Critical Limb Ischemia as a Rare Presentation of Malignant Solitary Fibrous Tumor of the Pleura

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Patient: Female, 57-year-old
Final Diagnosis: Malignant solitary fibrous tumor of the pleura • tumor embolism
Symptoms: Bilateral numbness and tingling
Medication: —
Clinical Procedure: —
Specialty: Cardiac Surgery • General and Internal Medicine • Oncology • Pathology • Radiology

Objective: Rare disease
Background: Solitary fibrous tumor (SFT) of the pleura is a rare fibroblastic neoplasm. It is commonly found incidentally on imaging and is usually benign but has significant potential to recur as a malignant tumor. Patients present asymptomatic or with pulmonary symptoms such as cough or shortness of breath. Cardiac invasion of an SFT can create an avenue for peripheral tumor embolization and critical limb ischemia, as in this case report. There is no prior published report of recurring malignant SFT presenting as critical limb ischemia.

Case Report: We report a rare presentation of malignant SFT recurrence in a 57-year-old woman with critical limb ischemia of both lower extremities secondary to bilateral tumor emboli. The patient’s primary tumor was treated with surgical resection alone. Upon recurrence, the tumor growth was so extensive that it was no longer amenable to surgical resection at the time of her critical limb ischemia. The patient presented with bilateral numbness and tingling, without any pulmonary symptoms.

Conclusions: Although it is sporadic, clinicians should know that an aggressive malignant SFT can embolize and present as critical limb ischemia. The possibility of tumor emboli provides a pressing reason to surgically resect SFT masses in their early stages before any cardiac invasion.

Keywords: Solitary Fibrous Tumor, Pleural • Embolism • Solitary Fibrous Tumors • Neoplasm Metastasis • Angiography, Digital Subtraction

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Background

A solitary fibrous tumor (SFT) is a rare mesenchymal tumor of fibroblastic origin that involves the pleura in 30% of cases [1]. SFT usually presents in older adults between ages 60 and 70 years old with pulmonary symptoms such as cough, shortness of breath, chest pain, pneumothorax, or hemothorax [2]. Overall, most patients with SFT are asymptomatic, and thus most cases are discovered incidentally on imaging. Diagnosis is confirmed by pathology. Although mostly considered benign, up to 30% of these tumors transform to malignant, and thus early detection is vital [3]. Presenting symptoms of SFT are usually nonspecific and variable, especially since SFT can present with local disease, locally invasive disease, and metastatic disease in rare cases [4]. This case presentation focuses on the sporadic presentation of critical limb ischemia caused by tumor emboli in a 57-year-old woman.

Case Report

A 57-year-old woman with a history of left-sided malignant thoracic SFT status after resection in 2018 with no history of chemotherapy or radiation treatment presented to the Emergency Department with sudden onset of bilateral lower extremity numbness and weakness. Her associated symptoms were notable for an ongoing non-productive cough, shortness of breath, intermittent palpitations, and myalgias in the months leading up to her hospitalization. She was being evaluated on an outpatient basis for a new left-sided 11-cm chest mass (Figure 1) that was presumed to be a recurrence of SFT but was still awaiting tissue diagnosis. Her past medical history included hypertension, prediabetes, COVID-19, and epilepsy, with her last grand mal seizure occurring over 20 years ago. The patient was a nonsmoker, and her family history was noncontributory.

Physical exam findings were significant for a complete absence of left-sided lung sounds and decreased sensation in her bilateral lower extremities. Pedal pulses were intact, and the bilateral lower extremities were pink and warm. Blood pressure and heart rate were elevated at 241/134 mmHg and 145 beats per minute, respectively. Oxygen saturation was 94% on room air.

Several hours later, the patient’s left toes appeared cyanotic, and her left lower extremity felt cool upon palpation. While the bilateral venous duplex ultrasound was negative, the arterial duplex showed probable obstruction of the popliteal arteries (Figure 2A). An angiogram later confirmed bilateral popliteal artery occlusions (Figure 2B), which were likely acute in nature given the lack of other chronic vascular disease or collateralization. Due to significant pain, the patient could not tolerate percutaneous thrombectomy during angiography.

Figure 1. (A) Fused coronal PET/CT shows the large hypermetabolic left chest mass (light yellow arrow) as well as a contralateral metastatic pulmonary nodule (blue triangle). (B) Although the heart is typically hypermetabolic on PET, there is suspicion of invasion of the left basilar ventricle on axial fused PET/CT imaging (blue arrow).
Figure 2. (A) Color Doppler image of the left popliteal artery shows no flow within the artery and no waveform detected within the cursor. The ABI of the left lower extremity was less than 0.5. (B) There is confirmation of the popliteal occlusion on the left lower extremity angiography. (C) A post-thrombectomy angiogram shows a restored flow in the popliteal with a two-vessel runoff to the foot, with mild residual non-occlusive thrombus remaining.
had to be taken to the operating room for immediate surgical revascularization with bilateral thrombectomy and fasciotomy. Additional oncology workup included a chest, abdomen, and pelvis computed tomography (CT), significant for a hypervascular tumor filling the left chest with parasitic recruitment of the left inferior phrenic, intercostal, and pulmonary arteries (Figure 3). Imaging findings demonstrated that the tumor directly invaded the left ventricle, making this a site suspicious for the origin of the tumor emboli. Smaller metstatic foci in the contralateral lung and left superior phrenic mass were also seen.

Postoperative angiography revealed restored popliteal flow (Figure 2C), corroborating the regained sensation in bilateral lower extremities. Histopathological examination of the unusually milky thrombus showed hypercellular tumor emboli with numerous mitotic figures (Figure 4A, 4B), aligned with the nuclear pleomorphism seen in malignant tumor cells (Figure 4C). Immunohistochemical stains were positive for STAT6 and CD34, which are sensitive SFT markers (Figure 5A, 5B) [5]. The patient was discharged a few days later and transferred to a tertiary care facility for further evaluation and cardiovascular surgical considerations. The chest mass was ultimately deemed unresectable due to its size and local invasion, and the patient was given a 1 to 2-year prognosis pending immediate chemotherapy. She was referred to palliative care for further management.
Discussion

This case documents the first presentation of a recurrent SFT as critical limb ischemia. It highlights the potential for malignant SFT to generate emboli and limb-threatening ischemia when left untreated. Peripheral arterial embolization of tumors is a rare etiology for limb ischemia and is an uncommon initial presentation of cancer. This patient's lung mass was delayed and may have contributed to this advanced presentation. While most cases of SFT are asymptomatic, the possible generation of tumor emboli provides an important reason for immediate treatment via resection. While surgery is the mainstay of treatment, neoadjuvant radiation or chemotherapy and adjuvant radiation are included on a case-by-case basis [6]. Anthracyclines are the chemotherapeutic of choice, although more data are needed to establish definitive outcomes [6]. Due to high vascularity, embolization is sometimes performed before resection, and antiangiogenic treatments are also a new option.

Cases of tumor emboli as the presenting disease symptom have been documented for malignant melanoma, germ cell tumor, lung cancer, and other cancers. Tumor emboli generally occur due to cardiac involvement of a pulmonary tumor, with few cases due to metastatic invasion of vessels or other
extracardiac involvement. In a similar case report, malignant fibrous histiocytoma presented as tumor emboli when part of the mass was directly attached to the thoracic wall, with aortic involvement creating a pathway for peripheral embolization [7]. Similarly, a metastatic parotid tumor presented with acute limb ischemia from left ventricle involvement and subsequent embolization [8].

In this case, CT demonstrates the tumor invading through the left ventricle, particularly in the sagittal view (Figure 3C). Nearby vasculature was recruited, including the pulmonary artery and vein, inferior phrenic artery, and intercostal vessels (Figure 3A, 3B). Cardiac involvement of the left ventricle probably provided the pathway for peripheral tumor emboli, but the recruitment of the pulmonary vessels, as demonstrated by CT angiography, can provide alternative pathways as well.

It is crucial to consider tumor emboli as a severe consequence of malignancy in patients with a known history of thoracic malignancy with evidence or potential for cardiac involvement. For patients presenting with symptoms of limb ischemia and typical chest pathology symptoms, malignancy and tumor emboli should be included in the differential diagnosis to prevent delays in diagnosis and resection of the primary mass. Because tumor emboli are likely a late-stage presentation of malignant SFT, the primary mass may not be amenable to surgical treatment, as in this case report. New research about the molecular signaling mechanisms that drive malignant SFT growth paves the way for advanced treatment options [6].

**Conclusions**

Critical limb ischemia may be the initial symptom of a recurrent malignant SFT, especially if treatment is prolonged. SFT can generate tumor emboli, providing a reason for immediate management of benign or malignant tumors. When pleural SFTs are left untreated, the cardiac invasion can lead to tumor emboli formation and manifest as critical limb ischemia secondary to peripheral embolization. Surveillance must be tailored to each patient to ensure early detection of recurrence in every patient presenting with SFT.

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**Declaration of Figures’ Authenticity**

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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