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A new onset pulmonary artery stenosis in a young man

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Background: Poorly differentiated and undifferentiated sarcomas are the most common primary tumors of the pulmonary arteries. They usually affect large caliber vessels and present with predominantly intraluminal growth. Dyspnea, cough, chest pain, and hemoptysis are the most common presenting symptoms; patients may have signs of chronic pulmonary hypertension. Clinical and imaging manifestations can mimic pulmonary embolism.

The overall prognosis is poor: combined therapy with surgical resection and chemo-radiotherapy offers the best survival rates.

Case presentation: A 31-year-old male was referred to our department because of recent onset dyspnea, a pre-syncopal episode and a new heart murmur.

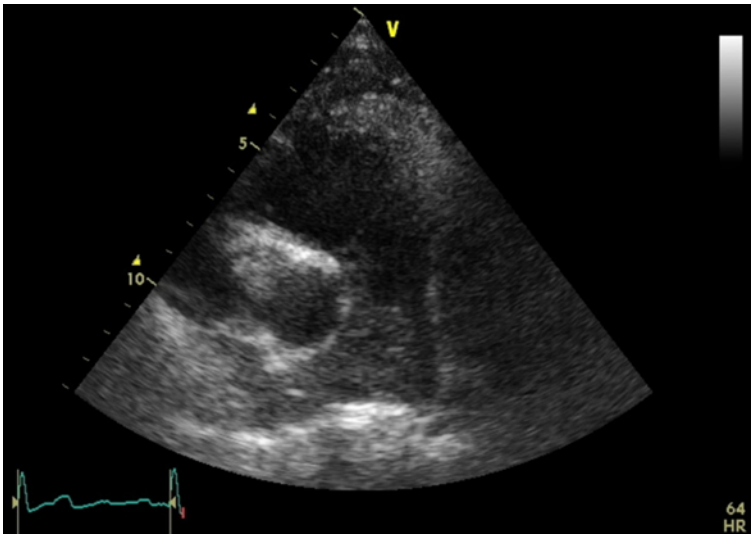
He had normal spirometry results but a significant desaturation during the 6MWT. A thoracic CT scan showed an incremented diameter of the pulmonary artery.The echocardiogram showed the presence of a mass with irregular borders attached to the pulmonary trunk almost obliterating its lumen. The mass determined a flow acceleration with maximal velocity of 3.8 m/sec, and a peak gradient of 60mmHg; Doppler findings on the pulmonary valve and right heart function were within normal values. Compression ultrasonography ruled out the presence of deep vein thrombosis.

The lesion showed a dishomogeneous impregnation in the contrastographic phase at cardiac MRI and had an intense glucidic metabolism at a PET-CT scan. These findings were highly suggestive of an angiosarcoma of the pulmonary artery. Biopsy specimens were taken through bronchoscopy.

The patient then decided to continue treatment in another hospital, where the histologic samples were sent; the cytologic results showed atypical cellular elements. The patient died a few months later.

Discussion: The presence of a unique mass in the main pulmonary artery or proximal branches and rapidly progressive dyspnea in a patient at low risk of pulmonary embolism should raise the suspicion of primary sarcoma of the pulmonary artery, which is a rare but aggressive tumor with a very poor prognosis.

Abstract P233 Figure.



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