A 38-year-old, previously healthy man, presented at our institution after a syncope during exertion. He referred worsening exertional dyspnoea over last year. On physical examinations, a grade II systolic murmur was detected. Electrocardiogram and chest radiography did not show abnormalities. Transoesophageal echocardiography revealed a $3 \times 1$ cm mobile mass that seemed to be attached to the pulmonary valve (PV) and another big mass inside the pulmonary artery (Figures 1A and B and 2A–2C; Supplementary material online, Videos 1 and 2). A pressure gradient of $50$ mmHg between the right ventricle and the pulmonary artery was detected (Figure 1C; Supplementary material online, Videos 3 and 4).

Cardiac magnetic resonance confirmed an intraluminal filling defect in the main pulmonary artery (MPA) extending to left and right pulmonary arteries (Figure 1D). Patient was scheduled for surgery. Through pulmonary arteriotomy, an irregular bluish-red $7 \times 4$ cm mass (†) arose from the lumen of the MPA (Figure 1E). Two more yellowish masses (‡/‡‡) were firmly attached to the ventricular side of PV leaflets, prolapsing into the right ventricle (Figures 1F and 3A–C). Complete resection was performed. The pulmonary outflow tract was reconstructed using a pulmonary homograft. Histopathological examination confirmed a high-grade pleomorphic intimal sarcoma (FNCLCC grade 3), dedifferentiated chondrosarcoma like with endocardial multifocal polypoid growth. Same histological diagnosis was found in all tumour masses. Immunohistochemical features demonstrated neoplastic cells positive for vimentin and $\SI{100}{\text{protein}}$. An extensive panel of immunohistochemical stains revealed focal areas with rhabdomyoblastic differentiation and positivity to desmin and actin. Focal immune-reactivity showed epithelial membrane antigen positivity. Staining with antibody to CD $31$/CD $99$/CD $57$ was negative.

Primary intimal sarcomas are weird malignant tumours arising in the mesenchymal tissue of great arteries. Intimal sarcomas are exceptional and commonly misdiagnosed. Echocardiography, gadolinium-enhanced magnetic resonance, and computed axial tomography scan allow narrowing a differential diagnosis. The prognosis of primary pulmonary artery sarcomas is poor due to its extreme aggressiveness and delayed diagnosis. Surgical removal remains the primary approach. AV, aortic valve. RA, right atrium.

Supplementary material is available at European Heart Journal online.