Double-Orifice Mitral Valve in an Adult with Atrioventricular Reentrant Tachycardia

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INTRODUCTION

Double-orifice mitral valve (DOMV) is a rare congenital cardiac anomaly. Since the advent of echocardiography, >200 cases have been reported. DOMV has been associated with several congenital heart defects, including atrioventricular septal defects, trisomy 21, Ebstein’s anomaly, coarctation of the aorta, and bicuspid aortic valves. DOMV is typically discovered during childhood as an incidental finding during evaluation for complex cardiac anomalies and is an exceedingly rare finding in adults. In this case, a 28-year-old patient who presented with palpitations and syncope was found to have DOMV with a bicuspid aortic valve and atrioventricular reentrant tachycardia from a concealed left-sided accessory pathway. To our knowledge, this is the first reported association of DOMV with an accessory pathway.

CASE PRESENTATION

A 28-year-old woman presented with palpitations and syncope. Physical examination revealed tachycardia with regular rhythm and a systolic murmur consistent with mitral regurgitation. Twelve-lead electrocardiography showed normal sinus rhythm without delta waves (Figure 1).

Subsequent event monitoring revealed wide-QRS tachycardia with regular rhythm and a systolic murmur consistent with mitral regurgitation. Twelve-lead electrocardiography showed normal sinus rhythm without delta waves (Figure 1).

The patient underwent electrophysiologic study and subsequent radiofrequency ablation of a left retrograde-only conducting lateral accessory pathway, with resolution of her palpitations and reentrant tachycardia. The patient’s mitral regurgitation was not affected by the ablation. At 1-year follow-up, the patient’s symptoms had not returned.

DISCUSSION

Three types of DOMV have been described by two-dimensional echocardiography, namely, the complete bridge, incomplete bridge, and hole types. In the complete-bridge type, the two orifices are configured like “spectacles,” with a dividing fibrous bridge dividing two well-defined circular valves from the annulus down to the leaflet edges. In the incomplete-bridge type, this tissue bridge is observable only at the leaflet edge, while at the level of the annulus, only a single ring is detected, resembling a funnel with two outlets. The hole type is characterized by a second orifice, smaller in size, that sits in the lateral commissure of the valve at the midleaflet level.

However, these subtypes have not been associated with variations in symptoms or severity. Likewise, presenting symptoms have not been attributed to the presence of a DOMV alone. Rather, symptoms were related to the degree of concomitant mitral regurgitation (26%–43% of patients), mitral stenosis (13%–26% of patients), or other cardiac anomalies as described above. As such, DOMV is usually detected in childhood as an incidental finding during workup for complex cardiac anomalies. In this case, our patient did not have clinically significant mitral valve regurgitation or stenosis. Instead, DOMV was detected during workup for tachycardia. Because of its associated congenital conditions, it is exceedingly rare to diagnose DOMV in adults, with studies of 27 and 46 patients with DOMV reporting median ages at presentation of 9 months and 2.6 years, respectively.

To our knowledge, this is also the first case of DOMV associated with an accessory pathway. The patient’s atrioventricular reentrant tachycardia was found to be the result of a concealed left-sided accessory pathway. In this case, electrical signals originating in the atria can conduct to the ventricles through the His-Purkinje system but cannot conduct antegrade through the accessory pathway, and as such would not generate the classic delta wave on electrocardiography (the accessory pathway is “concealed”). However, the impulse can reenter into the atria through the retrograde conducting pathway, thus initiating a reentrant tachycardia. It is possible that the development process that led to the patient’s DOMV was also responsible for the existence of this anomalous accessory pathway.

CONCLUSION

We present a novel case of an adult patient who was incidentally found to have a DOMV associated with a concealed accessory pathway participating in atrioventricular reentrant tachycardia and bicuspid aortic valve.
Figure 1  Electrocardiogram showing sinus rhythm without delta waves.

Figure 2  Event monitor strip showing wide-QRS tachycardia with left bundle branch block configuration transitioning to narrow-QRS tachycardia at similar cycle length, highly suggestive of an atrioventricular reentrant tachycardia with a left-sided accessory pathway.

Figure 3  Transthoracic echocardiogram, short-axis view, without Doppler, showing DOMV with a tissue bridge at the level of the mitral valve annulus, indicating a complete bridge-type DOMV.

Figure 4  Transthoracic echocardiogram, short-axis view (off-axis) without Doppler, showing two well-defined circular mitral valves divided by a tissue bridge, confirming the complete bridge-type DOMV.

Figure 5  Transesophageal echocardiogram, midesophageal long-axis (off-axis) view in diastole (left) and systole (right), without Doppler, providing further characterization of the DOMV.
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