CASE REPORT

A Multiparous Woman with Lately Diagnosed Multilevel Left Ventricular Obstruction

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ABSTRACT

A 56-year hypertensive, multiparous woman presented to the cardiology unit with Canadian Cardiovascular Society (CCS) class-III angina and worsening dyspnea for the past few weeks. Her clinical examination showed high blood pressure and mid-systolic crescendo-decrescendo murmur radiating to carotids. However, there was no radio-femoral delay or significant blood pressure difference between her arms. Her transthoracic echocardiography (TTE) revealed moderate aortic stenosis (AS) and mid cavity left ventricular outflow (LVO) obstruction. Left heart catheterization (LHC) showed coarctation of aorta with extensive collaterals, mid cavity LVO obstruction, and moderate AS. Thus, she was diagnosed as a case of multi-level LVO obstruction including mid cavity LVO obstruction AS and coarctation of aorta. She underwent stenting of aortic coarctation as the initial step of graded approach to her disease, and is doing well.


INTRODUCTION

Multilevel LVO obstruction is characterized by combination of two or more obstructive lesions in series to left ventricle. It is usually diagnosed in infancy or early childhood during routine clinical examination supplemented by imaging. However, if the diagnosis is missed, it may lead to serious complications, especially in females of child-bearing age.¹

We are presenting a rare case and probably the first of its kind in the literature, where a lady with multilevel LVO obstruction remained undiagnosed till the fifth decade of her life and multiple pregnancies.

CASE REPORT

A 56-year lady, mother of three children and having uncontrolled hypertension for the last 20 years, presented to the cardiology clinic with CCS-III angina and worsening dyspnea for the past few weeks. Her clinical examination showed high blood pressure and mid-systolic crescendo-decrescendo murmur radiating to carotids. However, there was no radio-femoral delay or significant blood pressure difference between her arms. Her ECG showed left ventricular hypertrophy. Transthoracic echocardiography (TTE) revealed moderate aortic stenosis (AS) and mid cavity LVO obstruction with a peak pressure gradient of about 45 mmHg and moderate AS (aortic stenosis) with mean pressure gradient of about 32 mmHg and peak pressure gradient of about 50 mmHg (Figures 1a and 1b). Keeping in view her CCS-III angina and the unreliability of echocardiographic assessment of AS in the presence of mid cavity LVO obstruction, it was decided to proceed with LHC to properly elucidate the gradients across the aortic valve.

LHC was planned through femoral arterial approach because there was no radio-femoral delay on clinical examination and bicuspid aortic valve was missed on initial TTE. After achieving right femoral arterial access with a 6.0 French (6F) sheath, a 0.035" wire (Cordis®) was introduced through the femoral sheath but could not be negotiated through the descending aorta above the diaphragm. An aortogram was performed with the help of a 6F pigtail catheter (Boston Scientific®), which showed coarctation of aorta (Figure 2a). Another access through right radial artery was achieved and aortogram performed again with the help of a 4F pigtail catheter (Boston Scientific®), which confirmed coarctation of aorta along with very well-developed collaterals across the coarcted segment (Figure 2b). A 6F multipurpose catheters- MPA (Boston Scientific®) and a 6F Judkins right (Boston Scientific®) were taken, one placed above and the other below the coarcted segment of the aorta;
and instantaneous pressure gradient across coarcted segment was taken that measured to be 90 mmHg.

Coronary angiography, performed through right radial approach, showed mild coronary artery plaquing in dominant left coronary circulation and severe disease in non-dominant right coronary artery. Then a 6F MPA catheter (Boston Scientific®) with end hole was introduced into the left ventricular cavity and mid cavity pressure gradient was taken that measured to be 40 mmHg and pressure gradient across the aortic valve was 30 mmHg.

Post LHC, repeat TTE especially for the aortic valve, showed bicuspid aortic valve which was missed on the initial TTE. Thus she was diagnosed as a case of multilevel LVO obstruction with mid cavity LVO obstruction, bicuspid aortic valve with AS, and coarctation of the aorta. She underwent stenting of aortic coarctation, through right femoral arterial approach, with a covered stent-Cheetham Platinum (NUMED®) (8x39 mm), using 4x16 mm balloon (NUMED®) and 260 cm long Amplatz wire (Cook®) (Figures 2c and 2d). On follow-up at 1 year, she was free of angina and symptoms of heart failure; and was doing fine.

We decided not to proceed with balloon aortic valvuloplasty as it was moderate aortic stenosis, both on non-invasive and invasive work-up.

DISCUSSION

LVO obstruction encompasses a spectrum of anomalies, ranging from a single lesion like aortic stenosis to multiple serial obstructions such as combination of bicuspid AS and aortic coarctation. The aortic stenosis may be valvular, subvalvular or supravalvular and the aortic coarctation may be anywhere from the beginning up to the abdominal aorta.1

The prevalence of LVO obstruction among the congenital heart diseases is 7%, of which 70% have aortic valve stenosis, 14% have subvalvular aortic stenosis, 8% have supravalvular aortic stenosis, and about 8% have obstruction at multiple levels.2 The clinical significance of LVO obstruction is the high afterload, against which left ventricle works, leading to left ventricular hypertrophy followed by dilatation and eventual heart failure. The prognosis is even worse when there is multi-level obstructive lesions because of significantly high afterload.3 The situation becomes further complicated when such kind of obstruction occurs in female, because stenotic lesions are poorly tolerated in pregnancy with significantly high morbidity and mortality for both mother and child.4

However, this case is unique as the patient tolerated three pregnancies and home deliveries despite having significant multilevel LVO obstruction and her LVO obstruction remained undiagnosed till she presented to us.

Diagnosis in most of the cases is established in infancy or early childhood through thorough clinical examination.
followed by imaging. Radio-femoral delay is the hallmark of clinical examination, especially in early childhood, in case of aortic coarctation. However, sometimes, this finding may be absent; especially, if there are sufficient collaterals across the coarcted aortic segment. Same was the situation in our case where there was no radio-femoral delay, probably as a result of extensive collaterals across the coarcted segment.

Radoslaw and colleagues reported a case of an adult lady with delayed diagnosis of aortic coarctation who presented with signs and symptoms of heart failure. But that was not a multilevel LVO obstruction. Delayed diagnosis of multilevel LVO obstruction in the form of LV mid cavity obstruction, bicuspid aortic valve with AS and aortic coarctation, in adult multiparous lady, to the best of our knowledge, has not been reported in literature previously.

The traditional diagnostic tool of LVO obstruction is invasive angiogram which enables us to measure pressure gradients across different levels of obstruction. However, non-invasive diagnostic modalities in the form of echocardiography, computed tomographic angiography and, especially, magnetic resonance imaging provide us with valuable information regarding pathology and gradients across the stenotic regions in patients with LVO obstruction.

Treatment is a major challenge in such patients, especially so in case of multilevel LVO obstruction as it is hard to accurately define as to which lesion should be addressed first and at what particular time. We suggest a graded approach dealing with the most significant lesion first and then assessing the response. The main stay of treatment, especially of aortic coarctation, is surgery. However, recent trends are in favour of endovascular treatment with covered or uncovered stents.

REFERENCES