

Cardiac Imaging

Fatty Myocardial Foci in Tuberous Sclerosis

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The patient was an asymptomatic 33-year-old woman who had tuberous sclerosis with heart involvement. Her transthoracic echocardiogram showed multiple intramyocardial homogeneous masses in the left ventricle, the largest being 19 × 15 mm in the interventricular septum (Figure 1), and normal biventricular systolic function. Computed tomography showed images in the interventricular septum with a density consistent with fat (Figure 2). The study was complemented by a cardiac magnetic resonance scan that revealed multiple nodular images in the interventricular septum and left ventricle free wall, with high intensity on T1- and lower intensity on T2-weighted images (Figure 3). There was no delayed enhancement.

Tuberous sclerosis is an autosomal dominant disorder characterised by hamartomatous changes in multiple organs, including the heart.¹ Cardiac rhabdomyomas are one of the most common manifestations of tuberous sclerosis. The presence of cardiac fatty tumours has also been described in patients with tuberous sclerosis, but their origin was unknown.² These fatty tumours may be remnants of rhabdomyomas that, according to the literature, are

rare in adults.³ The differential diagnosis of a fatty focus is a lipoma.⁴ Only histology can distinguish between them, but in this case there was no justification for performing a biopsy, since the lesions were not responsible for any symptoms.

The presence of well circumscribed fatty myocardial foci in tuberous sclerosis patients should be kept in mind as a differential diagnosis of the more classically described rhabdomyomas. In the future, these fatty foci may help identify patients suspected of having tuberous sclerosis.

References

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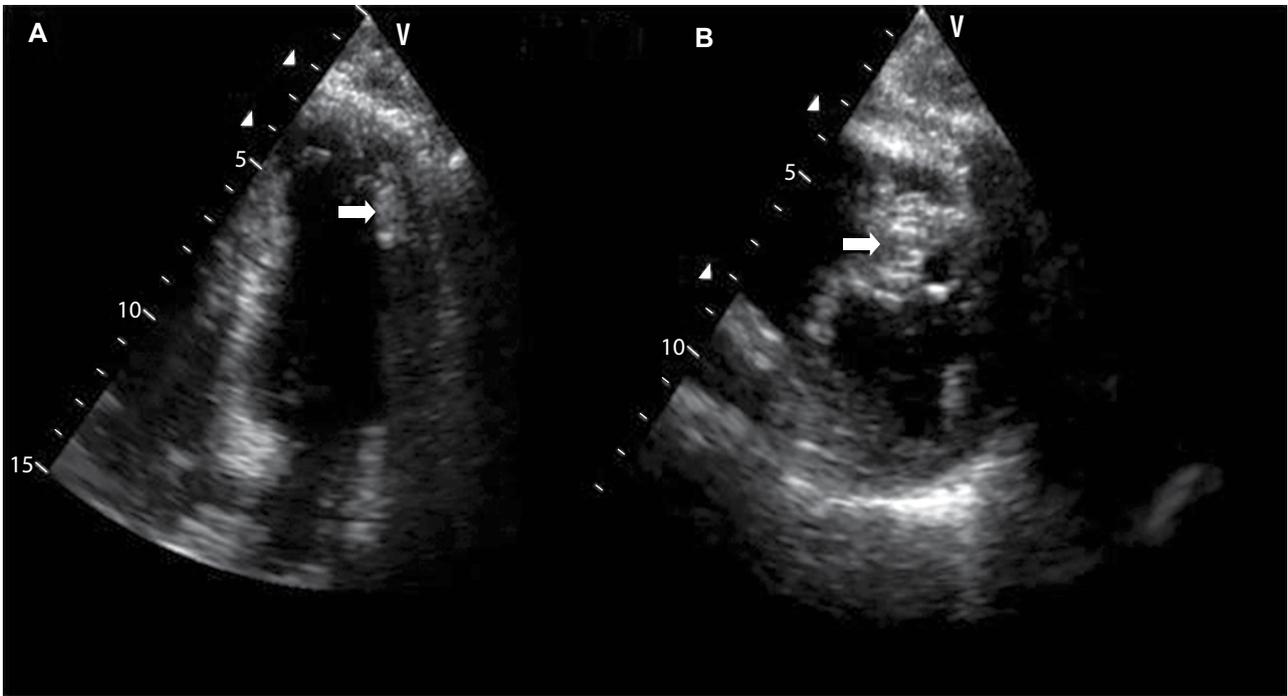


Figure 1. Transthoracic echocardiogram in apical four-chamber (A) and short-axis parasternal (B) views, demonstrating two focal areas of increased echogenicity (arrows) in the lateral left ventricular wall and in the interventricular septum, respectively.

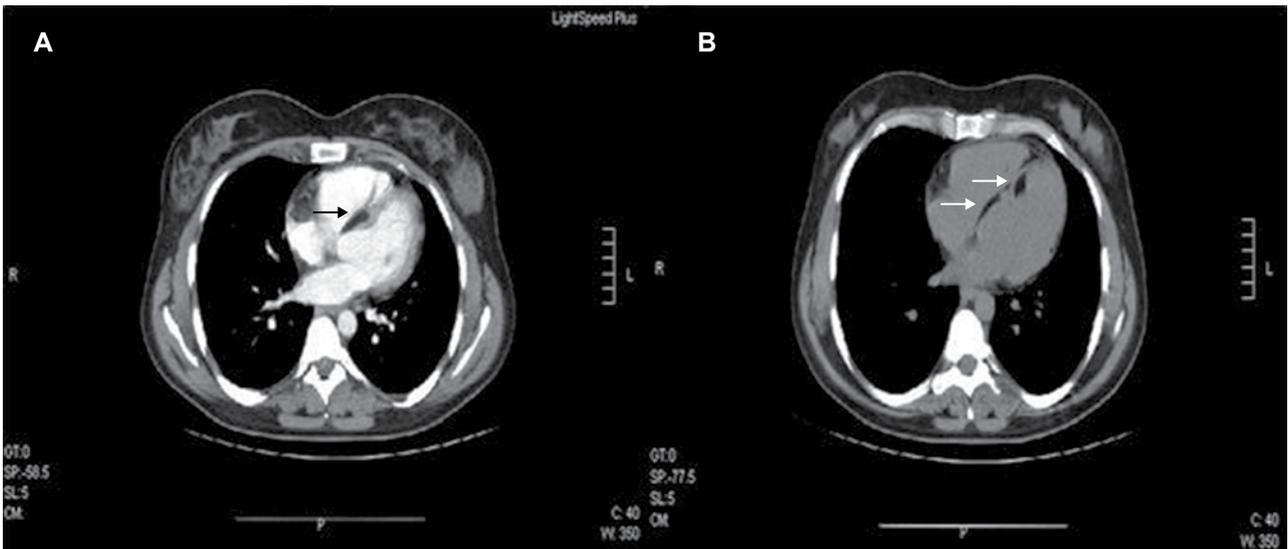


Figure 2. Thoracoabdominal computed tomography with (A) and without (B) intravenous contrast, revealing a nodular lesion in the interventricular septum with fat density (arrows).

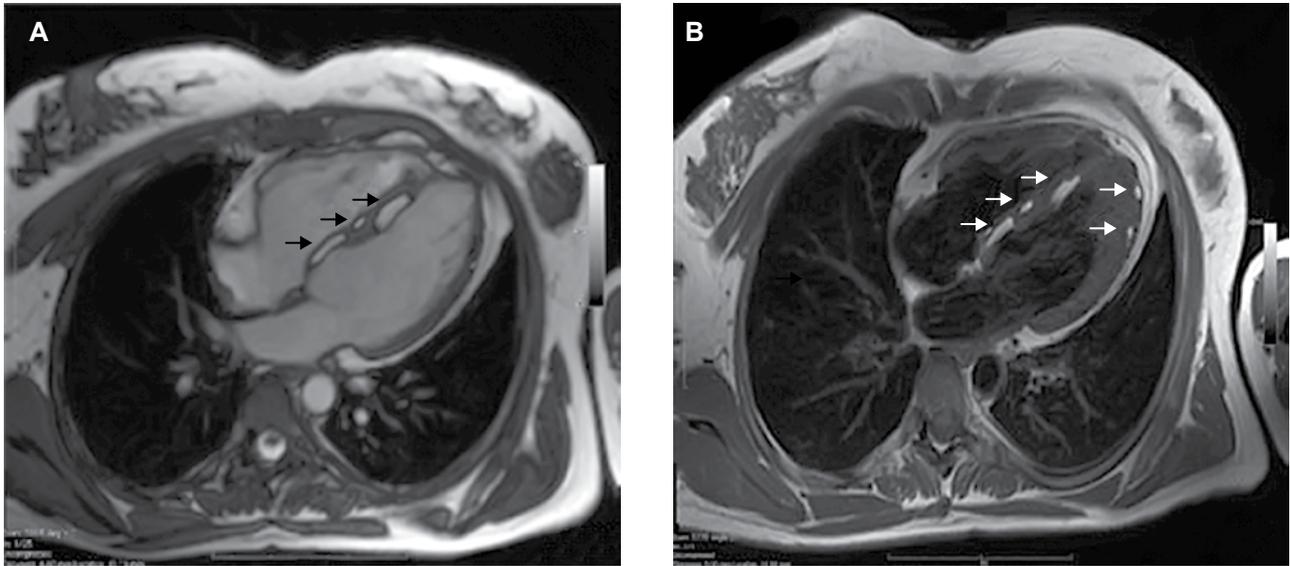


Figure 3. Cardiac magnetic resonance findings: four-chamber T1-weighted images, after (A) and before (B) gadolinium administration, showing multiple hyperintense images in the interventricular septum and left ventricle free wall (arrows), corresponding to fatty myocardial foci.