Steroid-induced metamorphosis of the heart: a case of cardiac sarcoidosis

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A 35-year-old man was admitted for acute heart failure. Electrocardiography showed epsilon waves in leads V1 and V2 (Panel A). Echocardiography showed asymmetric septal hypertrophy mimicking hypertrophic cardiomyopathy (Panel B and see Supplementary data online, Video S1). Enlargement of the mediastinal lymph nodes (Panel C) and multiple low-density nodules in both the liver and spleen were found on enhanced computed tomography (Panel D). Positron emission tomography revealed abnormal patchy areas of 18F-fluorodeoxyglucose uptake in the interventricular septum (IVS) and right ventricular (RV) wall (Panel E). A biopsy demonstrated non-caseating granulomas in the endocardial endothelium (Panel F). We diagnosed multivisceral sarcoidosis, including both ventricles. Six days after initiating steroid treatment, echocardiography surprisingly revealed a striking change in heart morphology characterized by a marked reduction in the IVS wall thickness from 17.0 to 7.4 mm, coupled with left ventricular dilatation, and contractile dysfunction, simulating dilated cardiomyopathy. Additionally, the steroids induced a large mobile thrombus adherent to the IVS (Panel G, arrows), although subsequent anticoagulation therapy resulted in disappearance of the thrombus (Panel H and see Supplementary data online, Video S2). Despite the striking change of the heart, the epsilon waves remained, implying that the steroid had no effect on the constituent elements responsible for the epsilon waves, i.e. islands of viable RV myocytes surrounded by scar tissue. Although steroid therapy can arrest the progressive myocardial damage in cardiac sarcoidosis, it only rarely induces a striking change in heart morphology due to the resolution of inflammatory oedema, which appeared like a metamorphosis of the heart.

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

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