

Focal Fatty Areas in the Myocardium of Patients With Tuberous Sclerosis Complex

A Unique Finding

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Abstract: With this collection of computed tomography and magnetic resonance images, we illustrate a recently described novel finding in the myocardium of patients with tuberous sclerosis complex.

Key Words: tuberous sclerosis complex, heart, fat, myocardium, computed tomography, magnetic resonance imaging

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With this collection of computed tomography (CT) and magnetic resonance images (MRIs), we illustrate a recently described finding in the myocardium of patients with tuberous sclerosis complex (TSC). TSC is an autosomal-dominant neurocutaneous disorder characterized by tumor-like malformations involving many organ systems, including the brain (cortical tubers, subependymal nodules, and subependymal giant cell astrocytomas), the lungs (lymphangiioleiomyomatosis), the kidneys (angiomyolipomas), and the skin (facial angiofibromas or forehead plaques, shagreen patches, and hypomelanotic macules).¹

CASE REPORT

Within the field of cardiology, patients with TSC can present at a fetal or pediatric age due to the development of cardiac rhabdomyomas, which tend to regress over time. After childhood, patients with TSC who develop cardiac dysfunction suffer mostly from cardiac arrhythmias.² In Figure 1, we show 2 different patients with TSC with focal circumscribed hypodense areas within the myocardium depicted on abdominal CT scans performed to monitor their renal angiomyolipomas. On MRI, these fatty foci show the signal intensity of fat (Fig. 2).

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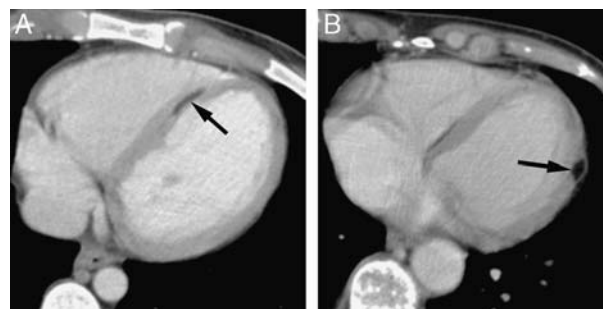


FIGURE 1. Fatty foci in the myocardium demonstrated on nongated contrast-enhanced abdominal CT scans in patients with TSC. A, Fatty focus in the interventricular septum (arrow) in a 25-year-old male patient. B, Fatty focus in the left ventricular wall (arrow) in a 59-year-old female patient.

DISCUSSION

In comparison with the known causes of fat in the myocardium (ie, epicardial fat deposits, old myocardial infarctions, arrhythmogenic right ventricular dysplasia, normal right ventricular fat, hemangiomas, and liposarcomas), the fatty foci in patients with TSC seem to have unique characteristics consisting of a combination of focality, well-circumscribed form, location into the mid-myocardium, pure fat density, absence of enhancement,



FIGURE 2. Fatty foci in the myocardium demonstrated on MRI in a 49-year-old female patient with TSC. A, Transverse dark-blood T1-weighted TSE image. Arrow points at fatty focus in the left ventricular wall. B, Short-axis dark-blood T2-weighted TSE image. Arrows point at fatty foci in the left ventricular wall. TSE indicates turbo spin echo.

and absence of invasive behavior.³ In a recently published case-control study, the majority of patients with TSC showed these well-circumscribed foci of fat density in the myocardium on abdominal CT scans, which were not found in an age-matched and sex-matched control group without TSC.³ To our knowledge, no MRIs of these characteristic fatty foci in TSC have been published yet. On echocardiography, these fatty foci can be seen as areas of increased echogenicity.⁴ Therefore, if in daily practice one encounters these types of fatty foci in the myocardium on cardiac CT, cardiac MRI, or echocardiography, one should remember to look for other features of TSC.

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