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CARDIOVASCULAR FLASHLIGHT

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Cardiac magnetic resonance characterization of atrial pseudo-mass in Erdheim-Chester disease

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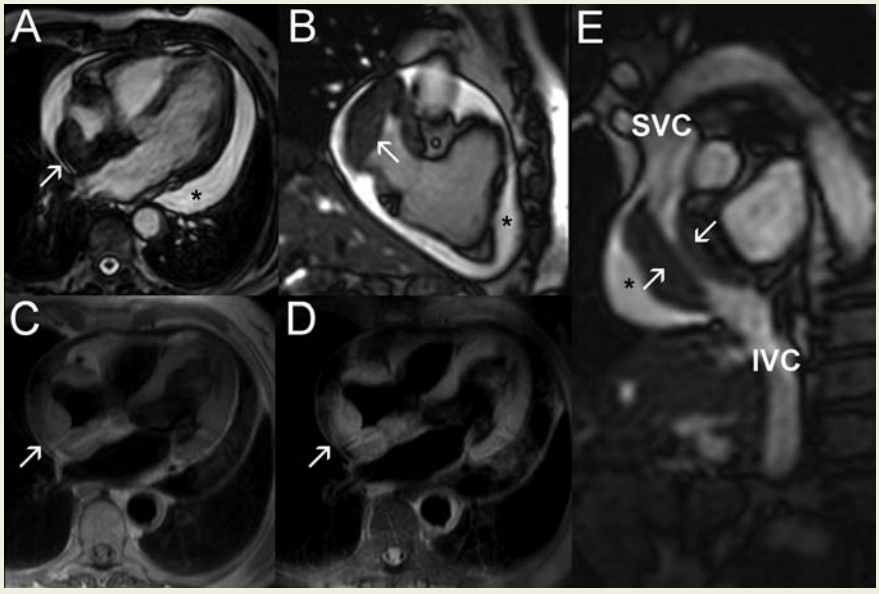

A 70-year-old man with end-stage renal failure due to Erdheim-Chester disease was admitted for dyspnoea. Cardiac auscultation revealed diminished heart tones. Transthoracic echocardiography showed an irregular thickening of the right atrium and a circumferential moderate-to-severe pericardial effusion. Thus, the patient was referred for further evaluation by cardiac magnetic resonance (CMR). Steady-state free-precession (SSFP) cine CMR images confirmed a pericardial effusion (asterisk on *Panels A*, *B*, and *E*) and showed a massive thickening (pseudo-mass) of the right atrium wall, which appeared hysointense to the normal myocardium (white arrows on *Panels A* and *B*); both on T1-weighted fast spin-echo (FSE) axial image without (*Panel D*) and with fat suppression (*Panel E*); the atrial pseudo-mass was hysointense (white arrows) to normal myocardium. As showed on parasagittal section by cine CMR (*Panel E*), the pseudo-mass causes obstruction (white arrows) of right atrial inflow through superior and inferior caval vein (SVC and IVC, respectively). Erdheim-Chester disease is a rare histiocytic disorder of unknown cause which frequently involves the appendicular skeleton and retroperitoneum. The diagnosis is based on the immunochemistry analysis (CD68+, CD1a−).

Cardiac pseudo-masses in Erdheim-Chester disease have largely been described previously. However, to our knowledge, this is the first case of severe limitation to the right atrial inflow due to the atrial pseudo-mass. We believe that CMR is a precious tool to diagnose cardiac involvement in Erdheim-Chester disease and quantifying the haemodynamics consequences.