Intramyocardial Calcification in a Patient with Apical Hypertrophic Cardiomyopathy

Satoshi Kaimoto, Tatsuya Kawasaki, Michiyo Yamano,
Shigeyuki Miki, Tadaaki Kamitani and Hiroki Sugihara

Abstract

Intramyocardial calcification is a very rare condition. We report a case of a 72-year-old man with apical hypertrophic cardiomyopathy, who was initially suspected of having a thrombus in the left ventricular apex on echocardiography, but was finally diagnosed as having apical intramyocardial calcification on multidetector computed tomography. The mechanism of developing intramyocardial calcification remains to be elucidated, but the patient has been stable for more than 2 years.

Key words: intramyocardial calcification, hypertrophic cardiomyopathy, echocardiography, multidetector computed tomography

(Intern Med 51: 1523-1526, 2012)

(DOI: 10.2169/internalmedicine.51.7323)

Introduction

Intramyocardial calcification is a very rare condition. We report a patient with apical hypertrophic cardiomyopathy, who was diagnosed as having intramyocardial calcification on multidetector computed tomography.

Case Report

A 72-year-old man was referred to our hospital because of an electrocardiographic abnormality. The patient reported no chest pain or palpitations. He had hypertension and diabetes mellitus that had been well controlled with metformin hydrochloride (500 mg once daily) and telmisartan (40 mg once daily). He was diagnosed as having idiopathic pulmonary fibrosis five years earlier and had received prednisolone (5 mg twice daily) and azathioprine (100 mg once daily) with domiciliary oxygen therapy. He had never smoked.

On examination, his blood pressure was 124/72 mm Hg, his pulse was 80 beats per minute and regular, and oxygen saturation was 90% while he was breathing oxygen at 2 liters per minute via a nasal cannula. The jugular veins were not distended; fine inspiratory crackles were heard throughout both lung fields; heart sounds were normal; and pretibial

edema was not present.

Electrocardiography showed premature atrial contractions and left ventricular hypertrophy (Fig. 1). A chest X-ray showed a diffuse reticular pattern in both lung fields that was unchanged compared with a few years earlier (Fig. 2). Blood examination was unremarkable; serum creatinine, 0.53 mg/dL; C-reactive protein, 0.10 mg/dL; low density lipoprotein cholesterol, 112 mg/dL; brain natriuretic peptide, 37 pg/mL. Transthoracic echocardiography showed a left ventricular ejection fraction of 58%, with apical hypertrophy accompanied by an abnormal mass in the apex of the left ventricle (Fig. 3; Supplementary movie 1). Mitral annular calcification was not detected. The remainder of the echocardiographic examination was normal.

The patient was admitted to our hospital and an initial diagnosis of apical hypertrophic cardiomyopathy with an apical thrombus was made. However, the size of the apical thrombus had not changed after the initiation of anticoagulant therapy with unfractionated heparin and warfarin for more than 2 weeks. A 64-detector computed tomography scanner (LightSpeed VCT, GE Healthcare, Milwaukee, Wisconsin, USA) revealed marked calcification in the left ventricular apex, without coronary stenosis or coronary calcification (Fig. 4). The apical calcification was not detected on chest X-ray. On the basis of the detailed morphological

Figure 1. Electrocardiography. Premature atrial contractions and left ventricular hypertrophy with negative T waves in leads I, II, aVL, and V3 to V6 are observed.

Figure 2. Chest X-ray. The cardiothoracic ratio was 58% and a diffuse reticular pattern compatible with pulmonary fibrosis was remarkable in both lung fields.

analysis of multidetector computed tomography, apical calcification was considered to be intramyocardial without a thrombus (Fig. 5). Intramyocardial calcification in the left ventricular apex was clearly demonstrated on three-dimensional computed tomography (Supplementary movie 2).

The patient was finally diagnosed as having apical hypertrophic cardiomyopathy with intramyocardial calcification. He had a normal blood calcium level (albumin-corrected total calcium concentration, 9.3 mg/dL) without a detectable disturbance in calcium metabolism, e.g., hyperparathyroidism (intact parathyroid hormone, 37 pg/mL [normal range, 10 to 65 pg/mL]), sarcoidosis, or malignancy (e.g., cytokeratin-19 fragments, 2.3 ng/dL [normal range, ≤ 3.5 ng/dL] and pro-gastrin-releasing peptide, 29.3 pg/mL [normal range, ≤ 46 pg/mL]). The apical calcification seemed to gradually increase in size (Fig. 6). After being discharged from our hospital, the patient has been stable for more than 2 years without anticoagulant therapy. Fortunately, morpho-

logical changes in the apical intramural calcification were not obvious on follow-up computed tomography.

Discussion

We report a case of a 72-year-old man with apical hypertrophic cardiomyopathy accompanied with intramyocardial calcification. The patient was initially suspected of having an apical thrombus on echocardiography, but was finally diagnosed as having intramyocardial calcification on multidetector computed tomography.

Coronary artery calcification and aortic valve calcification are known to occur in elderly patients. Mitral annulus calcification seems to be not uncommon in patients with hypertrophic cardiomyopathy (1-4). Some patients with hypertrophic cardiomyopathy were reported to have left ventricular endomyocardial calcification (5, 6) or papillary muscle calcification (7). Apical calcification was also reported in a few cases with apical hypertrophic cardiomyopathy, but all of the calcifications were more likely to be associated with an apical thrombus accompanied by apical aneurysm formation (8, 9). Neither an apical thrombus nor apical aneurysm, including a small pouch, was observed in the present patient.

The mechanism of developing intramyocardial calcification remains to be elucidated in the present case. Pulmonary tuberculosis has been reported to be associated with intramyocardial calcification (10, 11), but our patient did not have a history of tuberculosis. He had been taking prednisolone and azathioprine for years. Immunosuppressive agents may be a possible trigger for calcification (12). However, his drugs were not likely to be the cause of the apical calcification. A small calcification had already been detected in the apex of the left ventricle on computed tomography before he started taking these drugs. Stagnation or turbulence of blood flow associated with apical hypertrophic cardio-

Figure 3. Transthoracic echocardiography. An apical four-chamber view (A) and the close-up (B) show apical hypertrophy with an abnormal high-echogenic mass, 13 mm at its longest in diameter, in the apex of the left ventricle.

Figure 4. Multidetector computed tomography. Axial scans at the level of the left ventricular apex show marked calcification in the apex of the left ventricle (A, plain; B, with contrast media).

Figure 5. Morphological analysis on multidetector computed tomography. Detailed analysis of the left ventricular apex shows that attenuation values in Hounsfield units in the region close to apical calcification are almost all similar to those of the left ventricular myocardium. Note systolic thickening of the tissue between the calcification and left ventricular cavity (arrows; A, end-diastolic; B, end-systolic), and the direct link of the tissue to the left ventricular trabeculations (arrowheads; C and D), indicating that the apical calcification was surrounded by myocardium without a thrombus.

Figure 6. Change in size of the apical calcification. Note the increased size of apical calcification at the same level on computed tomography on this admission (A), as compared with 5 years earlier (B).

myopathy may traumatize the endomyocardium, resulting in calcification with extension into the myocardium (6) although his apical hypertrophy was not severe enough to cause systolic obliteration of the middle part of the left ventricle.

The intramyocardial calcification in the present patient did not seem to increase in size after his discharge, but careful follow-up is needed because left ventricular calcification may be a cause of adverse events, e.g., arrhythmia or embolism.

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

References

1. Wanderman KL, Margulis G. Coexistence of hypertrophic obstructive cardiomyopathy and mitral annular calcification: proposed etiologic relationship. *Isr J Med Sci* **15**: 422-425, 1979.
2. Lindvall K, Herrlin B. Mitral annulus calcification, systolic anterior motion of the anterior mitral leaflet and outflow obstruction in two patients without hypertrophic cardiomyopathy. An echocardiographic report. *Acta Med Scand* **209**: 513-518, 1981.
3. Akiya K, Hayashi T, Kobayashi S, et al. Echocardiographic observation of appearance and progression of mitral annular calcification in a patient with familial obstructive hypertrophic cardiomyopathy. *J Cardiol* **31**: 115-121, 1998 (in Japanese).
4. Okamoto H, Tamenishi A, Itoh Y, Niimi T. Mitral annular reconstruction with anterior leaflet flip-over in concomitant surgery for mitral regurgitation associated with extensive posterior annular calcification and hypertrophic obstructive cardiomyopathy. *Jpn J Thorac Cardiovasc Surg* **52**: 104-106, 2004.
5. Bolz KD, Bathen J, Skjaerpe T, et al. Massive left ventricular endomyocardial calcification associated with apical hypertrophic cardiomyopathy. *Rofo* **149**: 436-437, 1988.
6. Kvaerness J, Rinck PA, Bolz KD, et al. MR imaging of apical hypertrophic cardiomyopathy with left ventricular endomyocardial calcification. *J Comput Assist Tomogr* **15**: 489-491, 1991.
7. Ross EM, Rosing DR, Laidlaw JC, et al. Impaired left ventricular systolic and diastolic function without left ventricular dilatation associated with papillary muscle calcification in hypertrophic cardiomyopathy. *Am J Cardiol* **57**: 488-490, 1986.
8. Gradaus R, Lentschig M, Wichter T. Atypical hypertrophic cardiomyopathy with calcified, left ventricular apical thrombosis. *Z Kardiol* **88**: 290-295, 1999.
9. Juergens KU, Wessling J, Fallenberg EM, et al. Multislice cardiac spiral CT evaluation of atypical hypertrophic cardiomyopathy with a calcified left ventricular thrombus. *J Comput Assist Tomogr* **24**: 688-690, 2000.
10. Butz T, van Bracht M, Meissner A, et al. Mitral valve disease as well as uncommon extensive epicardial and intramyocardial calcification secondary to massive mitral annular calcification. *Eur J Echocardiogr* **11**: 85-88, 2010.
11. Shiga Y, Miura S, Nakamura A, et al. Intramyocardial calcification with mitral annular calcification. *Intern Med* **50**: 1857-1858, 2011.
12. Nemere I, Norman AW. Steroid hormone actions at the plasma membrane: induced calcium uptake and exocytotic events. *Mol Cell Endocrinol* **80**: C165-C169, 1991.