Apical Hypertrophic Cardiomyopathy Combined with Bilateral Renal Artery Stenosis in Leriche Syndrome: A Rare Coexistence

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ABSTRACT

We describe the case of a 60-year-old male hypertensive patient who was admitted to our department with intermittent claudication. An echocardiography evaluation detected apical hypertrophy without an intracavity pressure gradient. Transthoracic echocardiography has been the first-line imaging method for patients with suspected hypertrophic cardiomyopathy (HCM), but the method's shortcomings in evaluating the apex are well known. Thus, images from the patient's magnetic resonance imaging and angiography examinations confirmed the classic features of apical HCM. In addition, a 3-dimensional computed tomography evaluation disclosed Leriche syndrome concurrent with severe bilateral stenosis of the renal arteries. Apical HCM combined with severe renal artery stenosis is very rare and has not previously been reported with Leriche syndrome.

INTRODUCTION

Apical hypertrophic cardiomyopathy (HCM) is characterized by hypertrophy of the myocardium, predominantly in the left ventricular apex. This relatively rare variant of HCM constituted 13% to 25% of all HCM cases reported in Japan; however, HCM is distinctly uncommon in other parts of the world and probably constitutes only 1% to 2% of HCM cases in non-Japanese populations [Ates 2006; Ting 2007].

The term Leriche syndrome has commonly been used to describe a variety of clinical symptoms attributable to obstruction of the infrarenal aorta. Although the occlusion site is infrarenal in Leriche syndrome, atherosclerotic renal artery stenosis may coexist in some cases [Jian 2004; Shiraiishi 2005]. Renal artery stenosis is common in patients with known coronary and peripheral artery disease and has been recognized as a cause of chronic renal failure and secondary hypertension [Jian 2004; Nakamura 2007].

We present the first reported case, to our knowledge, of a hypertensive patient complaining of claudication with a diagnosis of apical HCM and concurrent Leriche syndrome with severe renal artery stenosis.

CASE REPORT

A 60-year-old man complaining of bilateral lower-extremity fatigue with exercise (walking distance of 100 m before stopping) was referred to our hospital. He also complained of calf and thigh claudication. The patient's medical history included hypertension that was not properly controlled with an angiotensin receptor blocker. A physical examination revealed a blood pressure of 150/95 mm Hg and a regular pulse rate of 69 beats/minute. A cardiovascular examination revealed the fourth heart sound in the apical area, but no murmur was heard. The examination also was remarkable for markedly diminished bilateral femoral and lower-extremity pulses. The ankle brachial index was 0.57 bilaterally. Laboratory findings on admission showed mildly elevated blood urea nitrogen (60 mg/dL; reference interval, 25-50 mg/dL) and a normal creatinine level (1.05 mg/dL; reference interval, 0.7-1.2 mg/dL). Other laboratory findings were within normal limits.

The electrocardiogram demonstrated sinus rhythm, high voltage in precordial derivations with marked T-wave inversion in leads V4, V5, V6, I, and aVL. An echocardiography examination detected apical hypertrophy without an intracavity pressure gradient. The valves and the ejection fraction were normal. The ankle brachial index was 0.57 bilaterally. Laboratory findings on admission showed mildly elevated blood urea nitrogen (60 mg/dL; reference interval, 25-50 mg/dL) and a normal creatinine level (1.05 mg/dL; reference interval, 0.7-1.2 mg/dL). Other laboratory findings were within normal limits.

The electrocardiogram demonstrated sinus rhythm, high voltage in precordial derivations with marked T-wave inversion in leads V4, V5, V6, I, and aVL. An echocardiography examination detected apical hypertrophy without an intracavity pressure gradient. The valves and the ejection fraction were normal. An apical 4-chamber view of the left ventricle revealed hypertrophy of the apex in the “ace-of-spades” configuration characteristic of apical HCM. The patient underwent a cardiac magnetic resonance imaging evaluation, which demonstrated a marked concentric thickening of the left ventricular apex and distal septum with an isointense signal in the remaining myocardium that gradually decreased to normal levels at the base (Figure 1). Given the magnetic resonance imaging and echocardiographic findings, we diagnosed apical HCM in the patient.

Coronary angiography and left ventriculography evaluations were performed from the right brachial artery. The left ventriculogram revealed a spade-shaped configuration of the left ventricular cavity at end-diastole, which is typical for apical HCM (Figure 2). The coronary arteries were free of...
obstructive disease (Figure 3). No intraventricular or subaortic pressure gradient was present.

One week later, the patient underwent a 16-row multidetector computed tomography angiography examination, and 3-dimensional volume-rendered reformations were obtained. The 3-dimensional volume-rendering anterior-, anterolateral-, and lateral-view images showed both complete abdominal aorta occlusion just below the renal level and concomitant occlusive disease affecting the renal arteries. There was also total ostial occlusion of the left renal artery and an ostial lesion with 95% stenosis of the right renal artery. Atrophy of the left kidney was also found (Figure 4). In addition, a prominent collateral pathway between the superior epigastric artery originating from the internal thoracic artery as the medial terminal branch and an inferior epigastric artery arising from the external iliac artery were seen bilaterally. There was significant enlargement of the right and left internal thoracic arteries and epigastric arteries to supply circulation to the lower limbs (Figures 5 and 6).

The patient’s clinical condition was complex and included apical HCM, significant bilateral renal artery stenosis, and total occlusion of the infrarenal aorta. The patient was offered surgical revascularization, which was the consensus of the cardiologists and cardiovascular surgeons, but he has refused any surgical intervention. The patient has shown improvement while remaining on medical treatment and is being followed up for vascular surgery. Although bilateral renal artery stenosis was present, follow-up has shown renal functions to be within normal limits.

**DISCUSSION**

We have presented a case of abdominal aortic occlusion (ie, Leriche syndrome) along with bilateral severe renal artery stenosis causing secondary hypertension in a patient with...
apical HCM. To the best of our knowledge, the coexistence of apical HCM, Leriche syndrome, and bilateral severe renal artery stenosis in the same patient has not previously been reported.

Patients with apical HCM tend to have few or no symptoms. Apical HCM generally has a more benign course than other variants of HCM [Ting 2007]. Despite a relatively favorable prognosis for apical HCM, sudden cardiac death, life-threatening ventricular arrhythmias, and apical aneurysm have occasionally been reported [Okishige 2001]. The mechanisms of these complications, although not well defined, have been proposed to be due to small-vessel disease caused by increased collagen deposition and to myocardial ischemia caused by a deficient myocardial oxygen supply and demand mismatch. This mismatch is driven primarily by the increased myocardial mass [Nakahashi 1995; Ting 2007].

Eriksson et al [2002] reviewed a series of 105 patients with apical HCM with a mean follow-up of 13.6 ± 8.3 years from presentation. The overall cardiovascular mortality rate was 1.9% (2/105), and the annual cardiovascular mortality rate was 0.1%. The overall survival rate was 95% at 15 years, and the probability of survival without morbidity was 74% at 15 years.

The diagnosis of apical HCM can be established with transthoracic echocardiography. If the apex is difficult to visualize on transthoracic echocardiography or if echocardiographic images are difficult to obtain or interpret, magnetic resonance imaging may occasionally be used to diagnose apical HCM [Ibrahim 2000; Ates 2006].

Apical HCM in patients with Takayasu arteritis has previously been reported [Satish 2002], but the coexistence of apical HCM, Leriche syndrome, and bilateral renal artery stenosis in the same patient has heretofore been unreported.

Normally, the superior and inferior epigastric arteries anastomose with each other if the abdominal aorta develops a significant stenosis and/or blockage (as may be caused by atherosclerosis or coarctation). This collateral pathway bypassing the obliterated segment may develop sufficiently over time to supply blood to the lower limbs [Yurdakul 2006]. In patients with occlusive aortoiliac disease, blood flow to the lower extremities can depend to a greater or lesser extent on the collateral circulation originating in the internal thoracic artery [Ferrer 2007].

Infrarenal abdominal aortic occlusive disease has traditionally been treated with aortic endarterectomy and/or aortobifemoral bypass grafting [Rutherford 1994]. With the advent of endovascular surgery, percutaneous transluminal angioplasty and stent placement have increasingly been used as an alternative to conventional surgery in the management of patients with focal aortoiliac disease [Karkos 2000]. Treatment with streptokinase [Bean 1985] and angioplasty [Bean 1985; Karkos 2000; Ferrer 2007] has been used in selected patients with Leriche syndrome.

For patients with renal artery stenosis and multisystem atherosclerotic disease, surgical revascularization or transaortic endarterectomy [Hagino 1997] is a great clinical challenge.
Less invasive endovascular procedures, such as renal angioplasty and stenting, seem to be the best way to stabilize renal function in selected patients or those with considerable morbidity [Jian 2004; Shiraishi 2005]. Supraceliac aortorenal bypass grafting is a safe and durable alternative to renal artery revascularization in carefully selected patients [Hagino 1997].

The experience with angioplasty and stenting for total aortic occlusions appears to be limited [Hagino 1997]. Thus, because of our patient’s symptomatic aortoiliac occlusive disease complicated by bilateral renal artery stenosis, we thought adequate treatment for this patient would consist of surgical revascularization or transaortic endarterectomy for the right renal artery and complete infrarenal aortic endarterectomy and/or aortobifemoral bypass grafting. The patient is being managed conservatively because of his refusal of surgical intervention.

The coexistence of different diseases in the same patient is very interesting and appears to be rare.

REFERENCES


