Myocardial Calcification with a Latent Risk of Congestive Heart Failure in a Patient with Apical Hypertrophic Cardiomyopathy

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Abstract

Myocardial calcification is rare. An 88-year-old man who had previously been diagnosed with apical hypertrophic cardiomyopathy exhibited left ventricular asynergy on echocardiography before undergoing cholecystectomy. Computed tomography revealed severe calcification in the apical region of the left ventricular myocardium, although the coronary arteries were intact and the hemodynamics on right heart catheterization were normal. The cause of the left ventricular asynergy appeared to be myocardial calcification, thought to be the result of rheumatic fever based on the patient’s past history. Stress echocardiography showed a latent risk for the development of heart failure due to the distensibility of the calcified left ventricular myocardium.

Key words: myocardial calcification, stress echocardiography, rheumatic fever, apical hypertrophic cardiomyopathy

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Introduction

Myocardial calcification is rare, with most reported cases having been identified postmortem (1). In each case, severe myocardial calcification was detected following the onset of symptoms of acute heart failure and the patient’s prognosis was poor, as early detection is difficult (2). The precise cause of myocardial calcification and its effects on cardiac hemodynamics remain unclear. In this report, we describe a case of myocardial calcification without symptoms in which stress echocardiography revealed a latent risk for the development of heart failure due to distensibility of the calcified left ventricular (LV) myocardium.

Case Report

An 88-year-old man with gallstones was scheduled for cholecystectomy. The patient had been hospitalized for coronary angiography after preoperative screening echocardiography revealed LV asynergy in the posterolateral and apical walls. He had not previously noticed any chest pain or dyspnea on effort; however, his personal history included rheumatic fever at 30 years of age and tuberculosis at 86 years of age. In addition, a high-density substance in the apical portion of the heart had been identified on a computed tomography (CT) scan performed at 65 years of age, which showed no other cardiovascular pathologies, including pericarditis or hemopericardium (after trauma or cardiac surgery). He was then diagnosed with apical hypertrophic cardiomyopathy on an electrocardiogram and echocardiography, although no specific treatments were started. On a physical examination, the patient’s blood pressure was 120/74 mmHg and his heart rate was 68 bpm. Auscultation revealed no cardiac murmurs or lung rales, and no rashes or lymphadenopathy were noted. The remaining findings of the physical examination were within the normal ranges, and a chest X-ray disclosed no increases in heart size or evidence of calcification in the cardiac silhouette. In contrast, an electrocardiogram showed abnormal Q waves in leads I and aVL and
negative T waves in leads V3-6 (Fig. 1). Nevertheless, 24-hour Holter monitoring recorded no life-threatening ventricular arrhythmias, and the values of hematological parameters, including eosinophils and blood chemistry data, such as the serum urea nitrogen, creatinine, bilirubin, total protein, albumin, calcium, magnesium, phosphorus and thyroxine (T4) levels, were within the normal limits. Furthermore, the B-type natriuretic peptide level was 118 pg/mL, and tests for antinuclear antibodies and rheumatoid factor were negative. A transthoracic echocardiographic examination showed a LV diastolic diameter of 64 mm, systolic diameter of 45 mm, end-diastolic volume of 80 mL, end-systolic volume of 43 mL and ejection fraction of 46%. The left atrium (LA) was also dilated (LA diameter: 43 mm, LA volume index: 57 mL/m²), although the mitral inflow pattern was consistent with the patient’s age (E wave peak: 71 cm/s; A wave peak: 103 cm/s; E deceleration time: 214 ms). A continuous wave Doppler recording indicated an estimated peak pressure gradient of tricuspid regurgitation of 21 mmHg, and an apical 4-chamber view of the LV revealed apical hypertrophy, with maximal an apical LV wall thickness of 21 mm, in addition to asynergy in the posterolateral and apical walls with high echogenicity (Fig. 2). There were no findings on Doppler echocardiography suggestive of constrictive pericarditis. Meanwhile, a CT scan of the heart revealed the presence of severe calcification, primarily in the LV myocardium (Fig. 3), and CT scans of the brain, abdomen and pelvis showed no evidence of calcium deposition. Thallium and ¹²³I-beta-methyl iodophenyl pentadecanoic acid dual-isotope single-photon emission tomography images at rest showed filling defects in the posterolateral wall and apex of the LV, and late gadolinium enhancement was observed in the hypertrophied apical portion on cardiac magnetic resonance imaging (MRI). MRI also demonstrated a low-intensity area of late gadolinium enhancement due to calcification on the endocardial side (Fig. 4). Moreover, coronary angiography disclosed normal coronary arteries, and contrast media did not flow into the apex on left ventriculography (Fig. 5). Although the patient’s hemodynamics on right cardiac catheterization were normal, stress echocardiography revealed a rise in the tricuspid regurgitation pressure gradient from 23 mmHg at rest to 50 mmHg at 70 W exercise (Fig. 6). In addition, the mitral valve regurgitation worsened from trivial to moderate during exercise. Therefore, apical hypertrophy with myocardial calcification, thereby slightly increasing the filling pressure under loading, was suspected. A cardiac evaluation was carried out prior to surgery, and we recommended that an adequate filling pressure be maintained for perioperative management for restrictive cardiomyopathy. The patient is currently awaiting surgery.

**Discussion**

It is rare that myocardial calcification can be accurately diagnosed while the patient is alive. Therefore, the precise cause of myocardial calcification and its effect on the cardiac function remain unexplained. Pathologic myocardial calcification occurs via two mechanisms: dystrophic and metastatic (1). Dystrophic calcification occurs in dead or de-
Concerning illnesses: tuberculosis (13), rheumatic fever (14), irradiation-related myocardial fibrosis (6-10), myocarditis (11), myocardial infarction (4), ventricular aneurysms (5), endomyocardial fibrosis is a disease of unknown etiology characterized by the presence of fibrous tissue in the endocardium eventually extending to the myocardium. It occurs almost exclusively in tropical and subtropical countries and is very rare outside these regions (10); the current patient had no history of travel to any endemic areas. Tuberculosis and rheumatic fever are other possibilities, as he had a past history of both conditions. However, the suspected myocardial calcification was already present when he developed tuberculosis. Therefore, we consider the etiology of myocardial calcification to be rheumatic fever rather than tuberculosis. No close inspections, such as electrocardiogram assessments, were conducted during the interval after rheumatic fever was noted and before apical hypertrophy was detected. Hence, if the calcification had been caused by the rheumatic fever, the temporal relationship between these disorders was unclear. The incidence of acute rheumatic fever is decreasing in developed countries, including Japan, as a result of improvements in sanitary conditions and upgrades to medical care systems (22, 23). Therefore, it is likely that this disease may not exist in the near future. On the other hand, based on the possibility that latent tuberculosis was not apparent as a result of the patient’s age, the myocardial calcification may have been produced by the tuberculosis at the time of the initial infection. Nevertheless, it is difficult to elucidate external factors of myocardial calcification. A similar case report described intramyocardial calcification with apical hypertrophic cardiomyopathy in which the etiology of the calcification was unclear (24). In the present case, we also allow for the possibility that the myocardial denaturation or necrosis indicated on MRI and scintigraphy resulted in the formation of myo-
Dystrophic calcification of the myocardium may occur in patients with severe myocarditis, serving as a marker of extensive damage, as reported in a patient with massive ventricular calcification who presented with diastolic heart failure (25). Pediatric myocardial calcification with myocarditis has also been reported to be a predictor of a poor outcome (26). Despite the detection of apical hypertrophic cardiomyopathy with occult myocardial calcification, which was identified fortuitously, our patient experienced no cardiac events until 88 years of age (without treatment with any medications). Overall, his clinical condition appeared to be rather favorable, with hemodynamic parameters within the normal limits. However, the myocardial calcification with apical hypertrophy induced elevation of the LA pressure under exercise stress, likely because the organic changes in the LV resulted in systolic and diastolic dysfunction with a reduction in the end-diastolic volume. It is possible that the diastolic dysfunction caused by hypertrophy further worsened due to the development of myocardial calcification. In other reports, myocardial calcification was identified at the time of onset of symptoms of acute heart failure, and the patient’s prognosis was poor (2). It is difficult to detect myocardial calcification on conventional chest radiography, and clinical signs suggestive of myocardial calcification remain unclear. In the present case, we were able to detect the myocardial calcification before the patient’s condition deteriorated to decompensated heart failure and subsequently...
evaluated the risk of acute heart failure using stress echocardiography.

We herein reported an interesting case of massive myocardial calcification with apical hypertrophic cardiomyopathy. This is the first report to evaluate the latent risk of congestive heart failure in a patient with myocardial calcification presenting with apical hypertrophic cardiomyopathy. In this case, the patient’s hemodynamic condition was stable, although there was a potential risk for congestive heart failure, and he had no history of acute heart failure because he avoided excessive physical loading. With the widespread use of CT and MRI, the number of asymptomatic patients with myocardial calcification found by chance may increase in the future. In such cases, the patient should be evaluated for the risk of heart failure using stress echocardiography, even if the cardiac activity is stable.

The authors state that they have no Conflict of Interest (COI).

References


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