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Typical Dyspnea in a Patient with an Atypical Diagnosis

Disnea típica en un paciente con diagnóstico atípico

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ABSTRACT

Primary pulmonary artery sarcoma is a rare tumor with a very poor prognosis. Its incidence is hard to estimate. It presents with nonspecific symptoms, delaying the diagnosis and making it challenging. We present a 62-year-old patient initially diagnosed with pulmonary thromboembolism, exhibiting discordant symptoms. Further investigation was pursued, ultimately confirming the diagnosis of primary pulmonary artery sarcoma through biopsy.

Key words: Dyspnea; Pulmonary sarcoma; Positron emission tomography

RESUMEN

El sarcoma primario de arteria pulmonar es un tumor poco frecuente y con muy mal pronóstico. Su incidencia es difícil de estimar. Presenta una sintomatología inespecífica dificultando y retrasando su diagnóstico. Presentamos un paciente de 62 años diagnosticado inicialmente de tromboembolismo pulmonar con una clínica discordante. Se decide continuar el estudio confirmándose posteriormente el diagnóstico de sarcoma primario de arteria pulmonar mediante biopsia.

Palabras claves: Disnea; Sarcoma pulmonar; Tomografía con emisión de positrones

A 62-year-old male with no significant medical history presented to the Emergency Department with NYHA (New York Heart Association) class II/IV dyspnea. He was diagnosed with pulmonary thromboembolism (PTE) at the level of the right pulmonary artery (Figure 1) via computed tomography (CT). During his hospital admission, the doppler ultrasound of the lower extremities showed no signs of venous thrombosis. Due to his good clinical progression, he was discharged with anticoagulation therapy. After 3 months, the dyspnea persisted despite treatment adherence. A CT scan was performed, which showed worsening of the condition compared to the previous study (Figure 2 a-c). Further evaluation with positron emission tomography (PET) revealed masses in both pulmonary arteries with a SUVmax >6 (maximum standardized uptake value) compatible with vascular neoplasm, along with distant lesions (Figure 2d). A biopsy of one of the masses was performed, and the results indicated pulmonary artery sarcoma. A multidisciplinary committee decided on oncological treatment, but the patient passed away six months after the initiation of therapy.

Primary pulmonary artery sarcoma is a rare tumor with very poor prognosis, more common in women in the sixth decade of life[1]. The symptoms are nonspecific and include dyspnea, fever, chest

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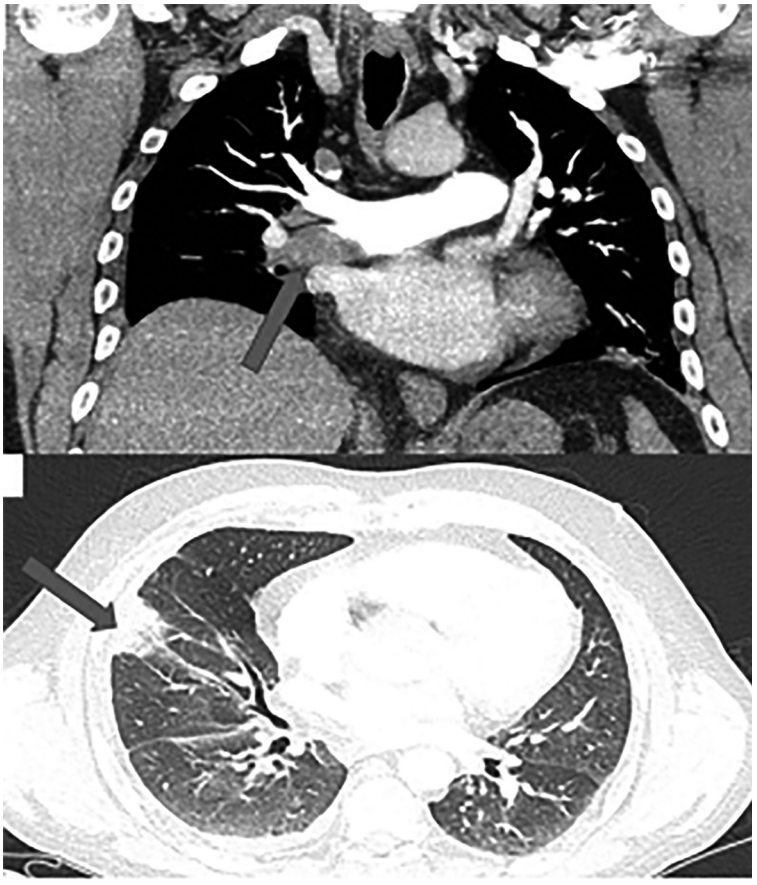

Figure 1. Upper: Coronal section of chest CT-angiography showing a thrombus in the right pulmonary artery (arrow). Lower: Axial section of chest CT-angiography showing pulmonary infarction (arrow).

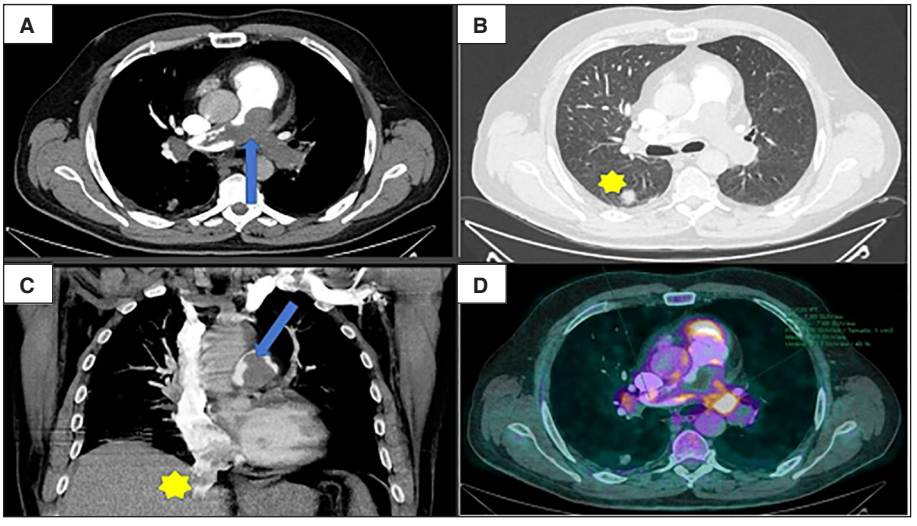

Figure 2. A and B. Axial section of chest CT-angiography showing PTE at both pulmonary arteries (arrow) and pulmonary nodule at the right lower lobe (RLL) level (asterisk). C. Coronal section of chest CT-angiography showing almost complete involvement of the left pulmonary artery (arrow) and reflux of the inferior vena cava (asterisk). D. Axial section of PTE showing increased uptake at the level of the pulmonary arteries.

pain, or syncope, which often lead to a delayed diagnosis.

The etiology is not entirely clear in the literature. Some authors[2] have associated it with the malignant degeneration of thrombi and the neoplastic transformation of mesenchymal cells. This occurs in 80 % of the cases in the trunk of the pulmonary artery and extends to the main branches in 60 % of the cases.[3]

The difficulty lies not only in the symptoms but also in imaging tests, as computed tomography detects masses in the lungs without being able to differentiate between a clot or a tumor. It's important to expand to other studies such as PET scans to guide the diagnosis. Additionally, there are some studies in the literature evaluating the use of PET scans to assess treatment response.[4]

Surgical treatment is the preferred option and can be successful if complete resection is achieved. The gold standard is endarterectomy with cardiopulmonary bypass. Other surgical techniques employed include resection with clear margins and pneumonectomy, with a median survival between 14 and 268 months in operable patients. Systemic treatment with chemotherapy and radiotherapy is given adjuvantly, with contradictory results in the literature.[5]

Systemic chemotherapy treatment can be effective in unresectable patients, such as our case, although there is limited literature on this. The few available studies demonstrate the effectiveness of various treatments such as adriamycin, doxorubicin, gemcitabine, and others.[4]

Conflict of interest
Authors have no conflicts of interest to declare.

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