CASE REPORT

A Rare Case of Hypertrophic Cardiomyopathy Associated with Congenital Mitral Stenosis

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ABSTRACT

Hypertrophic obstructive cardiomyopathy is mostly associated with mitral insufficiency rather than mitral stenosis. This association is very rare and no cases have been reported in Africa. Our case was about 22-month-old female child that was referred with a 1-year history of tachypnea and III to IV class of dyspnea. Transthoracic echocardiography showed serious mitral stenosis and a mean gradient of 27 mmHg. The interventricular septum was hypertrophic with a width of 8.5 mm with small aortic annulus, leading subaortic stenosis with a mean gradient of 73 mmHg. There was also a severe pulmonary hypertension at 79 mmHg. It was expected to doing a standard septal myectomy and mitral valve replacement.

Key words: Hypertrophic cardiomyopathy, mitral plasty, mitral stenosis, septal myectomy

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is defined as asymmetric (predominantly septal) left ventricular (LV) hypertrophy, of genetic origin, inconsistently associated with ejection obstruction (obstructive HCM or OHCM).

It is a common cause of sudden death in young patients that may be the first manifestation.[1] It can classically associate with mitral insufficiency, but its association with a mitral stenosis is very rare and the prognosis is reserved.[2]

CASE REPORT

A 22-month-old child from an apparently healthy couple was hospitalized at our center for a congestive heart failure syndrome. The clinical evaluation noted Stage IV dyspnea, regular tachycardia at 150 beats/min, quivering, and 4/6 aortic and mitral systolic murmur with preserved B2, accentuated pulmonary component of the second heart sound. SPO_2 was 100%, with polypnea at 30 cycles/min and crackles at both lungs. Painful hepatomegaly to two fingers was noted.

The electrocardiogram recorded sinus tachycardia at 153 beats/min, the right ventricular hypertrophy, axis 130° [Figure 1]. The chest X-ray showed cardiomegaly with a cardiothoracic index: 0.63, a supradiaphragmatic tip, an accentuation, and vascular redistribution to the apices [Figure 2].

Doppler echocardiography showed severe congenital mitral stenosis (mean gradient: 27 mmHg) by abutment-commissure fusion [Image 1]. No mitral leak associated.

There was no hypoplasia of the ring or supramitral membrane. There was concentric ventricular hypertrophy of the ventricle (septal thickness: 10 mm and posterior wall thickness: 11 mm) left responsible for a reduction of the ventricular cavity [Image 2] and systolic anterior motion (SAM) severe aortic stenosis (mean gradient: 73 mmHg). The aortic ring was hypoplastic 7 mm [Image 3]. There was also hypertrophy of the right ventricle and moderate tricuspid regurgitation estimating pulmonary pressure at 79 mmHg.

While waiting for surgery, a medical treatment with propranolol was initiated.

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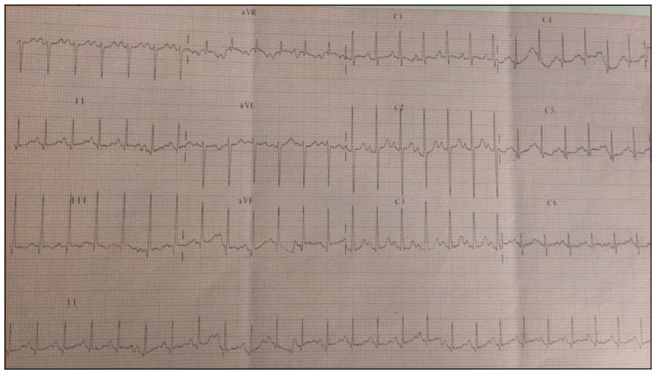

Figure 1: Electrocardiogram showing the right ventricular hypertrophy

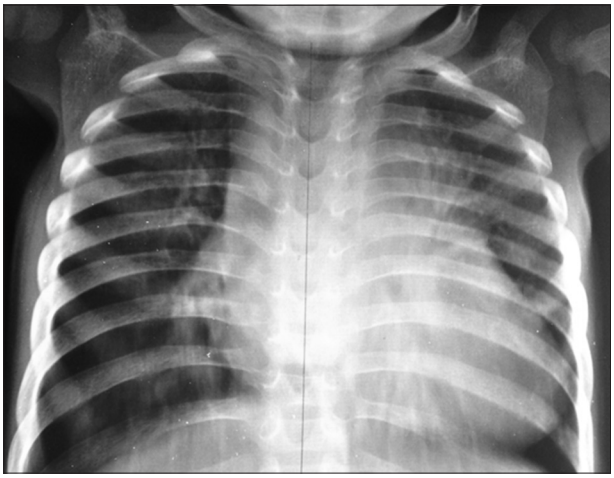

Figure 2: Chest X-ray showing cardiomegaly at the expense of the right cavities and vascular redistribution to the apices

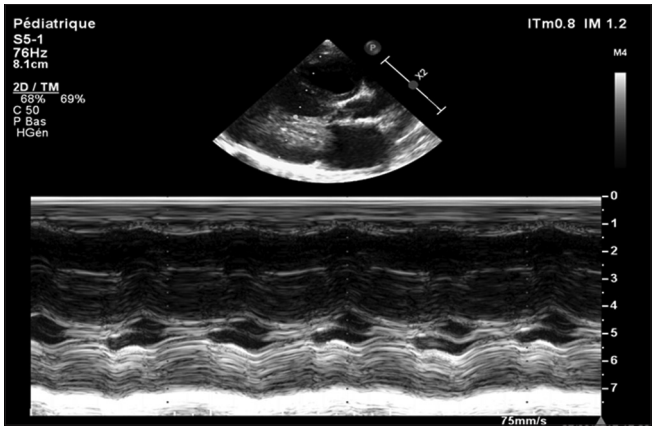

Image 1: M-mode parasternal long axis view revealing the left ventricular hypertrophy

While waiting for a possible transfer abroad for surgery, the patient died of sepsis in a context of gastroenteritis.

DISCUSSION

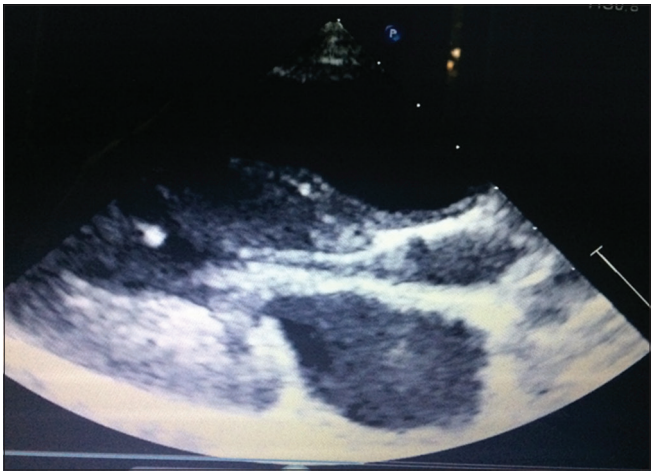

Image 2: Concentric left ventricular hypertrophy and small aortic ring

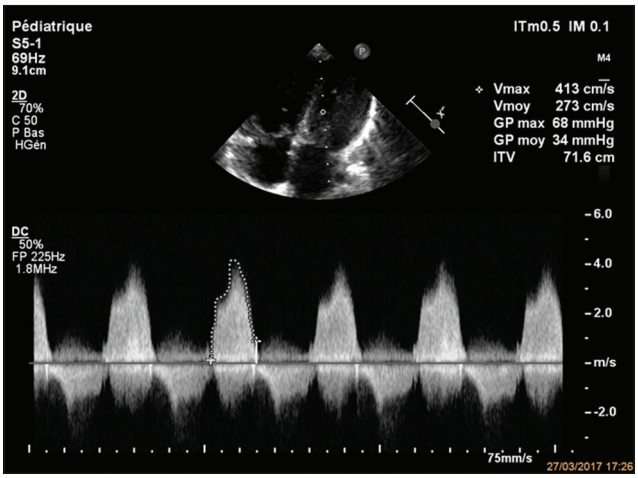

Image 3: Continuous wave Doppler on the mitral valve evaluating the elevation of the mitral gradient

The unusual presentation of our case is interesting due to its rarity and complexity of diagnosis and management. It is classic to find mitral regurgitation in HCM. There is a SAM that creates a mitral leak and participates in aortic obstruction.[2] On the other hand, it is rare that HCM is associated, as in our case, with mitral congenital valve stenosis.

Clinically, the patient may remain asymptomatic or has non-specific clinical signs. Sudden death may be the first manifestation.[1] We had observed a congestive heart failure syndrome with an episode of acute decompensation. This clinical presentation and pulmonary edema observed in our case are attributable to the sum of the physiopathological effects of OHCM and mitral stenosis. OHCM increased the LV end-diastolic pressure with anterograde airway obstruction resulting in upstream pressure elevation, while mitral stenosis caused dyspnea due to the LV filling

obstruction, thereby determining the high pressure of the left atrium and pulmonary capillaries.

Transthoracic echocardiography revealed concentric hypertrophy of the left ventricle responsible for ventricular cavity reduction and SAM tight aortic stenosis (mean gradient: 73 mmHg). We also noticed a small aortic ring. There was a high transmitral gradient created by congenital mitral stenosis (mean gradient: 27 mmHg). The mechanism of mitral stenosis was particular and complex. If SAM was at least partly responsible for mitral stenosis, there was also a commissural fusion. There was no associated mitral regurgitation despite the presence of SAM. We did not report hypoplasia of the ring or supramitral membrane. Mitral stenosis had determined a severe pulmonary hypertension estimated by tricuspid regurgitation at 79 mmHg. The right ventricle was therefore hypertrophied. There were no other associated malformations such as aortic coarctation that could define the Shone complex.[3,4]

Even though we know that there are incomplete forms, the lesions described above did not seem to support any form of Shone complex.

Medical treatment with beta-blockers or disopyramide is often beneficial in MHC.[5]

Thus, our patient was on propranolol at the initial dose of 0.5 mg/kg/day. The dose was gradually increased to 3 mg/kg/day. The child was also treated with Lasilix 2 mg/kg/day and spironolactone 2 mg/kg/day due to the congestive heart failure. The evolution under this drug treatment was marked by the improvement of the congestion. It is usual to offer an invasive treatment in patients who are refractory to medical treatment. Thus, septal alcoholic ablation may result in a significant reduction in obstruction and septal thickness.[6] This method is often proposed in elderly patients at high surgical risk. We had not considered carrying it out in our patient mainly due to the existence of an associated mitral obstruction. We also did not consider percutaneous mitral dilatation since the mechanism of the stenosis was not a commissural fusion. Surgery has been shown to have the best results in the long term with low morbidity and mortality.[7] In most cases, this is a conventional myomectomy.[8] In some cases, this intervention is sufficient.

However, the myomectomy may be insufficient and it is conventional to perform a mitral valve plasty or mitral valve replacement to remove the mitral obstacle.[8] In our patient, we planned to perform a myomectomy associated with a mitral plasty. However, waiting for a possible transfer abroad for surgery, the patient died of sepsis in a context of gastroenteritis.

CONCLUSION

HCM can exceptionally be associated with congenital mitral stenosis as was the case in our patient with a rather atypical clinical presentation. Diagnosis should be made by Doppler echocardiography and treatment discussed collegially with cardiac surgeons. Surgery should consist of myomectomy and mitral plasty for those patients with HCM associated with mitral stenosis.

REFERENCES

1. Maron BJ, Olivotto I, Spirito P, Casey SA, Bellone P, Gohman TE, *et al.* Epidemiology of hypertrophic cardiomyopathy-related death: Revisited in a large non-referral-based patient population. Circulation 2000;102:858-64.
2. Cavalcante JL, Barboza JS, Lever HM. Diversity of mitral valve abnormalities in obstructive hypertrophic cardiomyopathy. Prog Cardiovasc Dis 2012;54:517-22.
3. Shone JD, Sellers RD, Anderson RC, Adams P Jr., Lillehei CW, Edwards JE, *et al.* The developmental complex of "parachute mitral valve" supravalvular ring of left atrium, subaortic stenosis, and coarctation of aorta. Am J Cardiol 1963;11:714-25.
4. Bolling SF, Iannettoni MD, Dick M 2nd, Rosenthal A, Bove EL. Shone's anomaly: Operative results and late outcome. Ann Thorac Surg 1990;49:887-93.
5. Nishimura RA, Holmes DR. Hypertrophic obstructive cardiomyopathy. N Engl J Med 2004;350:1320-7.
6. Yavuzgil O, Tülüce K, Simsek E, Gölcük E, Özcan KS, Guler TE. Septal alcohol ablation in hypertrophic obstructive cardiomyopathy: a retrospective examination. Turk Gogus Heart Dama 2012; 20:427-33.
7. Unal EU, Ozen A, Işcan HZ, first son CL. Early and mid-term results of surgical treatment in obstructive hypertrophic cardiomyopathy. Turk Gogus Heart Dama 2012; 20:421-6.
8. Mataracı I, Polat A, Songur çm, Aydin C, Yanartas M, Seidman CE. Surgical treatment and results of hypertrophic obstructive cardiomyopathy. Turk Gogus Heart Checkers 2009; 17:243-8.

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