Apical Hypertrophic Cardiomyopathy of the Japanese Type Coexistent with a Coronary Muscle Bridge

A Case Report and Review

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SUMMARY

We describe the case of a 21-year-old Italian male who presented with giant negative T-waves and left ventricular hypertrophy on the electrocardiogram suggestive of apical hypertrophic cardiomyopathy. Clinically, he suffered from new onset of exertional angina, dyspnea and palpitations during soccer playing or heavy exercise beginning one week before admission.

Echocardiography and cardiac catheterization confirmed the rare combination of a nonobstructive apical hypertrophic cardiomyopathy of the “Japanese” type coexistent with an extensive muscular bridge involving almost the entire anterior interventricular branch of the left coronary artery.

Although the patient complained of exertional angina pectoris, absence of objective evidence of myocardial ischemia by means of treadmill stress test, exercise thallium scan, dobutamine stress echocardiography as well as atrial pacing stress emphasized the benign nature of this complex anomaly. (Jpn Heart J 1997; 38: 741-748)

Key words: Apical hypertrophic cardiomyopathy, muscle bridge, myocardial ischemia, atrial pacing, lactate metabolism

Apical hypertrophic cardiomyopathy (AHC) of the “Japanese” type is almost completely restricted to Japanese patients and is thought to be exceedingly rare in non-Asians. Although the clinical course is regarded to be benign, myocardial ischemia of multifactorial etiology is a relatively common finding in AHC. Occasionally a coexistent anomaly of the coronary vasculature can be identified as a potential cause of myocardial ischemia. However, the coexistence of AHC with muscular bridges is exceedingly uncommon. Usually the principal branches of the coronary arteries have a subepicardial

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course. However, when a segment of the artery runs within myocardial tissue a muscular bridge is formed.\textsuperscript{11} Again, this coronary anomaly is regarded as benign\textsuperscript{11} although reports have linked myocardial bridging with myocardial ischemia, infarction, and sudden cardiac death.\textsuperscript{12,13}

In order to provide optimal treatment, which largely depends up on the detection of myocardial ischemia, we performed an extensive metabolic, scintigraphic, echocardiographic and electrocardiographic evaluation for myocardial ischemia which is presented in the following case report.

**CASE REPORT**

A twenty-one-year old otherwise healthy Italian was referred for evaluation of new onset of exertional angina and dyspnea which occurred repeatedly during soccer playing. Electrocardiography revealed electrocardiographic abnormalities suggestive of left ventricular hypertrophy and myocardial ischemia. Family history was negative for cardiovascular diseases or thrombophilia. Physical exami-
Figure 2. Two-dimensional echocardiography (apical view) reveals hypertrophy limited to the left ventricular apex.

Figure 3. Left ventriculogram obtained in the right anterior oblique (30° RAO) projection, showing the spade-like appearance of the apex at end-diastole.

nation was unremarkable; in particular heart sounds were normal, and no murmur or friction rub was heard on cardiac auscultation. On palpation, the apical precordial impulse was slightly displaced laterally and sustained. Blood pressure was 130/80 mmHg.

All laboratory values including blood sedimentation rate and C-reactive protein, leukocyte and erythrocyte cell count and blood coagulation were
unremarkable. In particular, prothrombin time (INR) was 1.0, aPTT 29 seconds, leukocyte count 8.4/nl, erythrocyte count 5.3/pl, thrombocyte count 157/nl. Chest x-ray film findings were normal. The electrocardiogram showed sinus rhythm with tall R-waves, especially in leads V3 and V4 and deep inverted T-waves in the inferior and precordial leads, especially in leads V2–V4 (12 mm in V3) (Figure 1). Two-dimensional echocardiography revealed marked hypertrophy of the left ventricular apex and the distal one third of the septum and lateral wall, compatible with AHC (Figure 2). Localized hypertrophy was best visualized in
the apical view and appeared to be located below the papillary muscle level in the short parasternal axis plane. There was no intraventricular gradient detectable on echocardiographic Doppler examination. On cardiac catheterization, the left ventricular end-diastolic pressure was 8 mmHg, and there was no intraventricular pressure gradient. Left ventriculography revealed excellent systolic performance with apical obliteration and an "ace-of-spades" appearance at end-diastole (Figure 3). Selective coronary angiography revealed an extensive muscle bridge over almost the entire left anterior descending artery with subtotal systolic compression of the coronary artery, but normal diameter and unimpeded flow during diastole (Figure 4). The rest of the coronary vasculature was normal. A Doppler coronary flow measurement to reveal any abnormal flow velocity pattern was not performed in this patient.

On treadmill exercise test he completed stage 4 of the Bruce protocol, corresponding to 13.3 MET until he experienced retrosternal chest pain and a slight increase in preexisting ST depression in leads II, III, aVr and V4-6. An exercise thallium scan showed no reversible perfusion defects. No exercise-induced regional wall motion abnormality could be detected with conventional dobutamine stress echocardiography (dobutamine concentrations from 10–40 μg/kg/min and 0.5 mg atropine were given additionally for inadequate heart rate response). In addition, atrial pacing stress with determination of coronary sinus (CS)-femoral artery (FA) lactate difference was performed (Figure 5). Incremental increase of the atrial pacing rate by 20 bpm starting at 100 bpm and discontinuation after three minutes on 160 bpm due to palpitations revealed neither a pathological decreased cardiac lactate extraction ratio ((FA_lactate-CS_lactate)/FA_lactate x 100 < 10%) nor a cardiac lactate production (CS_lactate > FA_lactate).

A twenty-four hour ambulatory electrocardiographic recording did not show any malignant arrhythmias or late depolarisations.

The patient was given 120 mg diltiazem twice daily to improve diastolic function and was discharged in an apparently good clinical condition.

On an empirical basis, restriction of excessive exertional activity was recommended and the patient remained asymptomatic. At 6-month follow-up, the patient remained asymptomatic. However, giant negative T-waves persisted without evidence of any developmental progression.

**DISCUSSION**

AHC is a variant of hypertrophic cardiomyopathy (HCM) with predominant involvement of the apex. The typical or "Japanese" type is characterized by giant negative T-waves and a spade deformity of the left ventricle. In contrast to HCM in general and AHC in non-Asians, AHC of the "Japanese" type is
distinctly uncommon in other parts of the world and has been found to be clinically benign.\textsuperscript{3)}

In this case report, we described a male Italian who exhibited the typical features of a “Japanese” type AHC including a characteristic spade-like configuration of the left ventricle during end-diastole, giant negative T-waves on the electrocardiogram and the absence of an intraventricular pressure gradient.

Myocardial ischemia in patients with HCM and non-atherosclerotic coronaries is common and of multifactorial etiology, including reduced coronary flow reserve probably related to thickened and narrowed intramural coronary arteries, increased oxygen demand due to greatly increased muscle mass, and subendocardial ischemia resulting from elevated filling pressures.\textsuperscript{4)} Angina pectoris can usually be precipitated by atrial pacing, often in association with evidence of inadequate coronary flow reserve, elevated filling pressures, and abnormalities in lactate metabolism.\textsuperscript{15-19)} Other causes include coronary vasoconstriction\textsuperscript{20)} and congenital abnormalities of the coronary vasculature. Bilateral coronary-pulmonary artery fistulae,\textsuperscript{5,6)} and single\textsuperscript{7)} or multiple\textsuperscript{8,9)} coronary-left ventricular fistulae have been demonstrated in patients with AHC and proposed to cause myocardial ischemia by coronary steal mechanism.

Muscular bridging is an incidental finding at angiography with a reported frequency of 0.5–7.5\%,\textsuperscript{21)} the left anterior descending artery being affected in almost all cases. Systolic compression has been reported to range from 18–87\%; the length varied from 3 to 33 mm and could not be related with the severity of obstruction.\textsuperscript{11)} Although the lesion is regarded as benign, reports have linked myocardial bridging with myocardial ischemia, infarction and sudden cardiac death.\textsuperscript{12,13)}

In our patient, a variety of exercise tests including thallium scan, treadmill ergometry and atrial pacing with measurement of transcardiac lactate failed to demonstrate myocardial ischemia. Although the patient’s complaints occurred during heavy exercise rather than at rest, coronary vasospasm cannot be excluded completely as a relevant pathomechanism. However, tests for coronary vasospasm were not performed as no consequences other than already ongoing treatment with diltiazem for treatment of AHC would arise from a positive ergonovine test.

In general, coronary artery bypass grafting, excision of the overlying muscle, percutaneous coronary angioplasty and coronary stenting represent therapeutic options for the management of myocardial ischemia caused by “muscular bridging”.\textsuperscript{22-24)}

Theoretically, concomitant occurrence of hypertrophic cardiomyopathy and muscular bridging might be more common in a country like Japan than the sparse reports might suggest, and the benign character of typical AHC does not
justify routine catheterization in every case. One previous report described a case with coexistence of hypertrophic subaortic stenosis and a muscle bridge of a septal perforator,25 while another report describes the coexistence of AHC and a muscular bridge.10

Although both clinical entities may cause myocardial ischemia on their own and by a variety of different pathomechanisms, in this individual case extensive exercise testing did not reveal any electrocardiographic, echocardiographic, metabolic or scintigraphic evidence of myocardial ischemia, thus emphasizing the benign nature of this entity.

REFERENCES

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