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A rare case of left atrial undifferentiated pleomorphic sarcoma

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An 87-year-old male presented to hospital with progressive dyspnoea limiting exercise tolerance to 5 m and intermittent chest pain over the last 6 months. His medical history was significant for a previous aortic dissection repair 35 years ago, hypertension, paroxysmal atrial fibrillation, and prostate cancer treated with curative radiotherapy. He was functionally independent.

His electrocardiogram demonstrated rate-controlled atrial fibrillation. He had a normal full blood examination, mild renal impairment, and normal inflammatory markers. A trans-thoracic echocardiogram showed a large, mobile cystic mass on the lateral wall of the left atrium prolapsing into the left ventricle causing functional mitral stenosis and moderate mitral regurgitation with severe pulmonary hypertension (*Figure 1A* and Supplementary material online, *Video*). On cardiac magnetic resonance imaging, the mass showed no enhancement on delayed, post-contrast sequence (*Figure 1B*). Computed tomography of the chest, abdomen, and pelvis showed no metastatic disease elsewhere. After discussion with the patient and at multidisciplinary meeting, he underwent resection of his left atrial cystic mass via midline sternotomy (*Figure 1C*) which immediately lowered his pulmonary pressures. Histopathology showed large pleomorphic epithelioid cells with eosinophilic cytoplasm set in a fibromyxoid stroma with some blood vessels. There were marked irregular hyperchromatic nuclei and some multinucleated cells. Occasional pseudo-nuclear inclusions were also seen. Immunohistochemistry showed the cells to be negative for markers CD31, CD68, smooth SMA, AE1/AE3, S100, Melan-A, SOX10, CD20, ERG, MPO, CD2, and CD30. In conclusion, the markedly atypical cells demonstrated features of undifferentiated pleomorphic sarcoma (*Figure 1D*). He was discharged home on post-operative Day 11 with planned oncology outpatient review. Six weeks after surgery, his dyspnoea had markedly improved, and he remains functionally independent. To our knowledge, this is the first reported case demonstrating a cystic appearance of an atrial mass shown to be malignant sarcoma.

Supplementary material

Supplementary material is available at. *European Heart Journal – Case Reports* online.

Consent: The patient verbally consented to the publication of his medical case in a peer-reviewed medical journal. The authors of this article, who actively participated in the decision process and management, obtained written informed consent from the patient, in accordance with COPE guidelines.

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Data availability

De-identified data are available upon request.

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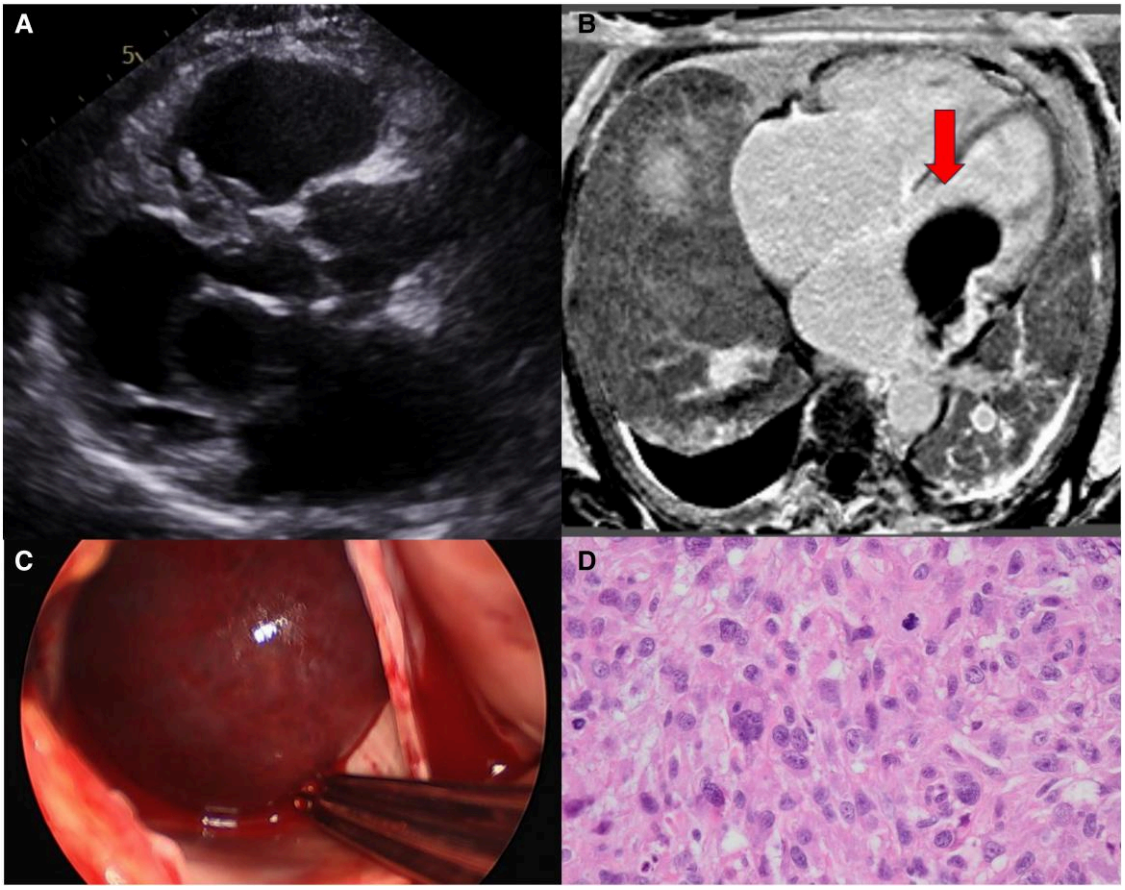

Figure 1 (*A*) Transthoracic echocardiogram in parasternal long axis view demonstrating a large mobile cystic mass prolapsing from the left atrium into the left ventricle. (*B*) Cardiac magnetic resonance imaging confirming the large cystic mass. (*C*) Surgical view of the large left atrial mass after incision through the inter-atrial septum. (*D*) Histopathology showing markedly atypical, undifferentiated, and pleomorphic cells.