Rare case of a multilocular primary cardiac intimal sarcoma presenting as left atrial mass with new onset atrial fibrillation

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A 70-year-old woman presented with atrial fibrillation and a new systolic murmur. Transthoracic echocardiography (see Supplementary material online, Video S1) and magnetic resonance imaging (see Supplementary material online, Video S2) revealed a 4 × 6 cm mass in the left atrium reaching to the mitral valve (Panel A) resulting in elevated pressure gradients and mild–moderate mitral regurgitation (Panel A). Transoesophageal echocardiography depicted a mass infiltrating the left atrial posterolateral wall and auricle with contact to the posterior mitral leaflet (see Supplementary material online, Videos S3 and S4). During surgery, a tumour of 1.5 cm was identified, arising from the confluence of the right upper pulmonary vein. A second tumour of 7 cm connected to the left auricle and three additional smaller tumours (≤1 cm), at the posterior mitral annulus the entrance of the left lower pulmonary vein and the lateral atrial wall were identified. Histopathology revealed an intimal sarcoma Grade 3 with a growth fraction of 15% with R1/R2 resection (see Supplementary material online, Figure S1). Computed tomography (CT)-staging was negative for metastases (Panel B) and due to lack of standardized chemotherapy, the patient was discharged with plan for regular follow-up imaging but presented 6 weeks later with a right humerus fracture due to a metastasis of the intimal sarcoma. Repeat-CT revealed metastatic disease to liver and the eight thoracic vertebra (Panel B). Unfortunately, radiation to the pulmonary artery main stem and chemotherapy did not limit disease progression and the patient passed away. Intimal sarcomas are extremely rare and typically originate outside the heart. To our knowledge, this is the first report of a multilocular primary cardiac intimal sarcoma originating in the left atrium.

Panel A. Large left atrial sarcoma with regular and smooth surface, slightly reduced echogenicity, and size measuring 4 × 6 cm recorded by transoesophageal echocardiography (A). During diastole, the tumour prolapses into the left ventricle, partially obstructing the mitral valve orifice resulting in elevated pressure gradients (Pmax = 25 mmHg, Pmean = 9 mmHg) (B) and turbulent diastolic inflow over the valve with a peak velocity of ~2.5 m/s. (C) Three-dimensional reconstruction of cardio-CT. Left atrial mass originating at the posterior wall indicated by arrow. (D) MRI steady-state free precession-sequence in four chamber view depicting a hypo-intense tumorous mass originating from the left posterolateral atrium (white arrow). A second hypo-intense tumour is depicted at the confluence of the right upper pulmonary vein by red arrow.

Panel B. Top row: CT-scans showing progression of hepatic metastases. No metastases were found in the first CT scan while follow-up scans at 5 and 7 months revealed increasing size and dissemination of hepatic metastases. Bottom row: CT-image showing relapse of sarcoma including pulmonary veins. Follow-up CT-scans depicting progression of metastasis involving the 8th thoracic vertebral body.

Supplementary material is available at European Heart Journal online.