Pseudoidiopathic Hypertrophic Subaortic Stenosis in a Patient with Cardiac Tamponade

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A 53-year-old woman with a large pericardial effusion and tamponade presented with signs of IHSS including a grade 4/6 apical systolic murmur, severe SAM, early systolic aortic valve closure and a small hypercontractile left ventricle but at most borderline left ventricular hypertrophy. Following pericardiotension, the clinical and echocardiographic signs of subvalvular obstruction resolved completely. One year later the patient died of bronchial carcinoma and no evidence of hypertrophic cardiomyopathy was found at autopsy. Pericardial tamponade should be added to the list of possible causes of dynamic subvalvular obstruction in a structurally normal heart.

Echocardiography is now the standard method for diagnosing idiopathic hypertrophic subaortic stenosis (IHSS). The three most important echocardiographic signs of this disease are: 1) asymmetric septal hypertrophy (ASH), 2) systolic anterior motion of the mitral echoes (SAM), and 3) mid-systolic closure of the aortic valve. While ASH is a sign of hypertrophic cardiomyopathy, SAM and premature aortic closure indicate dynamic subvalvular obstruction. However, none of these signs is totally specific for IHSS and in the literature there are occasional case reports on patients with dynamic subvalvular obstruction in the absence of hypertrophic cardiomyopathy. In this article, we report, for the first time, a patient in whom a large pericardial effusion with tamponade led to the appearance of clinical and echocardiographic signs of IHSS, which resolved completely after pericardiocentesis.

CASE REPORT

The patient was a 53-year-old woman who had been essentially healthy previously except for intermittent claudication of the left leg which was relieved by balloon dilatation of the femoral artery in 1979. On May 14, 1984, she was admitted to another hospital with progressive dyspnea, chest discomfort and fever (38.5°C). At that time, tachycardia, cardiomegaly and mild hepatomegaly were noted, but there was no systolic murmur. She was treated with antibiotics, diuretics and diuretics, yet deteriorated progressively. Three days later, a new grade 4/6 systolic murmur was noted at the apex and left parasternal area. She also developed bilateral pleural effusion, mural edema and marked hepatomegaly (4 cm below the costal margin). Bacterial endocarditis was suspected and the patient was transferred to our hospital. On arrival, she was sent to the echocardiography laboratory.

The echocardiogram (Fig 1) revealed no evidence of bacterial endocarditis. Instead, there was a large pericardial effusion with right ventricular compression as indirect evidence of cardiac tamponade. The left ventricle was small and hyperactive with an end-diastolic diameter (LVEDD) of 3.5 cm, an end-systolic diameter (LVEDSD) of 1.5 cm (fractional shortening =57 percent). The left ventricular outflow tract (LVOT) at the onset of systole was 1.6 cm, the septum 1.2 cm and posterior wall 1.2 cm. In addition, there was severe SAM of the mitral valve leaflets with broad contact to the interventricular septum. Using the method of Pollick et al., a subaortic gradient of 75 mm Hg was calculated. The aortic valve showed obvious mid-systolic closure with systolic fluttering. On the basis of these echocardiographic and clinical findings, the diagnosis of IHSS was made though there was no ASH and the left ventricular hypertrophy was unusually mild.

Since the patient was in very poor condition with tachycardia, pulsus paradoxus, low blood pressure, severe dyspnea and distended neck veins, pericardiotension was performed immediately and a pig tail catheter was introduced into the pericardial sac. After removal of 940 ml of clear fluid with high protein content, the systolic murmur became much softer. Serial follow up echocardiograms showed diminution and later disappearance of the echocardiographic signs of subvalvular obstruction with a simultaneous increase in the LVEDD, LVEDSD and LVOT (Fig 2 and 3).

On the sixth day, the echocardiogram was normal except for some residual pericardial effusion and upper limit of normal left ventricular wall thickness (LVEDD = 3.8 cm, LVEDSD = 2.2 cm, fractional shortening = 42 percent, posterior wall = 1.1 cm, ventricular septum = 1.1 cm). The phonocardiogram and pulse curve recorded at that time were normal at rest and during provocation with nitroglycerin. The patient progressively improved with supportive therapy and was discharged on the tenth day after complete recovery.

In July 1985 the patient was readmitted because of bronchial carcinoma and died shortly after admission. At autopsy the heart did not show any evidence of hypertrophic CMP.

DISCUSSION

In this report, we describe a patient who developed prominent SAM and early closure of the aortic valve and a new and varying systolic murmur during the course of pericarditis with effusion and tamponade. These signs resolved completely after treatment of the pericardial disease.

REFERENCES


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Severe SAM with complete systolic mitral septal contact and early systolic closure of the aortic valve are usually seen in patients with hypertrophic cardiomyopathy.\textsuperscript{3,4,8} Although the exact mechanism of the subaortic pressure gradient in IHSS is controversial, there is substantial evidence to incriminate subaortic obstruction due to SAM with mitral septal contact in the setting of a hypercontractile left ventricle and a narrow outflow tract. It is now well appreciated that diseases other than IHSS may also go along with dynamic subvalvular obstruction and corresponding clinical, hemodynamic and echocardiographic findings which may be indistinguishable from IHSS.

The long list of differential diagnoses can be grouped into two categories. First, patients in whom the narrow LVOT and subvalvular obstruction is caused by an anatomically thick septum, eg, the valvular aortic or discrete subaortic stenosis,\textsuperscript{9} systemic hypertension,\textsuperscript{4,9} children of diabetic mothers,\textsuperscript{7} glycogen storage disease,\textsuperscript{7} some forms of infiltrative cardiomyopathy.\textsuperscript{9} Second, patients with a small but anatomical...
ically normal nonhypertrophic left ventricle with hypercontractile function, e.g., hemorrhagic shock, pressor therapy, and idiopathically hyperkinetic hearts.

The patient described here also had a structurally normal heart. The observation that she developed dynamic subvalvular obstruction during the course of cardiac tamponade can be explained, as follows.

Pericardial tamponade causes compression of the heart with reduction in effective transmural filling pressure. This leads to a decrease in left ventricular size and outflow tract width. Systolic contraction was supranormal because of excessive sympathetic drive during cardiac tamponade. Thus, the conditions for the development of the subvalvular obstruction in a normal heart (small hyperactive left ventricle) were fulfilled. The treatment with diuretics and digitalis may have aggravated the obstruction by further decreasing effective filling pressure and further increasing contractility. These signs resolved completely after pericardiocentesis and withdrawal of therapy with digitalis and diuretics. At the same time, the LV size and outflow tract width increased, while fractional shortening decreased.

To our knowledge, this is the first report of obvious subaortic obstruction in cardiac tamponade. There has been one report from 1973 where SAM was described in a patient with pericardial effusion. However, SAM in this case was very mild and did not even get close to the septum and did not fulfill currently used criteria for outflow obstruction.

In conclusion, pericardial tamponade therefore should be added to the list of conditions which may cause dynamic subvalvular obstruction with an anatomically normal left ventricle.

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