Simultaneous Pressure Recording in Mid-Ventricular Obstructive Hypertrophic Cardiomyopathy

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Abstract

A 70-year-old man was diagnosed with mid-ventricular obstructive hypertrophic cardiomyopathy (MVOHCM) with apical aneurysm and paradoxic jet flow. At cardiac catheterization, pressure study showed that there was a markedly high pressure-gradient of 90 mmHg between the apex and the base in systole. Apical pressure was 350 mmHg after premature ventricular contraction. The apical aneurysm was already dilated and spherical in late systole; the absence of active relaxation was considered to be the cause of the paradoxic jet flow. In this report, we suggest the pathogenesis of left ventricular apical aneurysm and paradoxic jet flow in MVOHCM.

Key words: mid-ventricular obstructive hypertrophic cardiomyopathy, apical aneurysm, paradoxic jet flow


Introduction

Mid-ventricular obstructive hypertrophic cardiomyopathy (MVOHCM) with apical aneurysm is a rare condition that occurs in 2% of hypertrophic cardiomyopathy (1). It is frequently associated with unusual intraventricular isovolumic diastolic flow, which is termed “paradoxic jet flow” (2). In this case report, we simultaneously measured pressures at the left ventricular apex and at the base, and speculate on the pathophysiology of apical aneurysms and paradoxic jet flow in MVOHCM.

Case Report

A 70-year-old man was referred to our hospital because of shortness of breath. Although the patient had been made aware of abnormal findings in his electrocardiogram, he had not undergone further examination. Physical examination showed that his blood pressure was 166/94 mmHg, and that his pulse rate was 87 beat/min. Auscultation of the heart and respiratory sounds were normal. Blood sampling data indicated dyslipidemia (total cholesterol: 263 mg/dL, triglyceride: 387 mg/dL) with a normal plasma brain natriuretic peptide level (65.5 pg/mL). His chest radiograph was normal (cardio-thoracic ratio of 51%). A 12-lead electrocardiogram showed a high R wave and a negative T wave in precordial leads. Two-dimensional transthoracic echocardiography showed hypertrophy and intraluminal obstruction at the mid-ventricle. Continuous-wave Doppler echocardiography showed a flow pattern specific for MVOHCM: high flow from the apex toward the base in early systole, the disruption of flow in mid-systole, and resumption of high flow from the apex toward the base in an isovolumic relaxation termed a paradoxic jet flow (Fig. 1). Cavity obliteration time, corrected by RR interval (cCOT) defined by Matsubara et al (3), was 298 ms by continuous wave Doppler echocardiography, and 293 ms by color M-mode echocardiography (Fig. 2). A computed tomographic scan showed marked left ventricular hypertrophy with systolic obstruction at the mid-ventricle and an apical aneurysm. Contrary to the base, the apical aneurysm shrunk during diastole (Fig. 3).
cardiac catheterization, a pressure study determined markedly high systolic pressure of 220 mmHg at the apex and a pressure gradient of 90 mmHg between the apex and the base. The pressure gradient became more severe after premature ventricular contraction (PVC), and apical pressure increased to 350 mmHg. In early diastole, isovolumic relaxation was delayed at the apex and there was a significant pressure gradient between the apex and the base (Fig. 4).

We diagnosed this patient as having MVOHCM and treated him with calcium channel blocker and beta-blocker medication.
Figure 4. Pressure study shows markedly high systolic pressure of 220 mmHg at the apex (left panel) and a pressure gradient of 90 mmHg between the apex and the base (middle panel). The pressure gradient became more severe after premature ventricular contraction and apical pressure increased to 350 mmHg (right panel). In early diastole, isovolumic relaxation was delayed at the apex and there was a significant pressure gradient between the apex and the base (middle panel, arrow).

Discussion

Apical aneurysm in MVOHCM

MVOHCM is often accompanied by an apical aneurysm that may be a risk of lethal arrhythmia, thromboembolic stroke, and sudden death (1). The apical aneurysm may be difficult to visualize by echocardiography due to massive hypertrophy, but can be clearly demonstrated by computed tomographic scanning, as it was in the present case.

Although the pathogenesis of an apical aneurysm is unknown, high left ventricular apical pressure due to mid-ventricular obstruction may be the main cause of an apical aneurysm (4, 5). Moreover, progressive left ventricular dilation and shape change (more spherical) can lead to the upregulation of stretch response protein that may lead to a positive feedback loop of dilatation (6).

In the present case, simultaneous pressure recordings showed that the pressure gradient between the apex and the base was close to 90 mmHg. After PVC, apical pressure increased to 350 mmHg due to extrasystolic potentiation. This suggests that PVC and other arrhythmia may cause further apical expansion and apical rupture in this disorder.

Assessment of the severity of obstruction by echocardiography

Matsubara et al reported that sustained cavity obstruction was an important pathophysiologic condition of apical aneurysm and that cCOT measured by M-mode echocardiography correlated with hypertrophy, ischemia, ventricular tachycardia, and development of apical aneurysm (3). In the present case, cCOT was not measured by M-mode due to the poor image. Instead, cCOT was found to be 298 ms by continuous wave Doppler echocardiography and 293 ms by color M-mode echocardiography. According to their criteria, the severity of cavity obstruction was moderate in this patient (from 200 ms to 350 ms).

Mechanism of paradoxic jet flow

The paradoxic jet flow is a mysterious mid-ventricular flow directed from the apex toward the base in early diastole. Its pathophysiology has been rarely discussed and it remains unexplained. We recently reported intraventricular pressure during paradoxic jet flow, where we believed that delayed apical relaxation during isovolumic relaxation might contribute to its generation (7).

In the present case, we recorded apical and basal pressures simultaneously and confirmed delayed apical relaxation. As the delay was considerable and the pressure gradient continued to mid-diastole (Fig. 3), this delay was not the simple time lag, but the consequence of real relaxation abnormality.

During the initial phase of isovolumic relaxation, left ventricular shape may change to a spherical shape in the normal heart (8). We have also encountered distinct sphericalization...
during isovolumic relaxation in cardiac tamponade (9). However, in this case, the apex was already dilated and spherical in end-systole and active relaxation could not occur. Apical pressure decreased gradually with the shrinking of the apex by paradoxic jet flow. Thus, relaxation at the apical aneurysm might be passive and might conform to the intrinsic pressure volume curve.

This case is considered clinically significant in developing the understanding of the pathogenesis of left ventricular apical aneurysms and paradoxic jet flow in MVOHCM.

The authors state that they have no Conflict of Interest (COI).

References


