandrov[2], PhD; Vladimir Bagrov[1], PhD; Aleksandr Gusev[1], MD; Evgeniy Toneev[3], PhD; Viktoriya Surkova[1], PhD; Aznaur Dotdaev[1], MD

[1]P. Hertsen Moscow Oncology Research Institute (MORI), Branch of the Federal State Budgetary Institution "National Medical Research Radiological Centre" (NMRRC), Ministry of Health of the Russian Federation, Moscow, Russian Federation;
[2]University College London Hospitals NHS Foundation Trust (UCLH), London, United Kingdom;
[3]Department of Hospital Surgery, Anesthesiology, Reanimatology, Urology, Traumatology, and Orthopedics, T.Z. Biktimirov Faculty of Medicine, Institute of Medicine, Ecology and Physical Culture, Ulyanovsk State University, 42 Lev Tolstoy St., Ulyanovsk, 432071, Russian Federation

Correspondence:
Oleg Aleksandrov, PhD;
314 10 Elvin Gardens, Postal code: HA9 0GW, Wembley, London
Tel: +44 7383387626
Email: oleg.aleksandrov1@nhs.net
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What's Known

- Primary superior vena cava (SVC) leiomyosarcoma is extremely rare; fewer than 20 cases have been reported.
- Patients usually present with SVC compression syndrome—facial or upper-limb swelling, dilated neck veins, and dyspnoea.
- Due to its rarity, diagnosis relies on imaging and postsurgical histopathology.
- Biopsy is often unsafe.
- Radical *en bloc* resection with vascular reconstruction is the only treatment, and the role of adjuvant therapy remains undefined.

What's New

- Primary leiomyosarcoma of the superior vena cava is extremely rare and lacks standardized diagnostic and therapeutic guidelines.
- This case demonstrated successful radical resection using a xenopericardial graft and provided detailed intraoperative and histopathological information.

Abstract

Leiomyosarcoma of the superior vena cava (SVC) is an exceptionally rare vascular tumor. Fewer than 20 cases were reported worldwide. Because of the nonspecific nature of the presenting symptoms and their deep mediastinal location, a pre-operative diagnosis is often difficult or impossible. A 63-year-old woman presented with progressive facial and upper-limb edema. Computed tomography (CT) demonstrated a hyper vascular intraluminal mass originating in the SVC and extending towards the right atrium. Percutaneous biopsy was considered unsafe due to the risk of hemorrhage. Therefore, radical surgical resection of the SVC and brachiocephalic veins with xenopericardial patch reconstruction was performed. Histological and immunohistochemical examination confirmed a grade two leiomyosarcoma. No adjuvant therapy was given. At 8-month follow-up, there was no evidence of recurrence. Primary SVC leiomyosarcoma is a rare and diagnostically challenging condition. Pre-operative biopsy is often not feasible, and radical *en bloc* resection with vascular reconstruction remains the cornerstone of treatment, offering the best chance of recurrence-free survival. Histopathological confirmation is required to establish the diagnosis, and the role of adjuvant therapy remains uncertain.

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Introduction

Leiomyosarcomas arising from major blood vessels are a rare variant of soft-tissue sarcomas, accounting for less than 2% of all cases.[1] The inferior vena cava (IVC) is most frequently affected, with more than 400 cases reported.[2] In contrast, primary tumors of the superior vena cava (SVC) are far more uncommon, and fewer than 20 cases have been reported in the literature.[3] Patients typically present with signs of SVC compression syndrome–facial and upper-extremity swelling, dilated neck veins, and dyspnea. The differential diagnosis includes thrombosis, lymphoma, and other mediastinal tumors. Due to the extreme rarity of this condition, there are no standardized diagnostic or therapeutic guidelines. Treatment decisions are usually based on case reports and small case series. We reported a successful surgical treatment of a primary SVC leiomyosarcoma and review of the published literature, highlighting diagnostic and management difficulties.

Case Presentation

A 63-year-old woman presented in March 2024 with facial and bilateral upper-limb swelling discovered during the assessment of a hypertensive crisis. She was referred to the Moscow Oncology Research Institute of P.A. Hertsen (a branch of the National Medical Research Center for Radiology, Ministry of Health of the Russian Federation) for further evaluation. Her past medical history included hypertension, well-controlled type 2 diabetes mellitus, hyperlipidemia, atherosclerosis of the brachiocephalic arteries, and class I obesity. Previously, she underwent sigmoidectomy for complicated diverticulitis in 2020 and laparoscopic cholecystectomy in 2017. There was no prior history of malignancy.

On admission, the patient was alert with a Karnofsky performance status of 100 and an Eastern Cooperative Oncology Group (ECOG) performance status score of 0. Physical examination revealed facial and upper-limb edema and dilated superficial veins over the anterior chest and neck. Heart sounds were muffled, though regular. The lungs were clear on auscultation, and the abdomen was soft and non-tender. Urine output was preserved.

Contrast-enhanced computed tomography (CECT) of the chest demonstrated an intraluminal mass involving the SVC and left brachiocephalic vein, measuring 89 mm in length and 33×35 mm in cross-section. The lesion showed heterogeneous contrast enhancement and marked hypervascularity, with its caudal margin 10 mm proximal to the right atrium (figure 1).

Ultrasonography of the brachiocephalic veins showed no thrombosis. Based on the clinical signs of SVC syndrome and imaging findings, a vascular tumor was suspected. Percutaneous biopsy was judged unsafe owing to the high risk of hemorrhage and proximity to the right atrium. Following multidisciplinary team discussion, a radical resection was planned.

On July 5, 2024, the patient underwent median sternotomy. The bulky tumor surrounding the SVC and extending towards the left brachiocephalic was identified after dissecting the thymic tissue. Firm on palpation, it appeared to be invading the lateral vessels without a clear dissection plane. Before clamping the SVC, a temporary bypass was established between the right internal jugular vein and the right atrium to prevent intracranial hypertension. The SVC and brachiocephalic veins were excised *en bloc*, and vascular continuity was restored with a xenopericardial graft (Biolab, Russia). The right brachiocephalic vein was anastomosed to the proximal stump of the SVC (figure 2). The postoperative course was uncomplicated, and the patient was discharged on day 11. Postoperative imaging confirmed patency of the graft.

Histological examination of the resected specimen showed an intraluminal spindle-cell tumor occupying the SVC. Microscopically, the tumor showed a fascicular growth pattern, moderate nuclear pleomorphism, focal necrosis involving less than 50% of the tumor area, and a mitotic count of 9 mitoses per 10 high-power fields (×400). The tumor cells infiltrated the entire thickness of the venous wall with extension into the perivascular adipose tissue. Immunohistochemistry demonstrated diffuse cytoplasmic expression of vimentin, desmin, and smooth-muscle actin (SMA); the Ki-67 index was 35%. Tumor cells were negative for S100 and CD34 (except for vascular endothelium), and there was no expression of epithelial or endothelial markers. These findings excluded angiosarcoma, neural tumors, epithelial neoplasms, and lymphomas and supported a diagnosis of grade two leiomyosarcoma (figure 3).

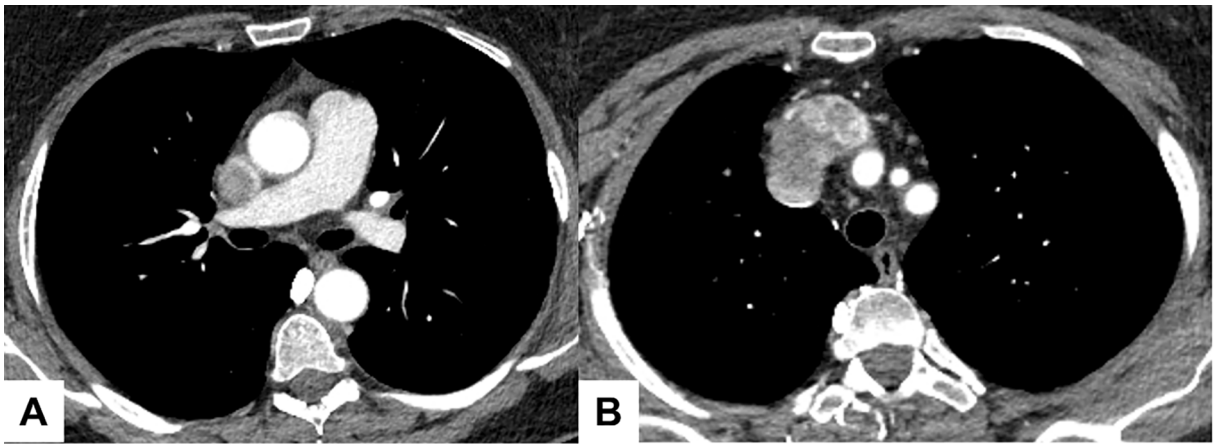

Figure 1: Contrast-enhanced computed tomography reveals a hypervascular intraluminal mass occupying the superior vena cava and brachiocephalic vein. (A) The contrast-enhanced computed tomography shows a hyper vascular intraluminal mass within the superior vena cava. (B) An axial computed tomography image demonstrates tumor extension into the brachiocephalic vein.

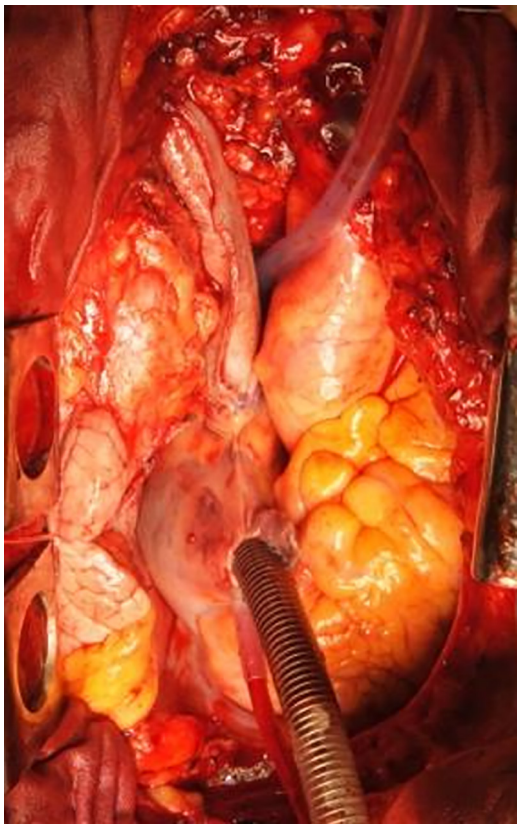

Figure 2: The figure shows the intraoperative view after reconstruction of the superior vena cava and brachiocephalic veins.

CECT performed 8 months after surgery revealed a patent graft of the reconstructed SVC without thrombosis or recurrent tumor. The patient remained asymptomatic and required no adjuvant therapy. Written informed consent was obtained from the patient to publish this case report and any accompanying images.

Discussion

Primary leiomyosarcoma of the SVC is an extraordinarily rare malignancy arising from smooth-muscle cells of the venous wall. Due to the rarity of this disease, clinical knowledge is derived only from small case series. To date, no systematic review has been conducted, which significantly complicates the development of standardized diagnostic and therapeutic approaches for this localization.

Previously reported cases of SVC leiomyosarcoma describe heterogeneous clinical presentations, ranging from incidental detection to classic features of SVC syndrome, including facial and upper-limb edema and dyspnea.[3, 4] Such patients are often misdiagnosed with venous thrombosis and empirically receive thrombolytic therapy without a proper understanding of the underlying cause.

Modern imaging modalities, especially

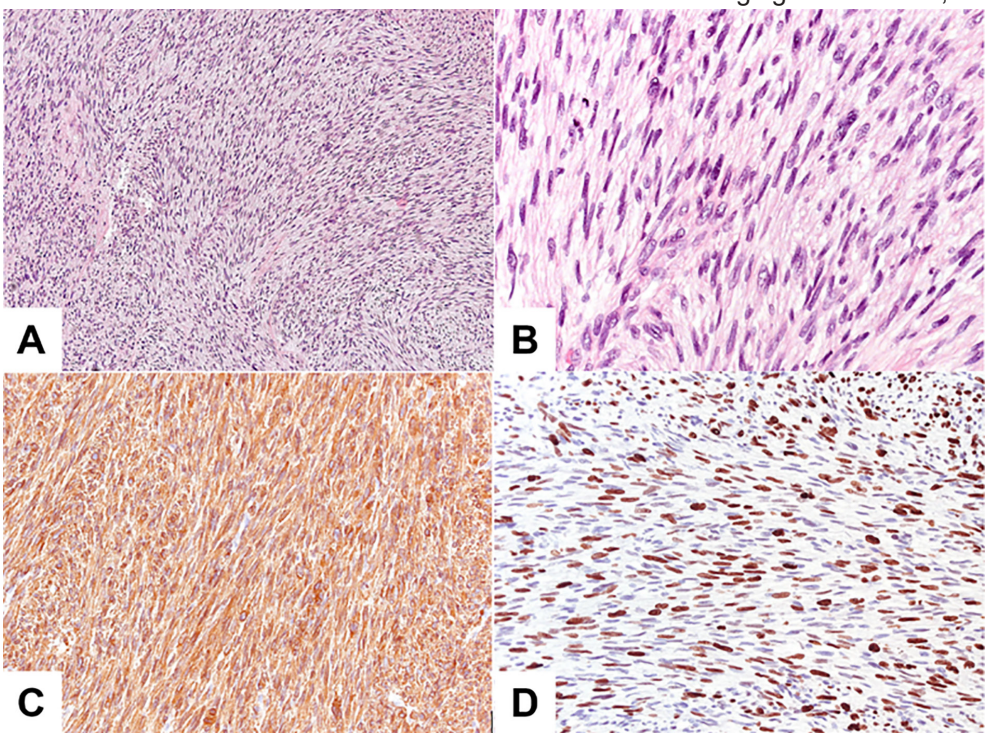

Figure 3: Histopathological and immunohistochemical features confirm a spindle cell neoplasm with smooth muscle differentiation and moderate proliferative activity. (A) Histopathological examination shows intersecting fascicles of spindle cells with moderate pleomorphism. Hematoxylin and eosin staining at ×100 demonstrates fascicular architecture of spindle-shaped tumor cells with infiltration of the venous wall. (B) Hematoxylin and eosin staining at ×400 shows spindle cell morphology, moderate pleomorphism, and mitotic figures (9 per 10 high-power fields). (C) Immunohistochemistry for smooth muscle actin at ×200 reveals diffuse cytoplasmic positivity, indicating smooth muscle differentiation. (D) Immunohistochemistry for Ki-67 at ×200 shows a proliferation index of approximately 35%, indicating moderate proliferative activity.

CECT and magnetic resonance imaging (MRI), allow accurate delineation of tumor extent and vascular involvement and guide surgical planning. According to the Delphi consensus published by Balaz and others in 2025, a preoperative assessment strategy combining CT and MRI is recommended for suspected tumors involving the IVC or SVC.[5] The consensus also highlighted the importance of multidisciplinary evaluation involving clinical oncologists, vascular surgeons, radiologists, and, in cases involving the SVC, cardiothoracic or vascular surgeons. Percutaneous biopsy is rarely feasible due to the risk of hemorrhage and anatomical inaccessibility. Existing catheter-based endovascular biopsy techniques remain technically limited and are not widely applicable in such scenarios. Definitive diagnosis is therefore established on the resected specimen.[6] In the present case, CT findings revealed characteristic features of a vascular-origin tumor, including intraluminal growth, hyper-vascularity, and proximity to the right atrium. These findings supported the decision to avoid biopsy and instead proceed directly with radical surgical intervention.

The definitive diagnosis of leiomyosarcoma is established by histological and immunohistochemical examination. The tumor is characterized by a spindle-cell component arranged in fascicles, moderate pleomorphism, and focal necrosis, positive expression of SMA, and absence of S100, CD34, and CD31 markers.[7]

Complete surgical removal remains the only treatment modality associated with the best oncologic outcomes for leiomyosarcomas.[2] In our case, complete resection of the SVC and brachiocephalic veins with xenopericardial graft reconstruction allowed an R0 resection.

Leiomyosarcomas, including vascular variants, are known for their poor sensitivity to systemic therapy. Classical anthracycline-based chemotherapy (doxorubicin, ifosfamide) has shown limited activity, with overall response rates rarely exceeding 20-30%, thereby making it unsuitable as monotherapy in localized disease.[7] Research is ongoing into targeted agents, such as mTOR inhibitors (rapamycin), antiangiogenic drugs (bevacizumab), histone deacetylase inhibitors, and poly (ADP-ribose) polymerase (PARP) inhibitors for tumors with impaired DNA repair mechanisms.[8] Promising directions include personalized molecular therapies guided by tumor genomic profiling, such as identification of TP53, RB1, and PTEN mutations, which are commonly observed in leiomyosarcomas.[9] However, these approaches remain experimental and require further clinical validation in clinical trials.

In the present case, the absence of metastasis and achievement of complete surgical resection supported the decision to omit adjuvant therapy, aligning with current management strategies for localized vascular leiomyosarcomas. The patient remains disease-free at 8 months.

Conclusion

Leiomyosarcoma of the SVC is a rare and diagnostically challenging tumor. Patients typically present with symptoms of SVC compression. However, imaging is essential to establish the extent of the disease. Percutaneous biopsy is usually avoided due to the risk of hemorrhage, and diagnosis is confirmed histologically after surgical resection. Vascular reconstruction with xenopericardial grafts allows for the achievement of complete resection and avoids thrombotic events. The role of adjuvant therapy remains undefined; nevertheless, current evidence suggests that complete surgical resection offers the best chance of recurrence-free survival.

Authors' Contribution

A.R: Conceptualization, supervision, surgical management of the patient and critical revision of the manuscript; S.C: Supervision, surgical management of the patient and critical revision of the manuscript; O.P: Conceptualization, supervision, surgical management of the patient and critical revision of the manuscript; O.A: Conception and design of the work, analysis, drafting the manuscript; V.B: Conception and design of the work, analysis, critical review of the manuscript; A.G: Conception and design of the work, analysis, anaesthetic management of the patient, critical review of the manuscript; E.T: Conception and design of the work, analysis, critical review of the manuscript; V.S: Review of pathology section, drafting, editing and reviewing the manuscript; A.D: Data verification, references, and final manuscript formatting. All authors have read and approved the final manuscript and agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Declaration of AI

The authors declare that no AI tools were used in the preparation of this manuscript.

Conflict of Interest: None declared.

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