Non-tropical endomyocardial fibrosis associated with sarcoidosis

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A 50-year-old female was admitted with a 2-year history of significant weight loss and a pyrexia of unknown origin. Hepatosplenomegaly and mildly enlarged retroperitoneal and mediastinal lymph nodes were seen on a computed tomography scan. The patient was profoundly cachectic with severe muscle wasting. Following an episode of acute pulmonary oedema, a transthoracic echocardiogram (TTE) was performed, revealing extensive thickening of the lateral and inferior left ventricular (LV) walls, extending from the mid-segments to the apex with a clearly defined tissue plane (Panels A and B, Supplementary data online, Video S1). The thickened tissue encased the posteromedial papillary muscle and obliterated the LV apex. The right ventricle was not involved.

There was moderate mitral regurgitation (Panel H) and severely restricted LV diastolic filling pressure (Panels C and F). A contrast TTE improved morphological assessment of the LV mass (Panels D and E, Supplementary data online, Video S2) and demonstrated significantly reduced perfusion in the associated underlying myocardium. Absent perfusion within most of the obliterative LV mass was seen on both contrast TTE and post-gadolinium cardiac magnetic resonance imaging, consistent with the thrombus/chronic inflammatory material (Panels G and I, Supplementary data online, Video S3).

The TTE findings met the diagnostic criteria for severe endomyocardial fibrosis (EMF). A TTE is the gold standard imaging technique used in the diagnosis of EMF and was pivotal in this case. The addition of contrast further enhanced the certainty of the diagnosis. With the liver biopsy confirming sarcoidosis, the patient was treated with corticosteroids, leading to a significant improvement in clinical state. Sarcoidosis has been reported in the literature as a possible but very rare cause of EMF.

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

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