Double-orifice mitral valve associated with bicuspid aortic valve: a rare case of incomplete form of Shone’s complex

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CASE REPORTS

Double-orifice mitral valve (DOMV) is a rare congenital malformation characterized by two separate valve orifices of varying sizes in association with the abnormalities of the subvalvular apparatus. It has been reported to be associated with a variety of other congenital malformations. We report a rare case of incomplete form of Shone’s complex composed of a complete bridge-type DOMV and bicuspid aortic valve, well demonstrated by both transthoracic and transoesophageal echocardiography and cardiac magnetic resonance imaging.

KEYWORDS
Double-orifice mitral valve; Bicuspid aortic valve; Shone’s complex; Transoesophageal echocardiography; Cardiac magnetic resonance imaging

Case report

A 32-year-old male was referred to our hospital after detection of a cardiac murmur during a routine clinical examination. He was suffering from shortness of breath during strenuous physical activity. On physical examination, there was a grade II/VI mid-systolic ejection murmur over right upper sternal border radiating through the neck. The blood pressure was 140/90 mmHg. Electrocardiography, chest X ray, and his blood tests showed no abnormality. Transthoracic echocardiography (TTE) revealed a mild concentric left ventricular hypertrophy with normal systolic function and a moderate degree of aortic stenosis with maximal and mean transvalvular gradients of 54 and 34 mmHg, respectively. A mild degree of aortic regurgitation was also detected. It was technically difficult to decide whether the aortic valve was bicuspid or not. The mitral inflow was normal and there was no regurgitation. On parasternal short-axis view, two separate mitral valve orifices of equal size were visualized. Transoesophageal echocardiography was performed to define the anatomy of the mitral and the aortic valves more accurately. Transgastric short-axis view confirmed the presence of a double-orifice mitral valve (DOMV) with circular orifices of equal size (Figure 1A, Supplementary data online, Video 1). Colour Doppler showed two separate diastolic inflow jets (Figure 1B). The modified transgastric TEE view at 45° demonstrated the atroventricular orifice divided into two inlets by the central fibrous bridge (Figure 2A, Supplementary data online, Video 2). The transgastric long-axis view disclosed that the chordae from each orifice were attached to their own subvalvular apparatus separately, and the tensor apparatus was composed of multiple papillary muscles. Both the posteroomedial and the anterolateral papillary muscles seemed to be formed by fusion of two separate muscles. The posteroomedial papillary muscle was displaced more posteriorly and another papillary muscle in the middle was attached to the central fibrous bridge (Figure 1C and D). Mid-oesophageal view at 90° revealed complete separation of the two orifices by a fibrous bridge (Figure 1C and D). Transoesophageal echocardiography also confirmed that the aortic valve was bicuspid.

No other congenital cardiac abnormality was detected on echocardiography. For more detailed evaluation, we referred the patient for cardiac magnetic resonance imaging (MRI). Cardiac MRI detected no other abnormality but the short-axis view well demonstrated the multiple papillary muscles (Figure 2B). The patient was scheduled for follow-up.

Discussion

Double-orifice mitral valve is a rare congenital malformation characterized by two separate valve orifices of varying sizes in association with the abnormalities of the subvalvular apparatus. It may have no haemodynamic significance or may cause clinically significant mitral stenosis or mitral
DOMV has been reported to be associated with a variety of other congenital malformations. According to the classification system proposed by Trowitzsch et al., DOMV in our case was a complete bridge type with equal circular orifices and was associated with bicuspid aortic valve. This combination can be considered as an incomplete form of Shone’s complex. The Shone’s complex has been defined as the presence of four left-sided defects such as supravalvular mitral membrane, parachute mitral valve, subaortic stenosis, and aortic coarctation. In the presence of less than four of these features, it is considered the incomplete form. The congenital mitral pathology in our case was DOMV with parachute chordal attachments and associating aortic arch anomaly was bicuspid aortic valve. Either parachute mitral valve or DOMV has been regarded to be the anatomical variants of the same malformation characterized by convergence of chordal insertion.

The presence of two separate regurgitant jets on transthoracic parasternal short-axis view should always arouse the suspicion of DOMV. However, in adults, transoesophageal echocardiography (TEE) is usually necessary for a definite diagnosis of DOMV. Moreover, TEE helps to differentiate DOMV from other possible acquired disorders such as perforation, rupture, and partial fusion of leaflets due to endocarditis. In differential diagnosis, the tensor apparatus is the key element. Bano-Rodrigo et al. concluded that DOMV was always associated with an anomaly of either the chorda tendinea or the papillary muscles. In the absence of a clinical suspicion for endocarditis, two separate mitral jets associated with the abnormalities of the subvalvular apparatus makes the diagnosis of DOMV definite. Das et al. defined the associated anomalies of the subvalvular apparatus such as accessory and/or redundant chordal attachments, chordal ring, accessory septal attachment, parachute chordal attachments and

Figure 1  TEE images; transgastric short-axis view showed two circular orifices of equal size (A) and the presence of two separate inflow jets (B). Transgastric long-axis view demonstrated multiple papillary muscles. The posteromedial orifice had chordal insertions to posteriorly displaced, two-headed papillary muscle (C). The anterolateral papillary muscle group was also formed by two separate muscles (D). The papillary muscle in the middle had a chordal attachment to the central fibrous bridge (arrowhead). Abbreviations: a, anterolateral orifice; b, posteromedial orifice; pm and al, posteromedial and anterolateral papillary muscle(s), respectively.

Figure 2  The modified transgastric TEE view at 45° demonstrated the central fibrous bridge dividing the atrioventricular orifice into two inlets (A). Cardiac MRI short-axis view showed multiple papillary muscles originating from the myocardium separately (B). Abbreviations: a, anterolateral orifice; p, posteromedial orifice; LA, left atrium; LV, left ventricle.
multiple, unequal, or fused papillary muscles. The associated subvalvular abnormalities in our case were accessory and parachute chordal attachments together with supernumerary, unequal, and fused papillary muscles.

We preferred to use the cardiac MRI as an adjunctive modality in order to investigate a probable concomitant congenital abnormality and to demonstrate the subvalvular apparatus more accurately. However, cardiac MRI adds little to the findings of the echocardiographic assessment. Despite its success in demonstration of the subvalvular apparatus, MRI failed in visualization of the structure of the leaflets itself and in flow analysis through the valves.

This case report raises the awareness about this rare syndrome and highlights the importance of a comprehensive examination. In the presence of a left-sided valvular abnormality on echocardiographic examination, other valves and the aortic arch should be screened with extra caution.

Supplementary data

Supplementary data are available at European Journal of Echocardiography online.

References


