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Two cases of atrial sarcoma: The dreadful puncture of the spindle

Short title: Two cases of atrial spindle cell sarcoma

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Patient 1: A 56-year-old woman with no previous medical history presented with progressive dyspnea, bibasilar crackles, and paroxysmal atrial fibrillation. Echocardiography revealed a large solitary mass in the left atrium, attached to the lateral free wall, with a wide base of implantation, interfering with mitral valve dynamics (Supplementary material, *Video S1*).

Patient 2: A 71-year-old male with arterial hypertension presented with tachycardia and peripheral edema. Echocardiography revealed a large multilobulated right atrial mass, partially obstructing the tricuspid valve.

Both patients underwent late gadolinium-enhanced (LGE) cardiac magnetic resonance (CMR) to obtain tissue characterization of the lesions. The two masses showed similar imaging features:

isointensity in FSE-T1 sequences (Figure 1A and D), strong hyperintensity in STIR sequences (Figure 1B and E) and inhomogeneous LGE (Figure 1C and F). In cine steady-state free precession sequences, the masses were mildly hyperintense and caused functional interference with atrio-ventricular valves (Supplementary material, *Figure S1*, *Videos S1–S2*). In case 1 the mass infiltrated the left superior pulmonary vein and the atrial appendage, as shown by cardiac CT (Supplementary material, *Video S3*). The histological examination of surgical specimens revealed spindle cell sarcoma in both cases. Both masses were successfully resected (Supplementary material, *Figure S2*). Follow-up after surgery lasted respectively 2 years and only 6 months.

Spindle cell sarcoma is the least common type among malignant cardiac tumors, with only nine cases reported in the literature so far [1]. It is a remarkably aggressive mesenchymal malignant tumor, mainly affecting the great vessels and rarely involving the hearts of patients between 20 and 50 years of age, with a reported mean survival ranging between 3 months and 1 year. Aspecific symptoms often cause diagnostic delay, although second-level imaging, including computed tomography and CMR, has significantly improved the diagnostic work-up. The differential diagnosis of myocardial lesions is often challenging. CMR is the gold-standard technique to perform tissue characterization and evaluate the size and location of myocardial masses. Moreover, the interobserver agreement rate at CMR for the differential diagnosis between benign and malignant tumors is high [2, 3]. While both benign and malignant masses are not distinguishable at T1 imaging, as they both commonly show isointense signal, on T2-weighted sequences, benign masses are more often hyperintense while malignant tumors display both hyperintensity and isointensity; also, first-pass perfusion is more commonly observed in malignant lesions. The most challenging differential diagnosis for spindle cell sarcoma is atrial myxoma, since they both typically involve the left atrium, showing similar intensity features at T1, T2, and LGE images. Spindle cell sarcomas, however, usually have a non-septal origin, a broad attachment to the atrial wall, and high vascularization features. The differential diagnosis with cardiac thrombus is mainly driven by the low-intensity signal of the clotted blood on contrast-enhanced CMR. As much as CMR-derived tissue characterization may help in suspecting the diagnosis, the final confirmation requires surgical exeresis and histological examination.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish_heart_journal.

Article information

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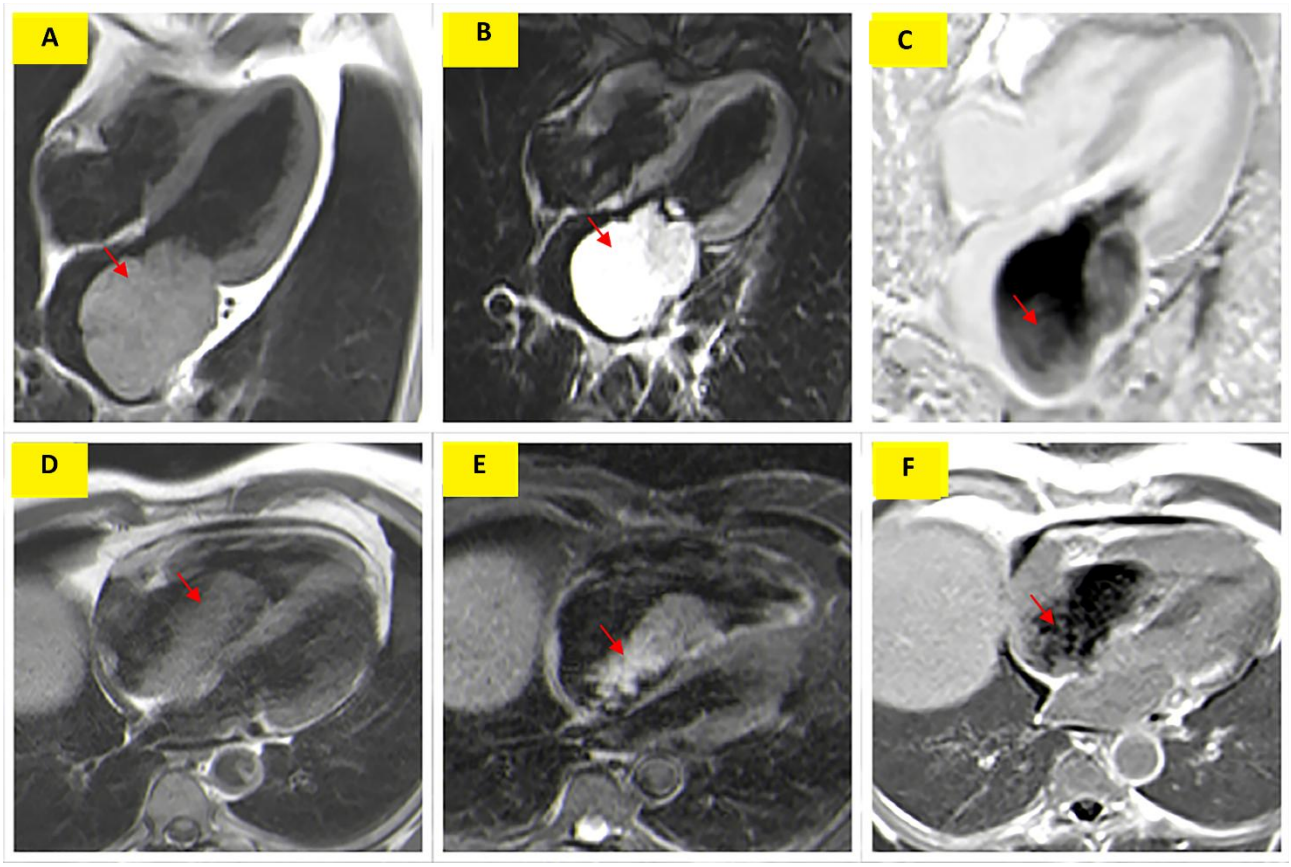

Figure 1. Cardiovascular magnetic resonance of the two atrial masses showing isointensity in FSE-T1 sequences (**A** and **D**), strong hyperintensity in STIR sequences (**B** and **E**) and inhomogeneous LGE (**C** and **F**)