CLINICAL IMAGE

From dyspnea through pulmonary embolism to angiosarcoma: a twisted diagnostic route in a young man with a history of COVID-19

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A previously healthy 31-year-old man was admitted to the hospital due to recurrent hemoptysis and exercise dyspnea, which had been progressing over a 3-month period preceding the admission. One month before the onset of the symptoms the patient had SARS-CoV-2 infection, and the manifestations were initially considered its sequelae. On admission, he presented with moderate exercise dyspnea with normal peripheral oxygen saturation (97%) and increased resting heart rate of 100 bpm. Laboratory work-up showed slightly elevated levels of C-reactive protein (7.4 mg/l, reference range [RR] <5 mg/l) and D-dimer (DD; 517 µg/l; RR <500 µg/l). The level of N-terminal pro–B-type natriuretic peptide was significantly increased (605 pg/ml, RR <125 pg/ml) but the high-sensitivity troponin T concentration was within the normal range. Echocardiography revealed significant right ventricular (RV) enlargement with elevated estimated RV systolic pressure of 68 mm Hg, a D-shaped left ventricle and a large pathologic mass filling the pulmonary trunk and floating into the RV through the pulmonary valve (FIGURE 1A and 1B). Computed tomography pulmonary angiography (CTPA) confirmed an extensive filling defect occluding almost entirely the pulmonary trunk and extending into both pulmonary arteries (FIGURE 1C). Initially, we suspected pulmonary embolism (PE) with intermediate-low early mortality risk[1] and started treatment with unfractionated heparin. No clinical improvement was observed in the following days and the DD level was still low; therefore, another CTPA was performed, which confirmed the lack of regression. Right heart catheterization could not be performed because of the risk of total obstruction of the RV outflow tract with a catheter. Due to the lack of clinical and radiological improvement despite anticoagulation, low DD levels, the lack of typical radiological signs of chronic thromboembolism, and a tight lumen of the RV outflow tract, the patient was urgently referred for pulmonary endarterectomy. The surgery involved resection of the tumor originating from the leaflet of the pulmonary valve, implantation of a biological bioprosthesis, and reconstruction of the RV outflow tract and pulmonary trunk with a patch. Immunohistochemical staining was positive for smooth muscle actin, murine double minute 2, and, focally, cyclin-dependent kinase 4. Additional histopathological findings were consistent, and a diagnosis of pulmonary artery intimal sarcoma (PAIS) was established. The resection margin was assessed as R1 and the tumor grade was classified as 2 according to the French Federation of Cancer Centers Sarcoma Group[2] (FIGURE 1D). Further hospitalization was uneventful. Positron emission tomography with ^{18}F-fluorodeoxyglucose performed 6 weeks post surgery showed no suspicious metastatic lesions.

The patient was then referred for adjuvant chemotherapy with doxorubicin (15 mg/m^2 intravenously, days 1–5) and dacarbazine (150 mg/m^2 intravenously, days 1–5) in 21-day cycles.[3] Due to the poor tolerance of chemotherapy (persistent high-grade mucositis), the treatment was discontinued after 5 of the 6 initially planned cycles. During a 12-month follow-up the patient has been in a good clinical condition and remains under strict monitoring by a multidisciplinary team.

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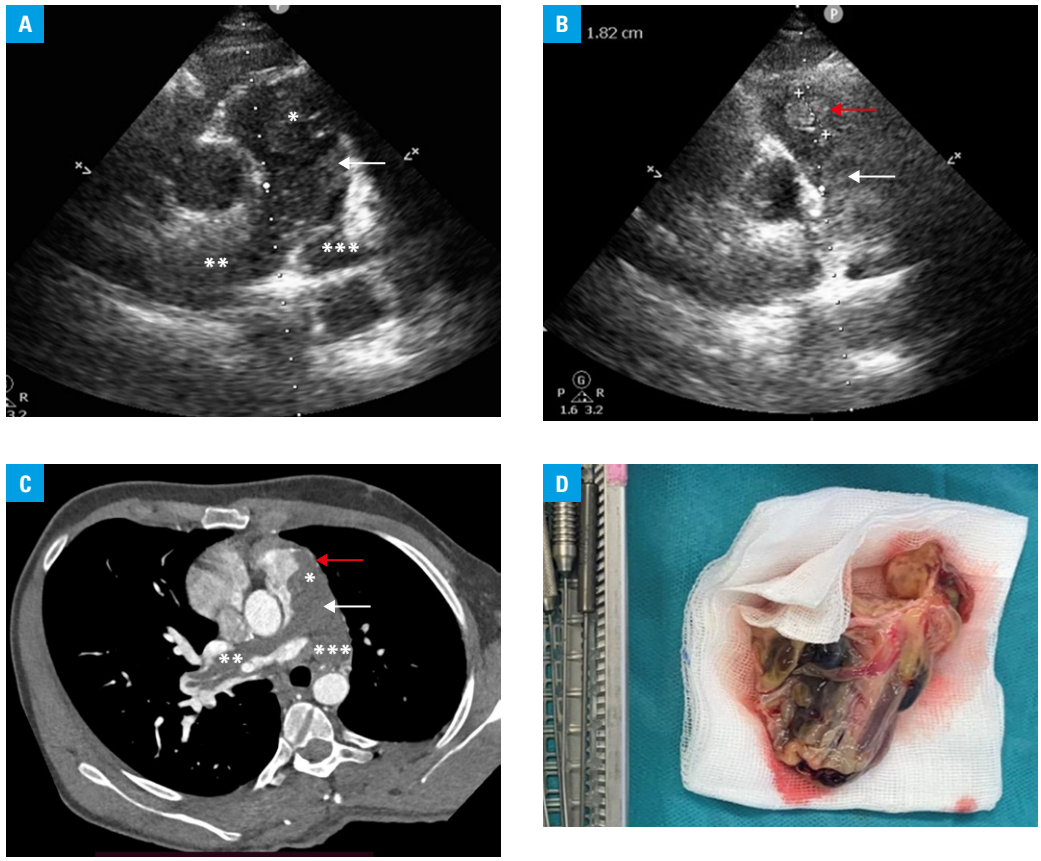

FIGURE 1 Ultrasound, radiological, and macroscopic imaging of bilateral pulmonary artery (PA) intimal sarcoma; **A** – transthoracic echocardiography, parasternal short-axis view, showing a mass extending from the pulmonary valve (single asterisk in panels **A** and **C**) filling the PA (white arrow in panels **A**–**C**), occluding nearly the whole pulmonary trunk, and extending to both PAs (double asterisk, right PA; triple asterisk, left PA; panels **A** and **C**); **B** – parasternal short-axis view focused on the pulmonary valve visualizing an additional structure of 18 mm floating to the right ventricle (red arrow in panels **A** and **B**); **C** – a computed tomography pulmonary angiography image corresponding with the aforementioned findings; **D** – a macroscopic photo of the resected mass

PAIS is a very rare and aggressive tumor with no specific treatment available. The prognosis is poor, especially in inoperable patients.[4] Even after surgical treatment the median survival is 6.6 months.[5]

To conclude, in a patient suspected of and treated for PE and not responding to anticoagulation there is a need for re-assessment with respect to nonthrombotic causes of PE, including rare malignant tumors, especially when the levels of fibrin degradation products are low.

ARTICLE INFORMATION

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