AN UNUSUAL CASE OF ACUTE PULMONARY EMBOLISM

Image in cardiology

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A 54 year old obese female with a history of hypertension presented to the emergency room with progressive dyspnoea over a few weeks. There was no relevant past history. Clinical examination revealed an elevated jugular venous pressure, pedal oedema and tenderness in the right hypochondrium. An ejection systolic murmur was noted across the pulmonary valve. She was haemodynamically unstable with an oxygen saturation of 88% on oxygen and a D-Dimer of 7.17 ug/ml. The X-Ray showed clear lung fields with prominence of the hilar regions. The echocardiogram (ECHO) revealed a long linear mass arising in the inferior vena cava (IVC) and extending into the right ventricular outflow tract. Inotropic support and a heparin infusion were initiated. After an initial improvement she deteriorated with increasing tachycardia, tachypnoea, hypotension, metabolic acidosis and progressive renal failure (creatinine of > 200umol/L and a urea of 11 mmol/L). Computed tomography demonstrated an intraluminal filling defect as described in (Figure 1).

A median sternotomy was performed and cardio-pulmonary bypass instituted. The pulmonary arteries and right atrium were opened exposing a long snake-like contiguous tumour that did not attach to nor invade the wall of the heart or IVC. It was delivered into the wound via the tricuspid valve and by gentle traction from the IVC (Figure 2).

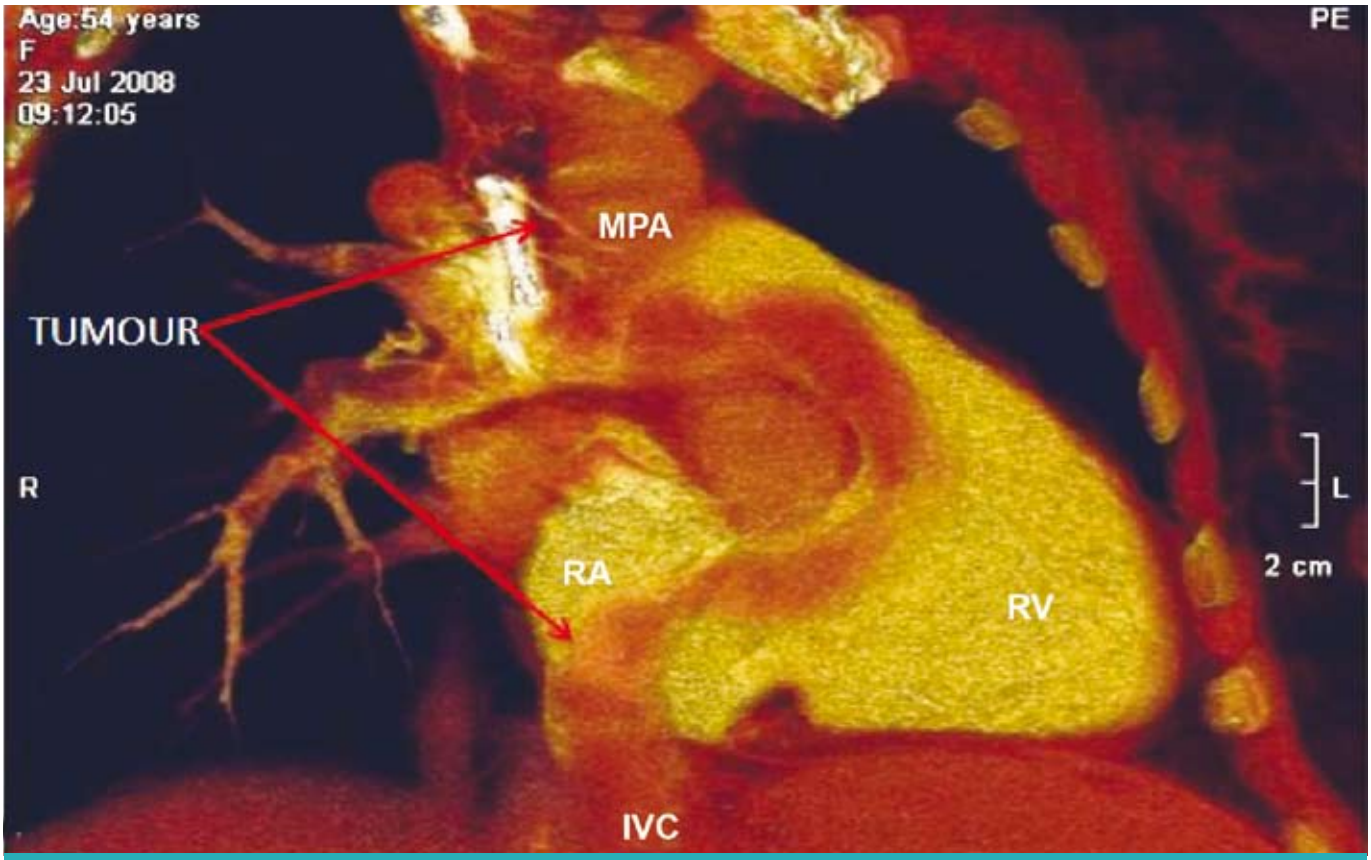

FIGURE 1: Curved multiplanar reconstruction showing filling defect extending from the IVC through the right heart chambers into the pulmonary arteries.

MPA: main pulmonary artery, RV: right ventricle, RA: right atrium, IVC: inferior vena cava.

She was transferred back to the ICU with good postoperative blood gases and a normal cardiac output. She remained stable over the next 48 hours and was extubated. Subsequently her organ function deteriorated, probably due to sepsis (leukocytosis of 25.8 $\times 10^{(9)}$L and a procalcitonin of 5,99ng/ ml (normal < 0.05). Serial blood cultures were negative. She was initiated on broad spectrum antibiotics as well as aggressive supportive therapy but died four days later.

Acute massive pulmonary embolism (PE), defined as an occlusion of the pulmonary artery exceeding 50% of its cross-sectional area is associated with a high mortality. Cardiac neoplasms of uterine origin are rare and have seldom been reported in clinical practice and may mimic acute massive PE.[1]

Dyspnoea, hypoxemia, right heart strain, and clear lung fields are common features of both tumour and thrombotic emboli and these entities are frequently clinically indistinguishable.[6,7] The treatment and prognosis differ considerably and therefore important to determine the correct diagnosis in each patient.

Pathology in this case revealed a tumour with features of an endometrial stromal cell sarcoma (ESCS) which is an aggressive tumour of the uterus with a propensity for local invasion as well as systemic metastasis rarely involving the large vessels or the heart.[1,2,3] They are the least common tumours of the uterus comprising only 0.2% of all uterine malignancies and less than 10% of all uterine sarcomas. The pathogenesis remains unknown.[9]

Based on available data complete surgical resection utilising cardiopulmonary bypass with or without hypothermic circulatory arrest is the mainstay treatment of intravenous uterine leiomyosarcomas with intracardiac extension.[1,3,4,5,8,10]

The findings of such an extensive "thrombus" seen on CT scan or ECHO should always raise the suspicion of a more sinister underlying pathological process such as a primary malignant tumour of an abdominal organ.

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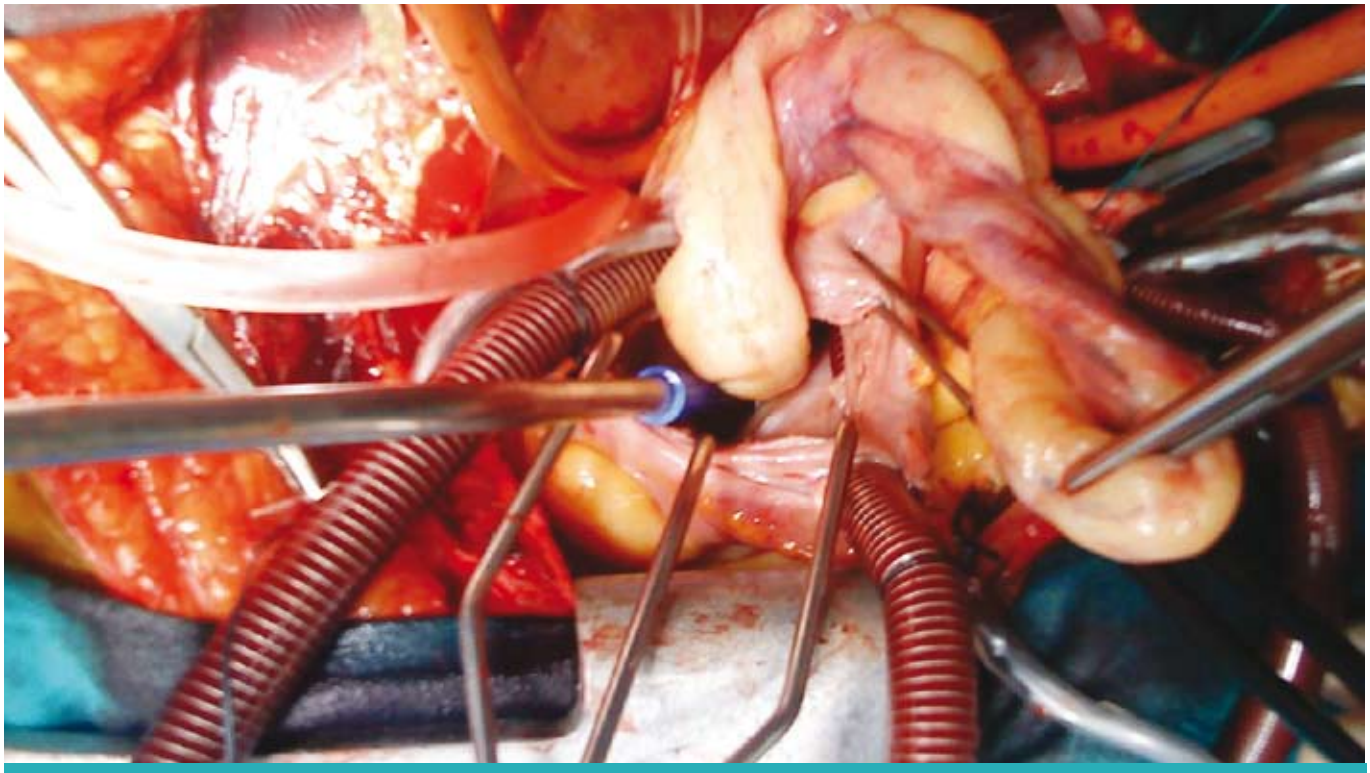

FIGURE 2: A long rubbery solid lesion withdrawn from the heart, pulmonary vessels and inferior vena.