

Case Report

Double Orifice Mitral Valve and Bicuspid Aortic Valve: Pieces of the Same Single Puzzle?

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Double orifice mitral valve is a very rare congenital abnormality. Well known associations of this pathology with other congenital lesions point to a complex and central pathophysiological mechanism leading to a sequence of pathologies. These associations have long been realized and arbitrarily defined as Shone complex. We would like to present a 21-year-old patient with double orifice mitral valve associated with bicuspid aortic valve, with a brief review of the literature on possible central mechanisms leading to different subsets of congenital abnormalities involving these two.

1. Introduction

Double orifice mitral valve is a very rare congenital abnormality [1]. The left atrium opens to the left ventricle via mitral valve orifice divided into two by a fibrous tissue. Isolated occurrence of this abnormality is an even rarer phenomenon. Well known associations of this pathology with other congenital lesions point to a complex and central pathophysiological mechanism leading to a sequence of pathologies. These associations have long been realized and arbitrarily defined as Shone complex. We would like to present a case with cooccurring double orifice mitral valve and bicuspid aortic valve, with the aim of drawing attention to possible underlying mechanisms.

2. Case Report

A 21-year-old man presented to our outpatient clinic with exercise-induced chest pain and palpitations. There was nothing remarkable in medical or family history. Questioning of the functional status revealed NYHA class III dyspnea. Vitals were normal. Levine 3/6 apical diastolic murmur was noted in auscultation. ECG, chest X-ray, and routine blood work yielded normal results.

Transthoracic echocardiography clearly showed the mitral valve with 2 separate orifices. The gradients with

continuous wave (CW) Doppler demonstrated severe mitral stenosis (24 mmHG mean gradient), in apical four-chamber and parasternal short axis (PSSX) view (Figures 1(a)-1(b)). An accompanying bicuspid aortic valve (BAV) with fusion of the left and right coronary cusps with insignificant gradient was observed in basal PSSX view (maximal and mean gradients of 18 and 12 mmHg, resp.). Transesophageal echocardiography (TOE) was performed to clarify mitral and aortic valve anatomy. DOMV was clearly visualized in TOE, and the gradient over the left ventricular outflow tract was localized to the level of the valve (Figures 2(a) and 2(b)). Coarctation of aorta or other associated congenital abnormalities was not detected. The patient was referred for surgery.

3. Discussion

DOMV is a very rare abnormality in which there are two separate orifices with separate respective chordal and papillary structures. This abnormality was first reported in 1876 by Greenfield [2]. Banerjee et al. reported incidence of this rare abnormality to be 0.05% [1]. DOMV is found to be associated with transposition of the great arteries, atresia of the left ventricle outflow tract, single coronary artery [3], ostium primum defect [4, 5], ostium secundum defect, ventricular septal defect and hypoplastic left heart

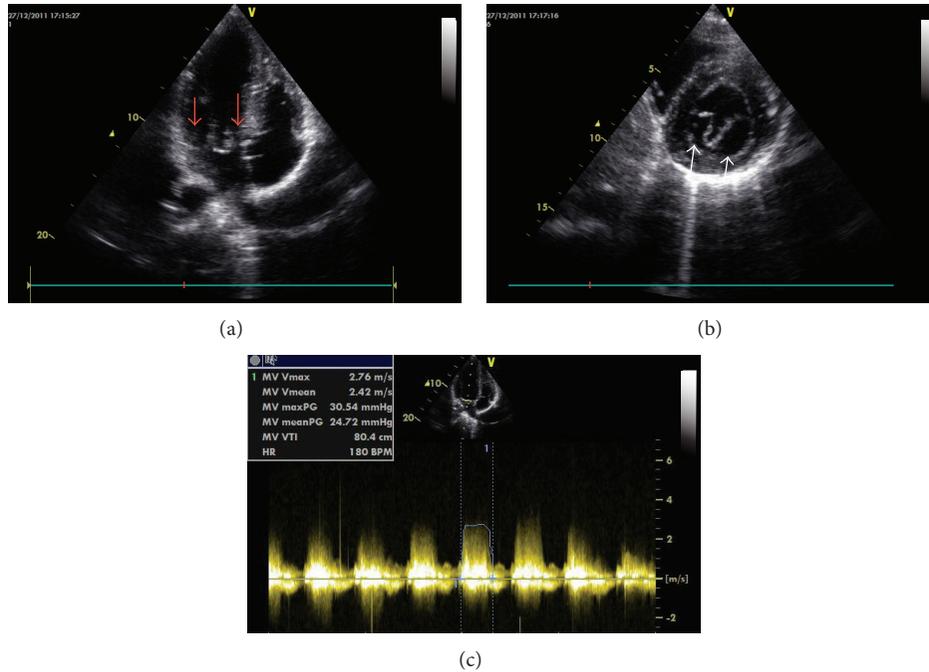


FIGURE 1: (a) Apical four-chamber view and (b) parasternal short axis view showing double orifice mitral valve. (c) Continuous wave Doppler tracings showing severe gradient over mitral valve.

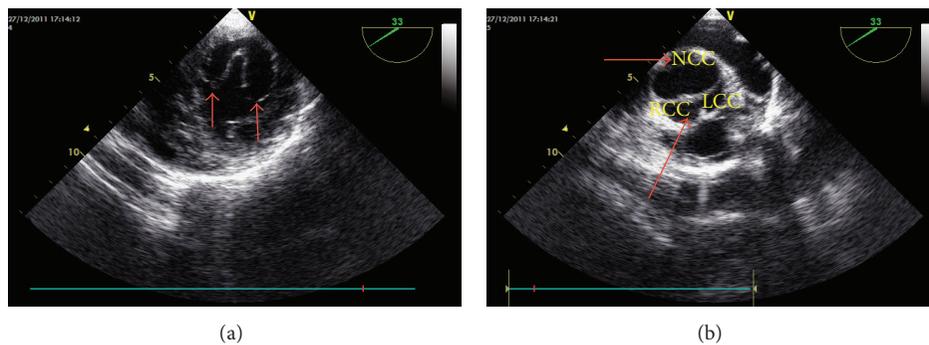


FIGURE 2: Modified section of mitral valve and short axis view of the bicuspid aortic valve with fusion of the left and right coronary cusps (LCC and RCC) in transesophageal echocardiography.

syndrome [5], persistent left superior vena cava, complete endocardial cushion defect [6], sinus venosus type ASD, Ebstein's abnormality and right atrial mass [7], left ventricular noncompaction [8], and cardiac papillary fibroelastoma [9] and as part of various syndromes with extracardiac and cardiac manifestations [10]. Isolated DOMV is very rare [11].

Valve development is a complex process which is not completely understood. Biomechanical properties of the valve and the surrounding as well as signalling pathways regulating migration and differentiation of cellular components of precursor are thought to be responsible for the embryogenesis and pathogenesis of subsequent valvular diseases.

Certain genes implicated in pathogenesis of BAV are found to regulate overall differentiation and morphogenesis of all the valves. For example, NOTCH-1 signalling, which is implicated in pathogenesis of aortic valve disease, is shown to

play important roles in normal valvulogenesis of all the valves by regulating activity of various other genes and transcription factors. Abnormalities in this gene or other associated genes and/or transcription factors might be responsible for the pathogenesis of DOMV and other associated valvular abnormalities, vascular diseases, and congenital heart defects.

4. Conclusion

DOMV is a very rare lesion which might be associated with various other left sided obstructive lesions. The hypothesis that a central genetic abnormality may result in DOMV and other associated abnormalities is attractive and should be tested. In the clinical settings, patients with DOMV should undergo cautious and detailed assessment for other components of Shone complex.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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