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IMAGES IN CV APPLICATIONS

Atypical presentation of a primary cardiac malignant peripheral nerve sheath tumor

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A 51-year old female patient was seen at the emergency department with complaints of palpitations, dyspnea and abdominal pain. Her prior medical history was uneventful. At physical examination, the p atient was afebrile, blood pressure was 125/75 mmHg, the peripheral oxygen saturation was 96% and there was diffuse abdominal tenderness. The ECG showed a sinus tachycardia with low QRS voltages.

The chest X-ray demonstrated an enlarged heart and right-sided pleural effusion. Transthoracic and transesophageal echocardiography demonstrated the presence of a left atrial tumor, however the origin and extension of the tumor could not be visualized (Fig. 1a). A thoracic-abdominal computer tomography (CT)-scan was performed which revealed a tumor in the left atrium, with growth through the interatrial septum in the right atrium, compression of the intraventricular septum and both pericardial and pleural effusion. Cardiac magnetic resonance imaging (MRI) demonstrated the tumor to be $6.7 \times 6 \times 4.7$ cm in size with heterogeneous late-enhancement after gadolinium administration (Fig. 1b). A fluorodeoxyglucose (FDG)-positron emission tomography (PET) disclosed inhomogeneous FDG avidity in the tumor with central necrosis, but no uptake elsewhere in the body (Fig. 1c). Biopsy of the tumor was performed showing non-atypical spindle-cells in a myxoid stroma consistent with cardiac myxoma. The patient underwent cardiac surgery with the intention to radically remove the tumor. However, perioperative infiltrative growth of the tumor into the coronary sinus was seen and the process appeared malignant. Resection was not possible and only debulking of the process could be performed. Histopathology now revealed myxoid stroma with spindle-cell architecture, but with high mitotic activity and focal necrosis. Immunohistochemical stains for S100, CD56, CD57 and SOX10 (Fig. 1d, e) were focally positive, whereas Pankeratin, melan-A, Tyrosinase, E-cadherin, P16, MiTF, HMB45, BRAFV600E, SMA, desmin, CD34, ERG, MYO-D1 and MYF-4 were negative. This confirmed

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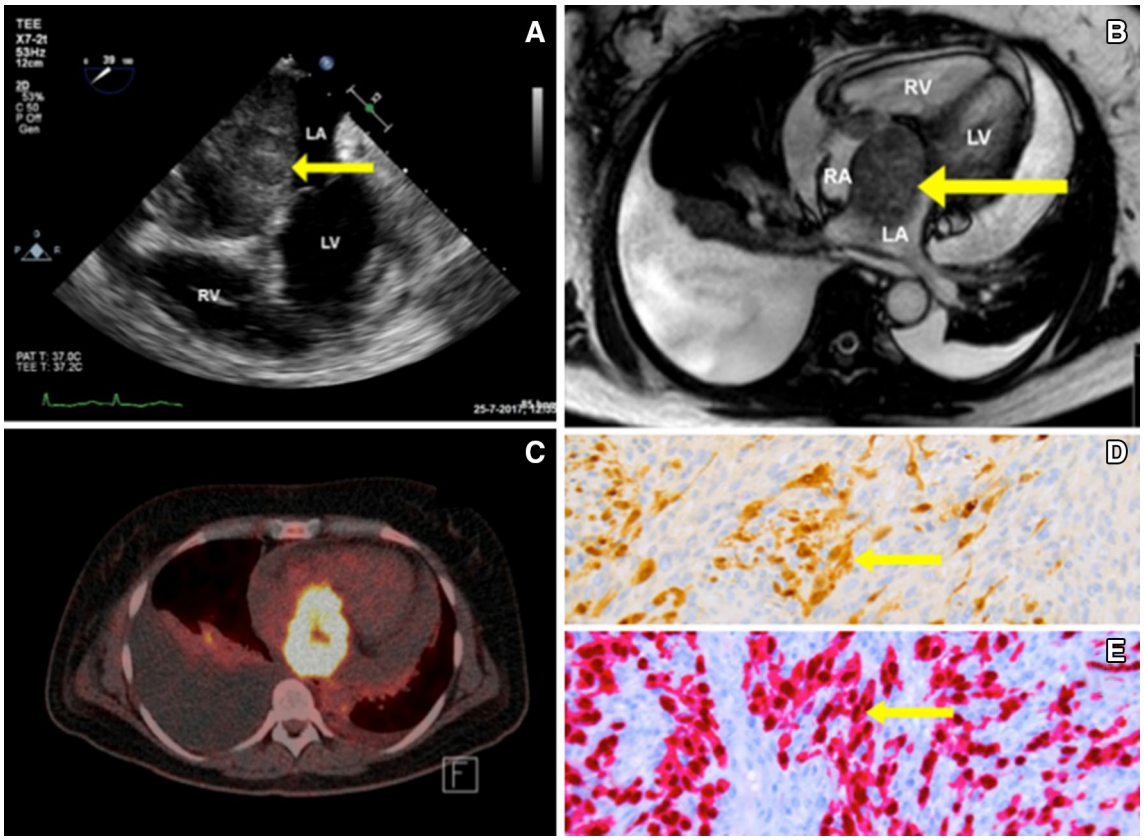

Fig. 1 Transesophageal echocardiography revealed a large tumor in the left atrium (**a**, arrow). Cardiac MRI showed the extensiveness of the tumor (**b**, arrow) with pleural and pericardial effusion. PET-CT-scan revealed inhomogeneous FDG uptake and central necrosis in the tumor (**c**). Immunohistochemical stains of spindle cells with S-100 (**d**, arrow) and SOX10 (**e**, arrow). *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

the diagnosis primary malignant peripheral nerve sheath tumor, a rare variety of soft-tissue sarcoma. No treatment options were available. The patient deteriorated quickly and died 3 months later.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.