A 45 year-old man had hypercholesterolemia with well diet control. He complained sharp pain at the left side of chest wall, without aggravation during exertion, for 3 months. Last attack was about 10 hours ago. He denied any dyspnea or syncope. He had one brother who had sudden cardiac death at the age of 44. Physical examination revealed normal findings. ECG was performed as shown in Figure 1. There was left ventricular hypertrophy (LVH), ST depression in V4-V5, giant negative T wave in V3-V6, I, II, III, and AVF. Cardiac enzymes were obtained to rule out non-ST elevation MI and the result became negative. Since the chest pain was atypical and there were ECG clues, apical hypertrophic cardiomyopathy was suspected. Transthoracic echocardiography showed LVH with good left ventricular (LV) contraction, no wall motion abnormality; however the apex could not be thoroughly inspected due to limited image quality. Cardiac magnetic resonance imaging (MRI) was requested to evaluate the possibility of apical hypertrophic cardiomyopathy. Cine MRI from two and four chamber views demonstrated predominant hypertrophy of LV apex with a spade-like configuration of LV at the end-diastole and an obliteration of LV apex at the end-systole as shown in Figure 2a to 2d. LV ejection fraction and wall motion were absolutely normal. The unique technique of MRI, delayed enhancement, revealed hyperenhancement at the area of LV apex (Figure 3) that was the typical pattern in this disease. Regarding to characteristic MRI and ECG results, the diagnosis of apical hypertrophic cardiomyopathy (HCM) was made in this patient.
Figure 2. Shows cine MRI findings, from two-chamber view during diastole (A) and systole (B), and from four-chamber view during diastole (C) and systole (D)

Discussion

HCM is the most common inherited cardiomyopathy with a wide variety of clinical and morphological expression. Apical HCM is one of the variant forms. This disease has been first discovered in Japan in 1976. The prevalence is relatively common in Japan up to 25% of all HCM cases, whereas the prevalence in the other countries is only 1-2% (1). From these reasons, it was called Japanese-type HCM. Atypical angina is the most frequent symptom, whereas typical angina may also happen due to impaired vasodilatory reserve. In addition, dyspnea and palpitation from atrial and ventricular arrhythmia can occur. According to nonspecific symptoms, primary clinical suspicion comes from ECG finding. ECG hallmark is giant negative T wave in the precordial leads in addition to LVH. There are other differential diagnoses from this ECG such as myocardial ischemia and central nervous system lesions, especially subarachnoid hemorrhage. Left ventriculogram in right anterior oblique projection demonstrated a spade-like (ace

Figure 3. Delayed enhancement MRI images in long axis (A) and 4-chamber view (B) demonstrate the typical pattern of myocardial fibrosis in apical hypertrophic cardiomyopathy
of spades) configuration of the left ventricle at end-diastole (2). MRI provides multimodality assessment in many cardiac diseases and is the gold standard in chamber assessment. In HCM, it can assess LV morphology, including LV thickness, mass, and volume, function, LV outflow tract obstruction, mitral regurgitation, and papillary muscle abnormalities, that can occur in this patient group. Moreover, the typical spade-like configuration similar to ventriculogram, can be demonstrated on long axis MRI images (3). Delayed enhancement technique is the unique property of cardiac MRI. It can localize and quantify the infarct in the setting of ischemic cardiomyopathy. In aspect of non-ischemic cardiomyopathy, the characteristic patterns in different diseases can help to make a diagnosis. In HCM, delayed enhancement is attributed to myocardial disarray, myocardial ischemia, and replacement fibrosis. The common site of this fibrosis corresponds to the area of thickness, such as typical area of superior and inferior insertion of septum in asymmetrical HCM, and apex in apical HCM. MRI is therefore a valuable technique for diagnosis, risk stratification, and follow-up in patients with HCM.

References