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CASE REPORT

When a pulmonary embolism is not a pulmonary embolism: a rare case of primary pulmonary leiomyosarcoma

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Arterial leiomyosarcomas account for up to 21% of vascular leiomyosarcomas, with 56% of arterial leiomyosarcomas occurring in the pulmonary artery. While isolated cases of primary pulmonary artery leiomyosarcoma document survival up to 36 months after treatment, these uncommon, aggressive tumors are highly lethal, with 1-year survival estimated at 20% from the onset of symptoms. We discuss a rare case of a pulmonary artery leiomyosarcoma that was originally diagnosed as a pulmonary embolism (PE). A 72-year-old Caucasian female was initially diagnosed with 'saddle pulmonary embolism' based on computerized tomographic angiography of the chest 2 months prior to admission and placed on anticoagulation. Dyspnea escalated, and serial computed tomography scans showed cardiomegaly with pulmonary emboli involving the right and left main pulmonary arteries with extension into the right and left upper and lower lobe branches. An echocardiogram on admission showed severe pulmonary hypertension with a pulmonary artery pressure of 82.9 mm Hg, and a severely enlarged right ventricle. Respiratory distress and multiorgan failure developed and, unfortunately, the patient expired. Autopsy showed a lobulated, yellow mass throughout the main pulmonary arteries measuring 13 cm in diameter. The mass extended into the parenchyma of the right upper lobe. On microscopy, the mass was consistent with a high-grade primary pulmonary artery leiomyosarcoma. Median survival of patients with primary pulmonary artery leiomyosarcoma without surgery is one and a half months, and mortality is usually due to right-sided heart failure. Pulmonary artery leiomyosarcoma is a rare but highly lethal disease commonly mistaken for PE. Thus, we recommend clinicians to suspect this malignancy when anticoagulation fails to relieve initial symptoms. In conclusion, early detection and suspicion of pulmonary artery leiomyosarcoma should be considered in patients refractory to anticoagulation, prompting initiation of early intervention.

Keywords: *pulmonary artery leiomyosarcoma*; *pulmonary embolism*; *dyspnea*; *right-sided heart failure*

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Arterial leiomyosarcomas account for up to 21% of vascular leiomyosarcomas, with 56% of arterial leiomyosarcomas occurring in the pulmonary artery (1). While isolated cases of primary pulmonary artery leiomyosarcoma document survival range up to 36 months after treatment (2), these uncommon aggressive tumors are highly lethal, with 1-year survival estimated at 20% from the onset of symptoms (3). We discuss a unique case of a pulmonary artery leiomyosarcoma that was originally diagnosed as a pulmonary embolism (PE).

Case report

A 72-year-old Caucasian female who was initially diagnosed with 'saddle pulmonary embolism' based on an outpatient computed tomography (CT) chest angiogram approximately 2 months prior to the current presentation and subsequently placed on warfarin for anticoagulation presented with persistent dyspnea. Her dyspnea had been progressively worsening during this time, and serial CT scans showed cardiomegaly with pulmonary emboli involving the right and left main pulmonary arteries with extension into the right and left upper and lower lobe branches (Fig. 1). The CT scans also showed a 47×42 mm right upper lobe paratracheal mass that had significantly increased in size over the 2-month period prior to this presentation. An echocardiogram shortly after admission showed severe pulmonary hypertension with a pulmonary artery pressure of 82.9 mm Hg, and a severely enlarged right ventricle. The patient developed respiratory distress and was placed on mechanical

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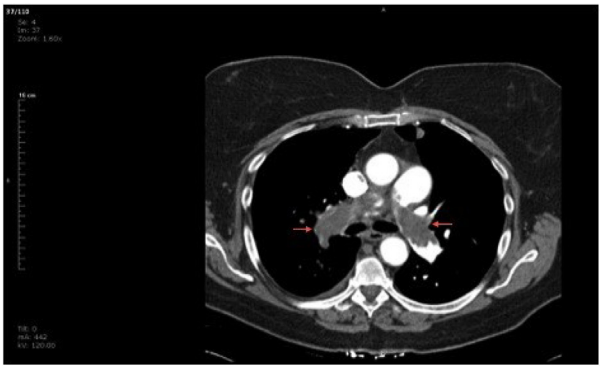

Fig. 1. Extensive intravascular filling defects of bilateral pulmonary arteries (red arrows).

ventilation. Multiple laboratory values at this time indicated multiorgan failure with worsening right ventricular function. Subsequently, the patient became hypotensive and bradycardic. Despite aggressive resuscitative measures, the patient expired shortly thereafter. A chest-only autopsy performed with the family's consent revealed biatrial dilatation, and the right ventricular cavity was dilated with a thickened wall. The interventricular septum was distorted and pushed toward the left ventricular cavity (Fig. 2). The lungs were markedly congested. A lobulated, pale, yellow mass occupied the entire volume of the right and the left main pulmonary arteries, loosely adherent to the pulmonary artery at multiple locations and measuring 13 cm in greatest dimension. The pulmonary artery mass extended into the parenchyma of the right upper lobe, 6 cm in greatest dimension (Fig. 3). On microscopy, this mass showed a highly malignant neoplasm, characterized by a sheet-like growth pattern with alternating areas of higher and lower cellularity; highly pleomorphic nuclei with abnormal mitotic figures, over 10 mitoses per 10 high-power fields; and extensive areas of necrosis. These cells

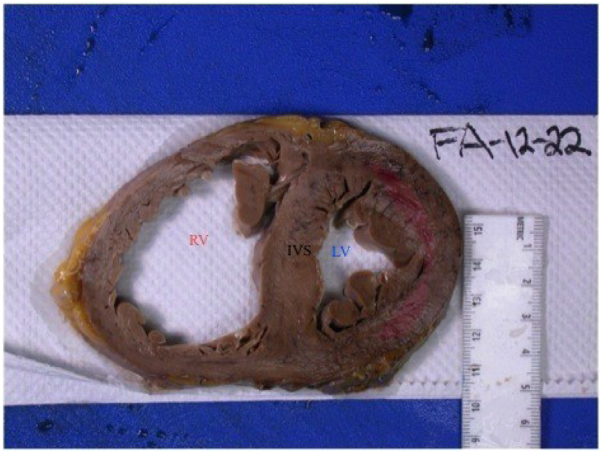

Fig. 2. The right ventricular (RV) cavity dilation with a thickened wall. The distorted interventricular septum (IVS) is pushed toward the left ventricular (LV) cavity.

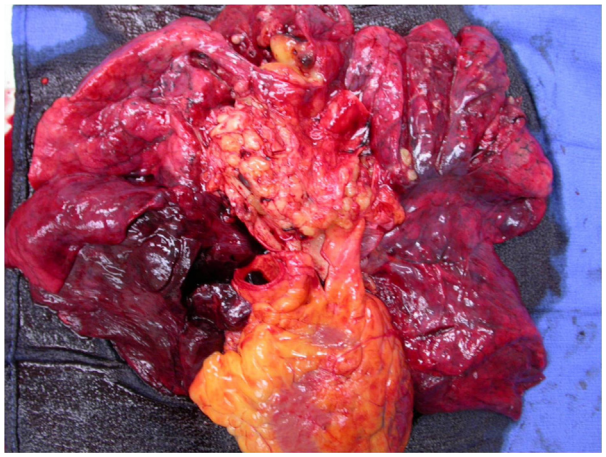

Fig. 3. The markedly congested lungs. A lobulated, pale, yellow mass is occupying the entire volume of the right and left main pulmonary arteries, loosely adherent to the pulmonary artery at multiple locations and measuring 13 cm in greatest dimension. The pulmonary artery mass extended into the parenchyma of the right upper lobe, 6 cm in greatest dimension.

were positive for desmin and vimentin and showed focal reactivity for actin. Overall, this was consistent with high-grade primary pulmonary artery leiomyosarcoma (Fig. 4).

Discussion

Pulmonary artery leiomyosarcoma is a rare disease – so much so that conflicting epidemiological data reflect rates of pulmonary artery sarcoma in general and not that of pulmonary artery leiomyosarcoma. More recent reviews suggest a predominance of two to one in women, with a median age at diagnosis cited between 49 and 52 years (4–6). Because of its rarity, it is often mistaken for chronic venous thromboembolic disease with similar symptoms of

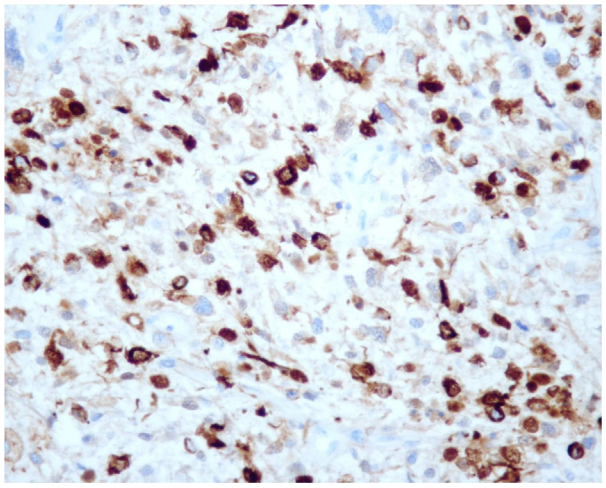

Fig. 4. The cells are positive for desmin and vimentin and showed focal reactivity for actin. This is consistent with a high-grade primary pulmonary artery leiomyosarcoma.

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dyspnea, chest pain, and right-sided heart failure. In addition, palpitations, syncope, cough, chest distress, and weight loss have been reported (4, 7–9). Radiographically, a wall eclipsing sign on a pulmonary CT angiogram is suggestive of pathogenicity for some pulmonary artery malignancies, such as sarcoma (10). While ventilation/perfusion scans make it difficult to differentiate between primary non-thrombotic intravascular lesions and PE (8), a fluorine-18 fluorodeoxyglucose positron emission tomography scan (18F-FDG PET/CT) has shown 18F-FDG uptake by malignant tumor and not by blood thrombi (8, 11). Magnetic resonance imaging may be able to differentiate a mass from thrombus based on signal homogeneity (12). Endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) has also been described as an effective tool to diagnose intraluminal malignant neoplasms in certain cases (13–17). Median survival without surgery is one and a half months (18), and mortality is usually due to right-sided heart failure. Pulmonary artery leiomyosarcoma is a rare but highly lethal disease commonly mistaken for PE (19, 20). Thus, we recommend clinicians to suspect this malignancy when anticoagulation fails to relieve the initial symptoms. In conclusion, early detection and suspicion of pulmonary artery leiomyosarcoma should be considered in patients refractory to anticoagulation, prompting initiation of early intervention.

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Consent

No personal patient information has been disclosed.

Disclosure

None of the authors have any financial or personal bias that would inappropriately compromise the publication of this work.

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