

Intimal Sarcoma of the Pulmonary Artery: a Rare Cause of Pulmonary Hypertension

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Intimal sarcoma of the pulmonary artery is a rare tumor that is usually diagnosed during surgery or autopsy. Such tumors are characterized by local growth, with only slight ability to metastasize. Diagnosis is difficult and often delayed owing to the nonspecific nature of the symptoms. Since intimal sarcoma of the pulmonary artery is so rare and insidious it is often confused with pulmonary thromboembolism and is therefore treated inappropriately with prolonged anticoagulation or thrombolysis. With a mean survival of 12 months from the onset of symptoms, the prognosis is poor. We present the case of a woman who was preoperatively diagnosed with intimal sarcoma of the pulmonary artery and who underwent surgical resection with no apparent recurrence at long term follow-up. A review of the literature is also included.

Key words: *Sarcoma. Pulmonary artery. Pulmonary hypertension.*

Sarcoma intimal de arteria pulmonar: una causa infrecuente de hipertensión pulmonar

El sarcoma intimal de la arteria pulmonar es un tumor infrecuente que habitualmente se diagnostica en el acto quirúrgico o la autopsia. Por lo general crece regionalmente, con poca capacidad de metastatizar. El diagnóstico es difícil y con frecuencia se retrasa debido a la naturaleza inespecífica de los síntomas. Su baja frecuencia y el crecimiento insidioso hacen que a menudo se confunda con una tromboembolia pulmonar, y por ello se trata inadecuadamente con anticoagulación prolongada o trombólisis. El pronóstico es malo, con una media de supervivencia de 12 meses desde el inicio de los síntomas. Presentamos el caso de una paciente a quien se diagnosticó preoperatoriamente un sarcoma intimal de la arteria pulmonar que se sometió a resección quirúrgica, sin que se evidenciase recurrencia en el seguimiento a largo plazo. También se realiza una revisión de la bibliografía.

Palabras clave: *Sarcoma. Arteria pulmonar. Hipertensión pulmonar.*

Introduction

Intimal sarcoma of the pulmonary artery is a rare tumor with a variety of clinical presentations. In most cases the initial diagnosis is pulmonary hypertension secondary to pulmonary thromboembolism.¹ A confirmed diagnosis is usually made when studies are extended with the intention of performing thromboendarterectomy, or the diagnosis might be made during surgery or autopsy.² Although intimal sarcoma shows little ability to metastasize, the prognosis is poor. Survival is reported to be months—only occasionally years.³ We report the case of a patient who was initially diagnosed with pulmonary thromboembolism but whose condition worsened despite initial anticoagulation treatment.

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Case Description

The patient was a 41-year-old woman who reported no substance addictions but who had a history of thrombophlebitis 15 years earlier (coinciding with the gestation of her second child) that led to secondary postphlebotic syndrome requiring surgical intervention for varicose veins a year before the onset of the symptoms involved in the present case. The symptoms were progressive dyspnea and chest tightness of 2 months' duration. At physical examination the patient's breathing was normal and blood pressure was 120/80 mm Hg; slight jugular vein engorgement was detected and breath sounds were normal. Auscultation at the lower left sternal border detected a grade III to IV systolic murmur indicative of tricuspid regurgitation; a harsh, grade V to VI systolic murmur in the area of the lung that radiated to the right hemithorax was also detected; there was tactile fremitus with no diastolic component. The patient's abdomen was soft, with no evidence of visceromegaly; her lower extremities showed signs of chronic venous insufficiency but no digital clubbing. Chest radiographs and results of a D-dimer test were normal. The erythrocyte sedimentation rate was 10 mm/h and electrocardiography showed sinus rhythm and diffuse repolarization abnormalities on the lower and

anterior walls. Transthoracic echocardiography revealed right ventricle dilatation with tricuspid regurgitation. This finding enabled us to estimate pulmonary artery systolic pressure (PASP) at 90 mm Hg which, in light of the patient's medical history, led us to suspect pulmonary thromboembolism. Spiral computed tomography (CT) revealed a mass at the bifurcation of the pulmonary artery trunk suggestive of a thrombus occupying the proximal segment of the right pulmonary artery. There was no evidence of other thrombi in the rest of the respiratory tree (Figure). Perfusion scintigraphy of the lungs was also performed and showed a homogeneous decrease in perfusion throughout the right lung with no segmental defects; the results of a thrombophilia study were negative and venography revealed no filling defects of either the superficial or deep venous systems.

Anticoagulant treatment was started. However, since the patient's condition showed no improvement, repeat echocardiography was performed and disclosed an increase in pulmonary hypertension (estimated PASP, 105 mm Hg). The previously described intravascular lesion was seen to have increased in size on a repeat thoracic CT scan. Cardiac catheterization showed normal pressures and function for the left ventricle, marked dilatation of the pulmonary artery trunk, a filling defect that extended from the bifurcation into both pulmonary artery branches—especially the right branch, severe pulmonary hypertension, and pulmonary vascular resistance of $409.75 \text{ dyne}\cdot\text{s}\cdot\text{cm}^{-5}$. Both sides of the arterial tree distal to the stenosis were normal.

Given the patient's poor response, she was transferred to the referral hospital for assessment for surgical treatment (thromboendarterectomy). During the preoperative evaluation, transesophageal echocardiography revealed a mass in the lumen of the pulmonary artery, attached to the posterior wall of the main trunk at the bifurcation and extending into both branches—especially the right one, where it reached the upper lobe bifurcation. The mass measured 4 centimeters in diameter at the widest point, was rounded and slightly irregular in contour, and showed no clear signs of infiltration into the vascular and perivascular walls.

The possibility of an intravascular neoplasm was considered in light of the following evidence: continued growth of the intravascular lesion—as shown in the CT scan—regardless of anticoagulation treatment, the clearly demarcated location of the lesion in a zone with no apparent involvement of the rest of the pulmonary vascular tree, negative results of a D-dimer test, the absence of multiple pulmonary perfusion defects as shown in the perfusion scintigrams, and the appearance of the mass as rendered in the transesophageal echocardiogram. Accordingly, surgical intervention was indicated. A pulmonary arteriotomy was performed, the mass was excised at the base and the artery was reconstructed using a pericardial patch. The pathologist diagnosed the mass as intimal sarcoma of the pulmonary artery.

Following surgery the patient remained anticoagulate. She received chemotherapy with ifosfamide and adriamycin, and at 24 months, her evolution was satisfactory, with CT scans showing no evidence of recurrence and follow-up echocardiography demonstrating PASP values at 30 mm Hg.

Discussion

Pulmonary artery sarcoma can be classified as either intraluminal or intramural. The more common type is intraluminal, known as intimal sarcoma, and is usually

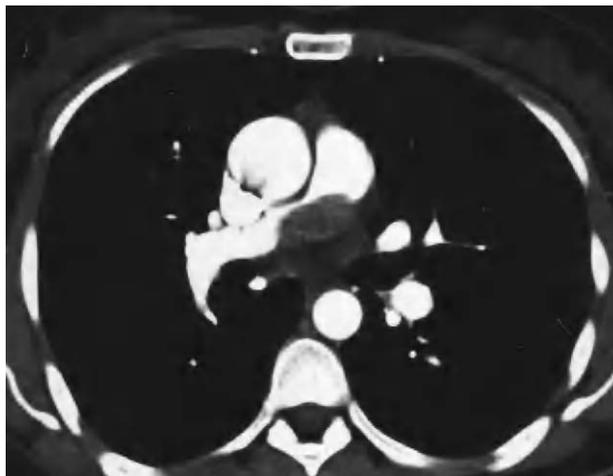


Figure. A computed tomography scan of the chest that shows a lesion at the bifurcation of the pulmonary artery.

formed of undifferentiated spindle-cells; intramural sarcomas are usually leiomyosarcomas.⁴ In general, intimal sarcoma of the pulmonary artery originates in the intimal layer and proliferates as polypoidal masses in the lumen of the vessel. In some cases, intimal sarcoma has been observed to grow proximally, affecting the pulmonary valve and the right ventricle.⁵ In approximately 50% of cases such tumors extend directly through the vessel wall to the adjacent lung, bronchial wall, lymph glands, or myocardium; distant metastasis occurs in 16% to 25% of cases.⁴

Intimal sarcoma has been reported to appear in an age range of 13 to 86 years, with the majority of cases occurring when the patient is 45 to 55 years of age.³ Isolated cases of thrombophilic conditions associated with intimal sarcoma have been reported.⁶ Faced with the lack of a specific diagnostic method for intimal sarcoma, medical personnel must exercise heightened awareness in order to make a precise early diagnosis. The entity presents in various ways. The most common initial symptoms are chest pain, hemoptysis, and dyspnea, although signs indicative of neoplasia, such as weight loss, clubbing, anemia, and an elevation in the erythrocyte sedimentation rate may also be apparent. A systolic murmur of pulmonary origin as well as signs of dysfunction of the right ventricle are usually observed in physical exploration.⁷

A chest radiograph showing hilar enlargement and an arteriogram showing intraluminal defects may indicate pulmonary thromboembolism, a mediastinal mass, bronchogenic carcinoma, or arthritis of the pulmonary artery. A spiral CT scan of the chest may reveal filling defects in the pulmonary artery that are identical to those observed in acute and chronic pulmonary thromboembolism. High resolution CT has occasionally shown a mosaic pattern of lung attenuation that is also indistinguishable from pulmonary hypertension caused by chronic pulmonary thromboembolism, possibly due to the growth of the tumor into the small vessels or

tumor embolism of the small peripheral vessels.⁵ Echocardiograms may show right ventricle dilatation and outflow tract obstruction, but distinguishing between a thrombus and a tumor is difficult if the obstruction is in the pulmonary trunk. Use of transesophageal echocardiography in the diagnosis of such tumors is yet to be well defined.⁷ Gadolinium enhanced magnetic resonance is reported to be a sensitive technique for differentiating thrombi from tumors since this imaging modality detects the depth of the intraluminal filling defect.⁸ Recently fluorine-18-2-fluoro-2-deoxy-D-glucose-positron emission tomography has been used to differentiate sarcoma from thromboembolism.⁹ Other diagnostic tools are transvenous catheter suction biopsy and pulmonary angiography, although in Spain such techniques are not generally used.

In the present case the diagnostic difficulty was compounded by the patient's history of intervention for varicose veins and the absence of tumor symptoms, such as clubbing, hemoptysis, general malaise, or an elevated erythrocyte sedimentation rate. Although preoperative imaging tests led us to suspect the possibility of a neoplasm, we were aware that most reported cases (60%) were diagnosed by angiography, or during surgery intended for thromboendarterectomy, or at autopsy.²

Surgery is the most effective treatment of intimal sarcoma and is successful only if total resection is performed.¹⁰ The mortality of patients with intimal sarcoma depends on the location and vascular extension of the tumor. The prognosis is poor, with short survival time reported: less than 2 months for patients receiving no surgical treatment and approximately 10 months for those treated surgically—albeit in some cases survival of longer than 10 years has been reported.² The roles of radiation therapy and chemotherapy in the management of intimal sarcoma and the duration of postoperative anticoagulation therapy have not yet been clearly defined. Post-operative distal arteriopathy and areas of perfusion deficit in the lung have been reported.³

In conclusion, intimal sarcoma of the pulmonary artery is a rare entity that can present in a variety of ways. It should be suspected in the investigation of chronic dyspnea, especially in cases with an initial diagnosis of pulmonary hypertension secondary to pulmonary thromboembolism and localized obstruction of the pulmonary vascular tree. Suspicion should be heightened if the initial lesion increases in size and/or there is evidence of neoplasm.

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