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 hypointense 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 hypointense (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.

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