

A Rare Case of Restrictive Cardiomyopathy with Massive Fat Accumulation on the Surface of the Heart

A 12-year-old female was admitted to our hospital for chest tightness, fatigue, and abdominal distension of unknown origin. On admission, chest x-ray displayed cardiomegaly. Transthoracic echocardiography demonstrated biatrial and right ventricular (RV) enlargement (Figure 1A) accompanied by left ventricular wall thickening and biventricular systolic dysfunction (left ventricular ejection fraction = 43.9%, tricuspid annular plane systolic excursion = 1.3 cm, RV fraction area change = 28%). Right atrial appendage aneurysm with thrombosis was also noted (Figure 1B, Video 1). Mitral valve flow spectrum and tissue Doppler imaging were

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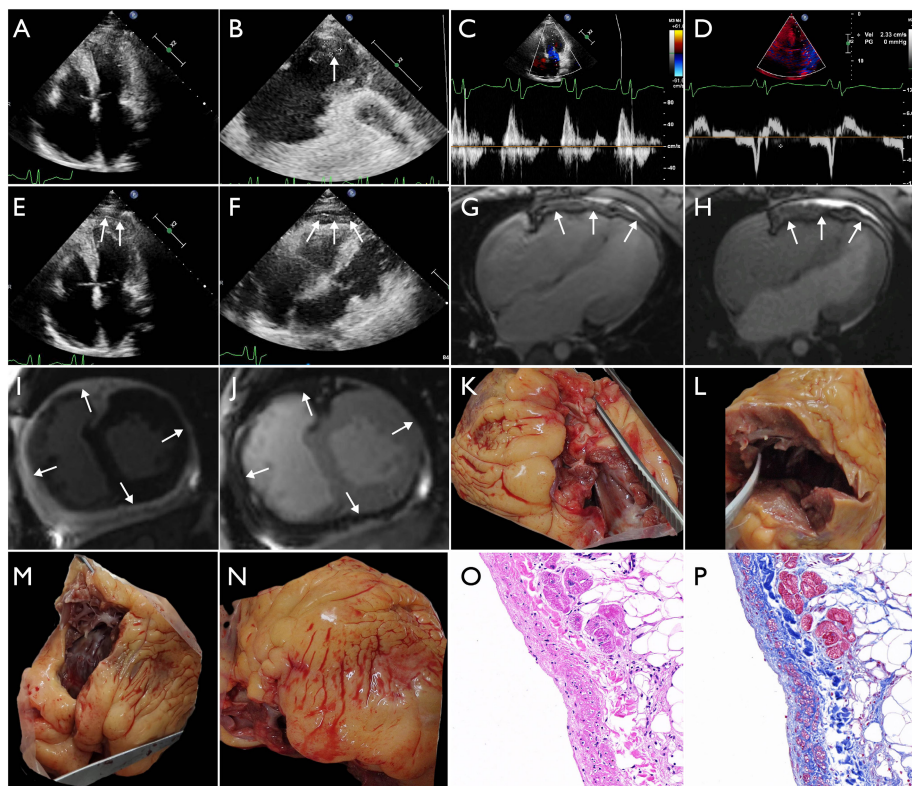


Figure 1. (A) Transthoracic echocardiography depicting biatrial and RV enlargement. (B) Transthoracic echocardiography showing right atrial appendage aneurysm with thrombosis (white arrow). (C and D) Mitral valve flow spectrum and tissue Doppler imaging revealing restrictive filling pattern. (E and F) Transthoracic echocardiography showing fat accumulation on the surface of the RV and LV (white arrows). (G-J) Cardiovascular magnetic resonance displaying fat attachment around the heart (white arrows). (K-N) Postoperative photographs confirming massive fat accumulation on the surface of the whole heart. (O-P) Histopathological examination demonstrating interstitial fibrosis and fatty metaplasia. LV, left ventricle; RV, right ventricle.

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notable for restrictive filling pattern (Figures 1C and D). Transthoracic echocardiography showed fat accumulation on the surface of the left and right ventricles (Figures 1E and F, Videos 2 and 3). Furthermore, cardiovascular magnetic resonance revealed a strip high signal area of fat around the heart, and there was fatty infiltration in the myocardium of the interventricular septum and RV free wall (Figures 1G-J). The diagnosis of restrictive cardiomyopathy with a great amount of fat accumulation on the surface of the heart was made on the basis of the aforementioned findings.

Long-term medical management failed to improve her symptoms, and she underwent heart transplantation after 1 month. Gross examination of the resected heart showed massive fatty infiltration on the surface of the whole heart (Figures 1K-N). Histopathologic examination showed interstitial fibrosis and fatty metaplasia (Figures 1O and P).

This case is an extremely rare form of restrictive cardiomyopathy, which highlights the integrated role of multimodality imaging in diagnosis and clinical decision-making.

Informed Consent: Written informed consent was obtained from the patient's parent.

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Video 1: Transthoracic echocardiography disclosing right atrial appendage aneurysm with thrombosis.

Video 2: Transthoracic echocardiography of the apical 4-chamber showing fat accumulation on the surface of the LV. LV, left ventricle.

Video 3: Transthoracic echocardiography indicating fat accumulation on the surface of the RV. RV, right ventricle.