

Detection of apical hypertrophic cardiomyopathy; which is the appropriate imaging modality

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Apical hypertrophic cardiomyopathy is a form of hypertrophic cardiomyopathy localized to the left ventricular apex. The disease is common in Japanese and other Asian populations but its presence has also been recognized, though less commonly, in non-Asian patients. Its prognosis is relatively benign in terms of cardiovascular mortality but morbid sequelae, such as diastolic dysfunction, left atrial enlargement, apical thrombi, ventricular aneurysms, and myocardial infarction, may occur. In patients with apical hypertrophic cardiomyopathy, the electrocardiographic changes and symptoms associated often mimic acute coronary syndromes. Invasive or noninvasive evaluation of the left ventricular cavity confirms the diagnosis of apical hypertrophic cardiomyopathy, with the “ace-of-spades” sign on left ventriculography being pathognomonic. It is therefore of utmost importance to use the appropriate imaging modalities to establish the correct diagnosis of apical hypertrophic cardiomyopathy. For instance, using doppler-echocardiography in 182 patients with apical hypertrophic cardiomyopathy, three morphologically distinct phenotypes i.e. (1) pure focal, (2) pure diffuse, and (3) a mixed type according to patterns of hypertrophy could be discerned [1]. Detailed subtyping turned out to be important in the prediction of

development of atrial fibrillation, left atrial volume index and left ventricular longitudinal function.

In the current edition of this issue, Duygu et al. [2] studied 17 patients with chest pain who had a pre-diagnosis of coronary artery disease but a final diagnosis of apical hypertrophic cardiomyopathy. Interestingly, all patients showed typical spade-like appearance of the left ventricle by contrast ventriculography and giant negative T-waves on the electrocardiogram. Transthoracic echocardiography showed a maximum wall thickness of 18 ± 4 mm and a mean gradient of 30 mmHg by Doppler echocardiography. None of the patients showed significant coronary artery disease; four patients showed myocardial bridging. The authors recommended that the diagnosis of apical hypertrophic cardiomyopathy should always be considered in certain patients with typical anginal symptoms and outspoken negative T waves on the electrocardiogram. As a result, physicians caring for patients with chest pain should consider apical hypertrophic cardiomyopathy in their differential diagnosis in case of a patient with chest pain and electrocardiographic changes suggestive of coronary artery disease. Stated by the authors, these patients should all undergo echocardiography to separate apical hypertrophic cardiomyopathy from acute myocardial ischemia. Although the authors mention that ischemic evaluation was not performed, they only refer to thallium myocardial scintigraphy as potential modality to identify ischemia in apical hypertrophic cardiomyopathy. Indeed, myocardial scintigraphy

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using perfusion tracers and metabolic markers have shown to be useful in the evaluation of cardiomyopathies [3–6].

It is, however, noteworthy that the authors did not refer to cardiac magnetic resonance imaging (CMR) as an accurate and reliable means of evaluating cardiac morphology, ventricular function, and myocardial perfusion, both for the left and the right ventricle therewith covering a whole spectrum of cardiac diseases [7–12]. In particular, CMR is very well suited for identifying and characterizing patients with hypertrophic cardiomyopathy [13, 14]. For instance, Tsukamoto et al. [15] showed systolic outward motion of the left ventricular apical wall in patients with apical hypertrophic cardiomyopathy using CMR tagging. In addition, CMR allows accurate evaluation of myocardial ischemia and infarction with the potential to establish the transmural extent of myocardial infarction with high spatial resolution [16–18]. Late gadolinium-enhanced CMR can clearly delineate subendocardial infarction and CMR appears to be more sensitive than other imaging methods in detecting small infarcts and ischemia [19–21]. In recent years, late gadolinium enhancement CMR has also been used to visualize myocardial interstitial abnormalities in patients with different forms of cardiomyopathies [22, 23]. In patients with apical hypertrophic cardiomyopathy, first-pass gadolinium delayed-enhanced CMR may show local delayed enhancement in the subendocardium in the hypertrophic lesion. The presence of perfusion defect and delayed enhancement may be a marker of a high-risk patient with apical hypertrophic cardiomyopathy [24]. On the other hand, patients with apical hypertrophic cardiomyopathy may show focal hyperenhancement of the non-hypertrophic basal lateral segment of the left ventricle, and absence of hyperenhancement of the hypertrophied apical segments [25].

It can be concluded that apical hypertrophic cardiomyopathy is a disease that may mimic coronary artery disease in a certain subset of patients. Hence, adequate diagnostic methods are needed to distinguish between these two entities. Although echocardiography remains the first line of investigation in these patients the final answer might be given by CMR.

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