Hypertrophic cardiomyopathy with apical aneurysm: Unraveling contribution and usefulness of gating to myocardial perfusion SPECT and a multi-modality investigation

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INTRODUCTION

Hypertrophic cardiomyopathy, as a primary condition or secondary to systemic hypertension, is a fairly common problem with a significant impact on the LV function and patient outcome. Multiple morphological variants have been described, in which, hypertrophic cardiomyopathy with apical aneurysm (HCM-AA) is among the rarer ones.[1,2] The present article deals with the contribution of gating to myocardial perfusion SPECT in unraveling such problem in a patient without prior diagnosis.

CASE HISTORY

A 47-year-old man was referred to our laboratory because of recent development of chest pain and dyspnea on exertion. Except a long history of hypertension, no other cardiovascular risk factor was present. On EKG, there were high-voltage R waves and somewhat symmetric giant negative T waves in favor of LV myocardial hypertrophy (Figure 1). The patient underwent a dipyridamole stress/rest gated myocardial perfusion SPECT. Perfusion images were negative for any regional defect, but some longitudinal elongation of the LV and nonuniformity of thickness of myocardial walls were noted (Figure 2A, B). Gated SPECT revealed obvious dyskinesia during stress (Figure 2C) and akinesia at rest (not shown) in the apex. Close inspection of gated tomographic slices (Figure 3) demonstrated an unusual appearance of the LV cavity during systole (i.e., dual-chamber appearance with a narrow interconnecting tunnel). As suspected of HCM-AA, to verify the presence of apical aneurysm and midventricular obstruction, an echocardiography (Figure 4) and invasive angiography and ventriculography (Figure 5) were subsequently performed that, then, confirmed the diagnosis.

DISCUSSION

HCM-AA is an uncommon morphological variant of HCM in that hypertrophy is more prominent in midventricular region of the LV accompanied by aneurysmal formation in the distal apical region. This phenotype is in direct contradiction to apical hypertrophy variant, which shows localized hypertrophy in the apex. Depending on the degree of hypertrophy, an obstruction to the outflow of apical LV cavity may occur and lead to an increased pressure and thus formation of aneurysm. EKG is frequently as the first-line tool and, in cases with marked hypertrophy, may show a typical diagnostic pattern as in our case. Echocardiography and

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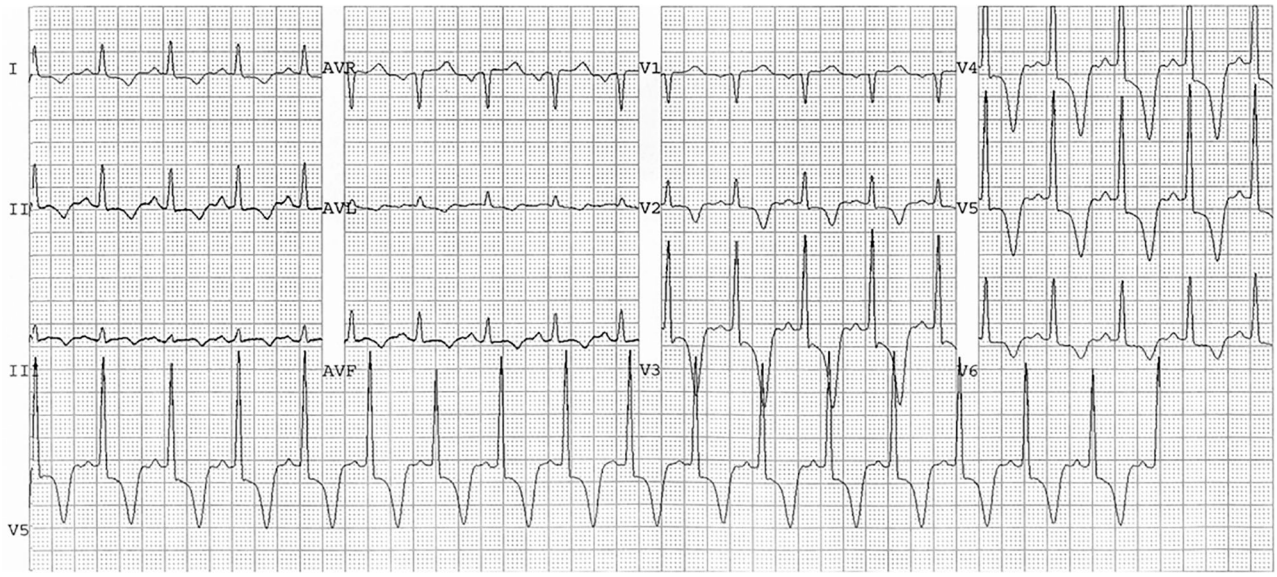

Figure 1. Resting 12-lead EKG demonstrated a sinus rhythm, normal electrical axis, high-voltage R waves, ST depression and somewhat symmetric giant negative T waves more prominently in precordial leads. Findings were in favor of LV hypertrophy.

particularly cardiac MRI depict anatomical details of the LV elegantly and are able to clearly differentiate the two variants and, therefore, are standard methods for assessment of such conditions. However, radionuclide imaging modalities may be utilized for perfusion and functional evaluation of the LV and in some circumstances may be the first modality in undiagnosed patients.[1–4]

Contrary to the typical findings on echocardiography, cardiac CT and MRI and also invasive ventriculography which are well described in the literature, gated myocardial perfusion SPECT is less investigated in such patients. However, similar findings are present as in this patient. The point, in this case, is the contribution of gating that helped unravel the regional functional abnormality, despite normal perfusion, of the apex and midventricular obstruction during systole directed us to the diagnosis. Against apical HCM, in HCM-AA, there is no solar pattern or downscaling effect of the apex on other walls in perfusion polar plots, but apical dyskinesia is present.

Apical aneurysm predisposes thrombus formation and even triggers arrhythmia from scar regions especially in large ones. For this reason, patients with HCM-AA may be considered as the subgroup with the highest risk and, therefore, differentiation from other subgroups using available imaging modalities is of interest.[5]

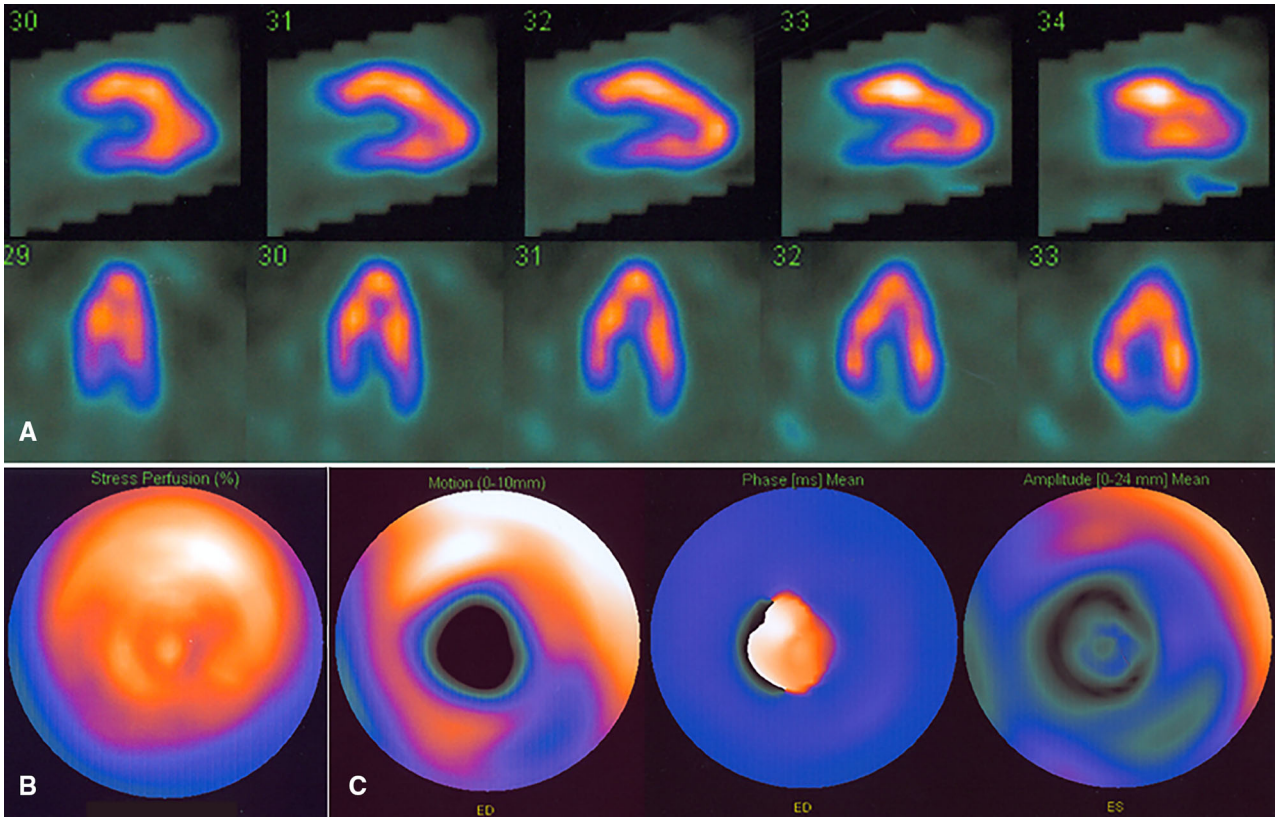

Figure 2. **A** Summed vertical- and horizontal-axis tomographic slices of the myocardial perfusion SPECT; no perfusion defect was noted but the LV seemed to be longitudinally elongated with seemingly ''bulged'' or ''outpouched'' apex. This appearance, in some way, may be resembled as the so-called spade-shaped LV. Polar plot of the perfusion **B** was fairly regular and uniform in brightness without any distinct regional dominance of intensity. **C** Polar plots of motion, phase and amplitude from gated imaging; polar plot of motion is strikingly abnormal in the apex and suggests an akinesia or dyskinesia. Polar plot of the phase confirms the presence of a dyskinesia in the apex.

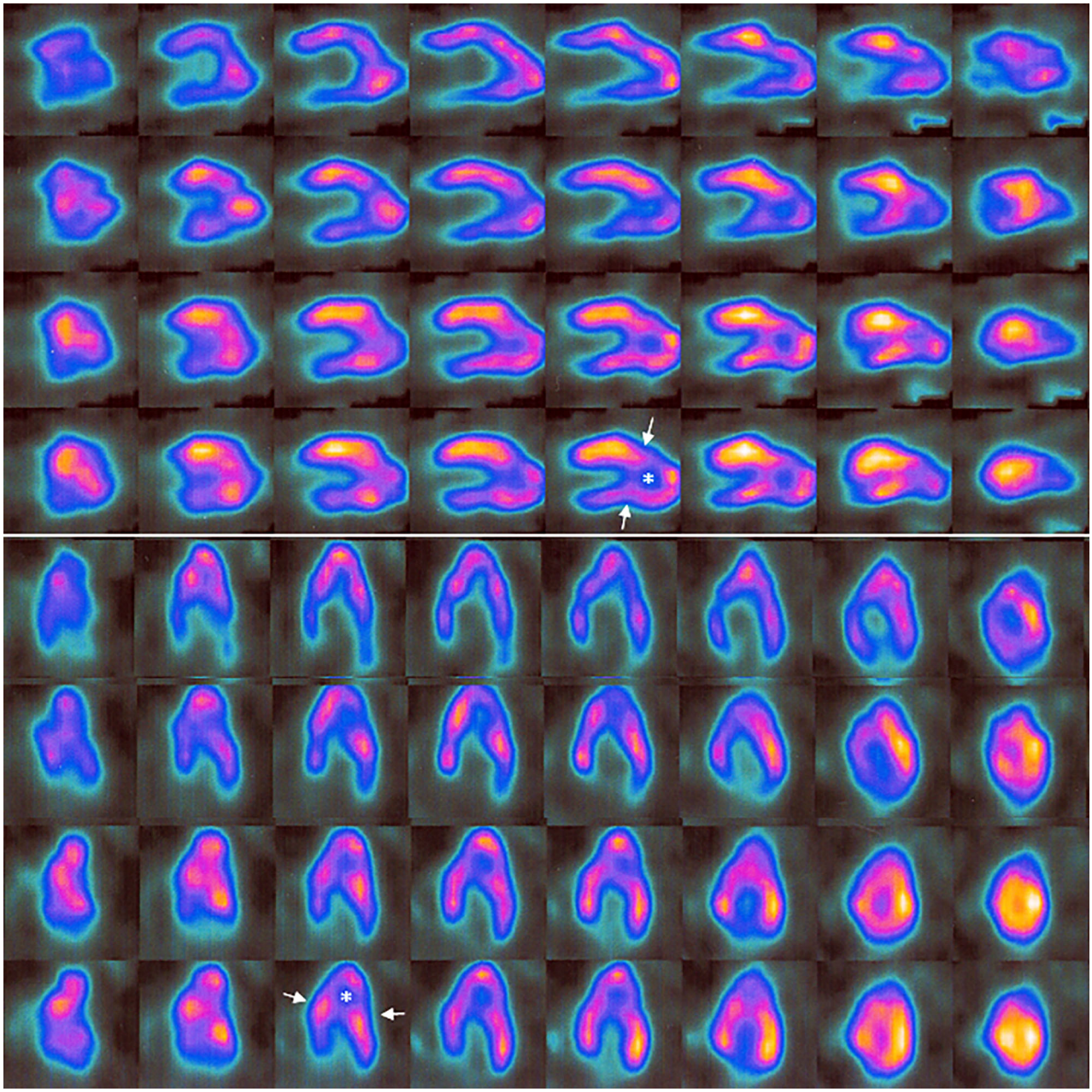

Figure 3. Gated vertical- and horizontal-axis tomographic slices from Frame 1 (end-diastole) to Frame 4 (end-systole) of the 8-framed gated SPECT; LV cavity did not show the normal conical shape, and the thickness of myocardial walls are irregular. In the end-systole, the cavity is divided into two chambers (distal and proximal) by a circumferentially hypertrophied myocardium in midventricular region of the LV (shown by small arrows). This appearance of the LV is similar to an ''hour-glass'' or a ''nutcracker'' in shape. These findings suggest aneurysmal formation in the apex (asterisk on the image) during systole with an acceptable myocardial thickness of the walls, although nonuniform with more thinning in lateral wall.

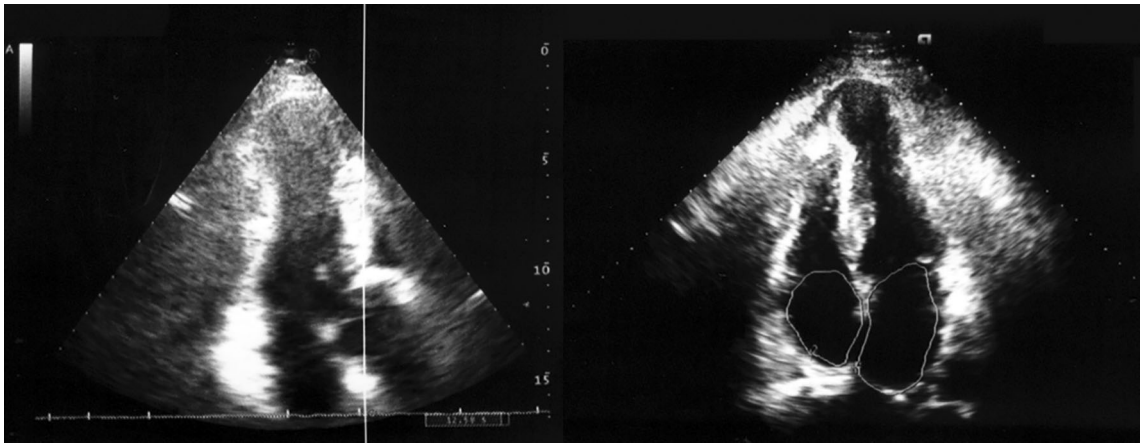

Figure 4. On echocardiography, interventricular septum, and posterior wall were 18 mm and 14 mm, respectively. Normal LV size and systolic function (EF: 50%), widening of the cavity in apical region, suspicious mild apical and midventricular hypertrophy and a mild gradient across LV cavity at rest were also evident.

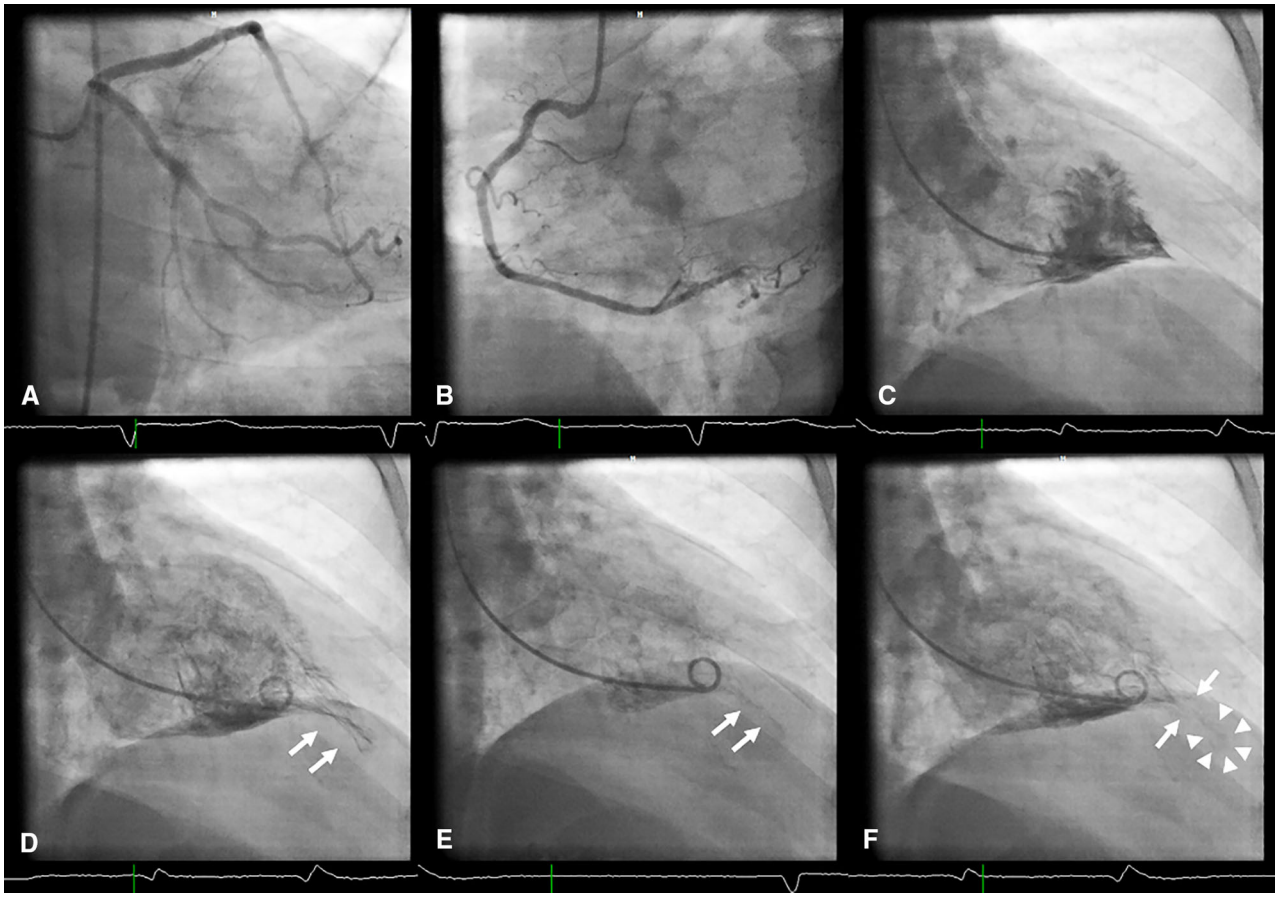

Figure 5. **A** and **B** Angiography showed normal coronary vessels. **C–F** Ventriculography revealed normal LV size, normal LV systolic function, no significant intracavitary pressure gradient but abnormal shape, or the so-called spade-shape appearance of the LV. During the systole, a ''bird's beak'' shape of the apical region of the LV cavity was evident (**C**) and during the diastole, a small amount of contrast moved forward in the apical region, giving an impression of the presence of another smaller chamber distally (**D**). During the systolic phase, when closely inspected, a small area of retention of contrast was noted distal to the main proximal chamber, interconnecting through a very thin tunnel (**E** and **F**).

Disclosure

The authors have no conflict of interest to declare.

References

1. Minami Y, Haruki S, Hagiwara N. Phenotypic overlap in hypertrophic cardiomyopathy: Apical hypertrophy, midventricular obstruction, and apical aneurysm. J Cardiol. 2014;64:463–9.
2. Kherada N, Vinardell JM, Mihos CG, Santana O. Apical hypertrophic cardiomyopathy with left ventricular apical aneurysm: Importance of multi-modality imaging. Echocardiography. 2017;34:1392–5.
3. Cianciulli TF, Saccheri MC, Masoli OH, Redruello MF, Lax JA, Morita LA, et al. Myocardial perfusion SPECT in the diagnosis of apical hypertrophic cardiomyopathy. J Nucl Cardiol. 2009;16:391–5.
4. Sun JP, Yang XS, Wong KT, Yu CM. Hypertrophic cardiomyopathy with apical aneurysm. Int J Cardiol. 2015;184:394–6.
5. Maron MS, Finley JJ, Bos JM, Hauser TH, Manning WJ, Haas TS, et al. Prevalence, clinical significance, and natural history of left ventricular apical aneurysms in hypertrophic cardiomyopathy. Circulation. 2008;118:1541–9.