CASE REPORT

Successful balloon mitral valvotomy in a rare coexistence of Ebstein's anomaly and rheumatic mitral stenosis

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Accepted 20 April 2016

SUMMARY

Co-existence of Ebstein's anomaly of the tricuspid valve with rheumatic mitral stenosis is a very rare occurrence. We report the case of a young man who presented with progressive dyspnoea and was found to have rheumatic mitral stenosis with pulmonary hypertension and Ebstein's anomaly of the tricuspid valve. The patient underwent successful balloon mitral valvotomy resulting in marked improvement of symptoms.

BACKGROUND

Ebstein's anomaly is a rare congenital heart disease with an incidence of 1 per 21 000 live births.¹ Various co-existent anomalies of the left side of the heart have been described with this condition, including mitral stenosis (MS), parachute mitral valve, supraventricular mitral ring and cleft mitral leaflet in association with endocardial cushion defect and coarctation of the aorta.^{2 3} Rheumatic heart disease associated with Ebstein's anomaly of

the tricuspid valve (TV) is an extremely rare clinical entity. We report the case of a young man with Ebstein's anomaly of the TV and severe rheumatic MS who underwent successful balloon mitral valvotomy (BMV).

CASE PRESENTATION

A 28-year-old man with no previous history of rheumatic fever, hypertension, diabetes or any other major systemic illness, presented with progressive New York Heart Association (NYHA) functional class II dyspnoea of 3-year duration. Clinical examination revealed a low volume regular pulse with a rate of 88 bpm, blood pressure (BP) of 112/82 mm Hg and finger oximetry arterial saturation of 97% on room air. The jugular venous pulse was normal in height, with a prominent 'a' wave. Apical impulse was in the fifth intercostal space directly medial to the mid-clavicular line. On auscultation there was a loud and split S1, a loud S2 and an opening snap. A mid-diastolic rumbling murmur

Figure 1 Twelve-lead ECG showing sinus rhythm with normal PR interval, bifid P wave with tenting of its initial component and a prominent negative terminal deflection along with an rSR' pattern in V1.

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To cite: Sidhu NS, Kondethimmanahally Rangaiah SK, Ramesh D, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-214674

Figure 2 Two-dimensional echocardiogram in four-chamber view demonstrating apical displacement of septal tricuspid leaflet with severe tricuspid regurgitation (TR) and thickened mitral leaflets with restricted opening.

with presystolic accentuation was present at the apex and a grade 3/6 pan-systolic murmur without respiratory variation was present at the left sternal border.

INVESTIGATIONS

Routine pathological and biochemical investigations were within normal limits. ECG showed sinus rhythm with normal PR interval, bifid P wave with tenting of its initial component and a prominent negative terminal deflection along with a rSR' pattern in V1. There was no evidence of pre-excitation on the ECG (figure 1). Chest X-ray revealed mild cardiomegaly with biatrial enlargement.

Transthoracic echocardiogram showed a large anterior tricuspid leaflet with apical displacement of the septal leaflet by 17 mm (11.33 mm/m²), consistent with the diagnosis of Ebstein's anomaly (figure 2 and video 1). There was biatrial enlargement with a moderate sized atrialised right ventricle (RV) and a smaller, though adequately sized, functional RV. The combined area of the right atrium and atrialised RV was much smaller than the combined area of the functional RV, left ventricle and left atrium (LA). There was severe tricuspid

regurgitation (TR) with calculated pulmonary artery systolic pressure (PASP) of 71 mm Hg, which is unlikely for an isolated Ebstein's anomaly. Mitral leaflets were thickened with bicommissural fusion, submitral disease and diastolic doming of the anterior mitral leaflet, consistent with rheumatic MS (figure 3 and video 2). There was no evidence of mitral regurgitation. The mitral valve orifice area (MVOA) by planimetry was 0.7 cm² and mean gradient across the mitral valve was 10 mm Hg (figure 4). Wilkin's echocardiographic score for the mitral valve was 6. The aortic and pulmonary valves were normal, and both ventricles had good systolic function. The interatrial septum (IAS) was intact and bulging towards the right atrium. A transoesophageal echocardiogram revealed similar findings with no evidence of left atrial thrombus.

TREATMENT

As the patient was acyanotic, with neither symptoms nor signs suggestive of heart failure or arrhythmia episodes, we thought that severe mitral stenosis was the cause of his symptoms. As the valve morphology was found to be suitable (Wilkin's score of 6), a percutaneous BMV was planned. After taking right femoral

Video 1 Two-dimensional echocardiogram in four-chamber view demonstrating apical displacement of septal tricuspid leaflet with severe tricuspid regurgitation (TR) and thickened mitral leaflets with restricted opening.

Figure 3 Two-dimensional echocardiogram in parasternal long-axis view showing thickened mitral leaflets with restricted opening and diastolic doming of anterior mitral leaflet (AML).

Video 2 Two-dimensional echocardiogram in parasternal long-axis view demonstrating thickened mitral leaflets with restricted opening and diastolic doming of anterior mitral leaflet (AML).

arterial and venous access, a trans-septal puncture was performed using a 70 cm curved Brockenbrough needle and an 8-French Mullins sheath-dilator combination. Mean left atrial pressure was 26 mm Hg and mean right atrial pressure was 11 mm Hg. A 26 mm ACCURA balloon catheter (Vascular

Figure 5 Still fluoroscopy frame in right anterior oblique (RAO) view demonstrating the inflated Accura balloon catheter across the mitral valve.

Concepts, Essex, UK) was introduced into the LA over the spring guidewire and positioned across the mitral valve. Two serial inflations were performed at 23 cm³ and 24 cm³ (figure 5), which resulted in splitting of the lateral commissure and a decrease in mean left atrial pressure from 26 to 18 mm Hg. Post-procedure echocardiogram showed a laminar forward flow across the mitral valve and improved opening of the mitral valve, with decreased diastolic doming of the anterior mitral leaflet (figure 6 and video 3). There was a decrease in mean transmitral gradient from 10 to 5 mm Hg, and an increase in MVOA from 0.7 to 1.5 cm² (figure 7). PASP decreased from 71 to 57 mm Hg post-procedure. There was no evidence of mitral regurgitation post-procedure.

OUTCOME AND FOLLOW-UP

The patient was discharged after 2 days, without any complications. He was asymptomatic at 1-year follow-up.

Figure 4 (A) Two-dimensional (2D) echocardiogram in parasternal short-axis view at mitral valve showing thickened mitral leaflets, restricted opening and bicommissural fusion. (B) 2D echocardiogram apical four-chamber view with continuous wave (CW) Doppler across mitral valve showing a mean gradient of 10 mm Hg.

Figure 6 Post-procedure echocardiogram in parasternal long-axis view demonstrating improved mitral valve opening after balloon mitral valvotomy (BMV).

Video 3 Post-procedure echocardiogram in parasternal long-axis view demonstrating improved mitral valve opening after balloon mitral valvotomy (BMV).

DISCUSSION

Although our patient had no history of rheumatic fever, the diagnosis of rheumatic MS was made from the typical appearance of the mitral valve with thickened leaflets, bicommissural fusion and diastolic doming of the anterior mitral leaflet. The septal tricuspid leaflet was apically displaced by 17 mm (11.33 mm/m^2), consistent with the diagnosis of Ebstein's anomaly, which requires apical displacement of the septal leaflet of the TV from the insertion of the anterior leaflet of the mitral valve by at least 8 mm/m^2 body surface area.⁴ The Ebstein's anomaly in our patient was not severe and the functional RV had adequate size (Carpentier type A Ebstein's anomaly).⁵

Figure 7 (A) Post-procedure echocardiogram in parasternal short-axis view demonstrating improved mitral valve opening with splitting of lateral commissure. (B) Post-procedure echocardiogram in apical four-chamber view demonstrating a mean gradient of 5 mm Hg.

Surgical intervention in patients with Ebstein's anomaly is indicated in those presenting with severe symptoms, NYHA functional class III-IV, substantial cyanosis, heart failure, occurrence of paradoxical embolism, progressive increase in cardiothoracic ratio on chest X-rays and recurrent supraventricular arrhythmias controlled by neither medical nor ablative therapy.⁶ Our patient had none of these features and was symptomatic primarily because of severe MS.

Association of Ebstein's anomaly of the TV with rheumatic MS is an extremely rare clinical entity. Most cases have been reported from India.⁷⁻⁹ Association of Ebstein's anomaly with MS may alter the haemodynamics, clinical presentation and natural history of MS and vice-versa.¹⁰ In the presence of interatrial communication, which is the case in 80–94% of patients with Ebstein's anomaly,^{11 12} high left atrial pressure due to mitral stenosis may lead to left-to-right atrial shunt causing volume overload of the right atrium and right ventricle, which may eventually lead to right heart failure. Decompression of the LA into the right atrium may prevent the development of pulmonary oedema. In patients with this association who have an intact interatrial septum, left ventricular abnormalities (such as systolic and diastolic dysfunction) as a part of Ebstein's anomaly may lead to further elevation of left atrial pressure and more chances for development of pulmonary oedema. Significant associated MS may cause earlier occurrence of atrial fibrillation, pulmonary hypertension and heart failure in patients with Ebstein's anomaly. However, in our patient, the interatrial septum was intact, and this prevented the left-to-right shunt and volume overload of the right-sided chambers but the MS resulted in severe elevation of pulmonary artery pressure, which is unusual in cases of isolated Ebstein's anomaly.

Our patient was symptomatic primarily because of severe MS. Hence BMV was considered, as the valve morphology was suitable. There was no indication for surgical repair of the Ebstein's anomaly.

The technical difficulties for BMV in case of co-existing Ebstein's anomaly of the TV include difficult septal puncture in view of an enlarged right atrium and altered morphology of IAS. In cases with significant enlargement of the right atrium, the free wall enlarges while the IAS is pushed rather inferiorly. This increases the chances of a high septal puncture with attendant risk of haemopericardium and cardiac tamponade. Transoesophageal echocardiography (TOE) can be helpful in avoiding this complication.¹³ A very large right atrium may not allow adequate tenting of the interatrial septum for the needle to enter the LA. To facilitate septal puncture in such cases, the curve of the standard needle may be increased or a needle with

a larger preformed curve such as a BRK-1 (St Jude Medical) may be used.¹⁴ We did not encounter any procedural difficulty probably because the right atrium in our patient was not massively enlarged.

There have been few case reports of BMV in a symptomatic patient with Ebstein's anomaly of the TV and rheumatic MS. In one such report, the valvotomy was performed using a double balloon technique.¹⁵ In another case, BMV was performed using an Inoue balloon catheter.¹⁶ In our case, it was carried out with an ACCURA catheter.

This case is significant since this rare combination of congenital heart disease with acquired valvular heart disease may be encountered in tropical countries where rheumatic heart disease is still prevalent; BMV can be performed safely and successfully in this complex clinical situation, without much technical difficulty.

Learning points

- ▶ The co-existence of Ebstein's anomaly and rheumatic mitral stenosis is a rare entity.
- ▶ Balloon mitral valvotomy can be performed in this situation in the usual manner, though technical difficulty may be encountered occasionally while puncturing the interatrial septum due to altered anatomy of the right atrium and interatrial septum in this entity.
- ▶ Long-term follow-up is needed for such cases to detect the recurrence of symptoms due to progression of rheumatic valvular disease and/or Ebstein's malformation.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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