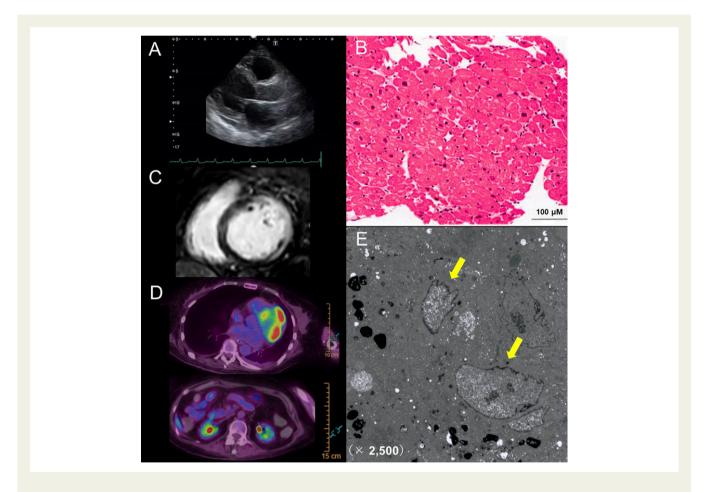
Fulminant cardiac and renal sarcoidosis revealed by electron microscope: challenging aspect of diagnosis

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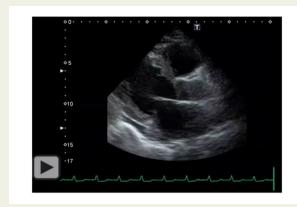


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Video I Echocardiography in parasternal long-axis view.

A 62-year-old woman with no prior history of cardiovascular and renal disease was referred to our department due to progressive cardiorenal dysfunction with fatigue and dyspnoea over the past 3 weeks. Chest X-ray revealed lung congestion and electrocardiogram showed intraventricular conduction disturbance (Supplementary material online, Figure S1). The laboratory tests revealed an increased level of serum N-terminal-pro-B-type natriuretic peptide (72 862 pg/mL) (reference value < 55 pg/mL), troponin T (466 ng/L) (reference value < 16 ng/L), creatinine (5.53 mg/dL) (reference value 0.65-1.07 mg/dL), angiotensin-converting enzyme (ACE, 26 U/L) (reference value 8.3-21.4 U/L), and soluble interleukin-2 receptor (s-IL2R, 6151 U/mL) (reference value < 613 U/mL). Echocardiography showed severely reduced left ventricular ejection fraction (LVEF) (25%) without basal thinning of interventricular septum, which is characteristic feature of cardiac sarcoidosis, or wall thickening as seen in acute infective myocarditis (Panel A, Videos 1 and 2). Coronary angiogram excluded coronary artery disease, and light-microscopy of endomyocardial biopsy specimen from the right ventricular septum did not show any findings of sarcoidosis or myocarditis (Panel B). The haemodynamic condition progressively worsened, resulting in the implantation of a intra-aortic balloon pump and extracorporeal membrane oxygenation. On the basis of elevated ACE and s-IL2R levels, we suspected the existence of fulminant cardiac and renal sarcoidosis and decided to administrate pulse methylprednisolone therapy (1000 mg/day for 3 days) followed by a maintenance dose of prednisone (started from 40 mg/day and gradually tapered to 5 mg/day). Her haemodynamics dramatically improved thereafter, and she was successfully removed from mech-



Video 2 Echocardiography in 4-chamber view.

anical circulation support in 3 days. Finally, renal function was normalized and LVEF remarkably recovered (58%) 60 days and 100 days after the administration of steroid therapy, respectively. We continued cardioprotective drugs and prednisone paying attention to the clinical course. Cardiac magnetic resonance imaging (CMR), which was performed after the recovery of renal function in the late phase of the myocarditis, showed non-patchy transmural or in mesocardium late gadolinium enhancement in the anterolateral LV wall (Panel C); ¹⁸F-fluorodeoxyglucose positron emission tomography (FDG-PET) revealed abnormal uptake in both LV myocardium and kidney (Panel D). Finally, electron microscopic images of the myocardium which was procured from the right ventricular septum on admission revealed non-caseating epithelioid granuloma (Panel E, yellow arrow), indicating cardiac sarcoidosis. In this case, not only light-microscopy but also CMR was not conclusive for cardiac sarcoidosis diagnosis. This emphasizes the choice of electron microscopy or FDG-PET for sarcoidosis diagnosis.

Supplementary material

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

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.