Pulmonary artery sarcoma: a rare cause of dyspnoea

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A 72-year-old woman with no significant medical history presented to the emergency room for severe dyspnoea. The initial clinical diagnosis was acute pulmonary embolism. Heparin infusion was initiated while awaiting a computed tomographic scan but the patient’s condition deteriorated dramatically and stat echocardiogram showed tamponade. Post-evacuation echo showed a dilated right ventricle with pulmonary hypertension and obstruction of the right pulmonary artery by a homogeneous mass attached to the pulmonary artery, suggesting a tumour rather than a thrombus. Computed tomographic scan confirmed the presence of an obstructive mass with almost no perfusion of the right lung. The patient was referred to cardiac surgery and the mass was removed, with anatomo-pathological diagnosis of a typical pulmonary artery sarcoma. Unfortunately, the patient died a few days after surgery. Primary pulmonary artery sarcoma is a rare tumour that arises in the central pulmonary arteries. Clinical presentation is often attributed to other causes of pulmonary hypertension, like pulmonary embolism. Magnetic resonance imaging could help to distinguish a soft tissue mass from a thrombus, but definitive diagnosis is almost always made at surgery or autopsy since patients usually present in acute and unstable situations. Prognosis is poor, from several months to a few years, and depends on how early the diagnosis is made, the presence of recurrence or metastasis after surgical resection, and the use of adjuvant therapy like radiation and chemotherapy.

Keywords
Pulmonary artery sarcoma • Echocardiography • Tamponade

A 72-year-old woman with no significant medical history presented to the emergency room for severe dyspnoea. She had noted shortness of breath with moderate exercise for 1 month and decided to come to the emergency room when her symptoms worsened acutely over 48 h. Physical examination at admission was remarkable for polypnoea with low oxygen saturation (89%), tachycardia (110 bpm), preserved haemodynamics (blood pressure of 125/68 mmHg) and jugular venous distention. Electrocardiogram was normal except for tachycardia. The initial clinical diagnosis was acute pulmonary embolism. Heparin infusion was initiated while awaiting a computed tomographic (CT) scan, but 20 min later, the patient’s condition deteriorated dramatically with a drop of her systolic blood pressure to 82 mmHg. A stat echocardiogram showed tamponade. Subxiphoid pericardiocentesis was immediately performed under echo guidance and >150 cc of haemorrhagic fluid was evacuated, with immediate haemodynamic improvement but persistent dyspnoea. Post-evacuation echo showed a dilated right ventricle with pulmonary hypertension (estimated pulmonary artery pressure 54 mmHg). Parasternal short-axis views showed obstruction of the right pulmonary artery by a homogeneous mass attached to the pulmonary artery, suggesting a tumour rather than a thrombus (Figure 1A; Supplementary data online, Videos S1 and S2). Computed tomographic scan confirmed the presence of an obstructive mass with almost no perfusion of the right lung (Figure 1B). Since the patient had persistent hypoxia with the challenges of anti-coagulation, she was referred to cardiac surgery and the mass was removed, with anatomo-pathological diagnosis of a typical pulmonary artery sarcoma. Unfortunately, the patient died a few days after surgery.

Primary pulmonary artery sarcoma is a rare tumour that arises in the central pulmonary arteries. No risk factors have been identified, although some studies have shown a slight female predominance.1,2 Clinical presentation is often attributed to other causes of pulmonary hypertension, like pulmonary embolism. Magnetic resonance imaging could help to distinguish a soft tissue mass from a thrombus,3 but definitive diagnosis is almost always made at surgery or autopsy since patients usually present in acute and unstable situations. In our case, heparin infusion complicated the
patient’s condition due to a major neoplastic bleeding in the pericardial sac. Bedside echo was a key diagnostic exam when the patient became haemodynamically unstable, although it should have been performed earlier pending CT availability, as recommended in the European Society of Cardiology guidelines. Prognosis is poor, from several months to a few years, and depends on how early the diagnosis is made, the presence of recurrence or metastasis after surgical resection, and the use of adjuvant therapy like radiation and chemotherapy.

Supplementary data
Supplementary data are available at European Journal of Echocardiography online.

Conflict of interest: none declared.

References

Figure 1 Obstruction of the right pulmonary artery by a large and homogenous mass (yellow arrow) as seen by echo (A) on a parasternal short-axis view and by computed tomographic scan (B), resulting in almost no perfusion of the right lung (green arrows).