Ebstein’s anomaly of tricuspid valve with rheumatic mitral stenosis: a rare association
Sanjeev Asotra, Rajeev Merwaha, Davinder Pal Singh, Kunal Mahajan

Department of Cardiology, Indira Gandhi Medical College, Shimla, Himachal Pradesh, India

Correspondence to Dr Kunal Mahajan, kunalmahajan442@gmail.com

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DESCRIPTION
A 16-year-old girl presented with progressive exertional dyspnoea and palpitations of 3-year duration. The medical history revealed one episode of fever with migratory polyarthritis at the age of 7 years, but it was not further evaluated. On presentation, she was in sinus rhythm with a pulse rate of 88/min and blood pressure of 110/80 mm Hg. Examination revealed a tapping apical impulse in the fifth intercostal space, inside the mid-clavicular line, and a grade 3 parasternal heave. Auscultation revealed loud S1 with an opening snap. There was a mid-diastolic rumble at the apex, with presystolic accentuation. A pansystolic murmur of tricuspid regurgitation was audible. Basal crepitations and peripheral oedema were absent. Transthoracic echocardiography revealed severe rheumatic mitral stenosis (MS) (video 1) (peak and mean mitral valve gradient of 25 and 18 mm Hg, respectively, and mitral valve area (MVA) of 0.74 cm² by pressure half time (figure 1) and 0.64 cm² by two-dimensional planimetry (figure 2)). In addition, the patient had Ebstein’s anomaly of tricuspid valve with apical displacement of septal tricuspid leaflet by 32 mm and an elongated anterior tricuspid leaflet (figure 3 and video 2). Severe tricuspid regurgitation (video 3) was identified, with a gradient of 58 mm Hg (figure 4). The interatrial septum was intact. The patient underwent successful balloon mitral valvotomy (BMV) using the Inoue technique. Post-BMV, MVA increased to 1.5 cm² and peak and mean mitral valve gradient decreased to 17 and 9 mm Hg, respectively. The patient showed a dramatic improvement in her symptoms. Ebstein’s anomaly was not corrected due to lack of associated right heart failure, cyanosis and...

Video 1 Transthoracic echocardiogram, parasternal long-axis view, showing a thick and doming anterior mitral leaflet. The posterior mitral leaflet is also thickened with markedly reduced mobility. Also note the apical displacement of the septal tricuspid leaflet.

Figure 1 Estimation of gradients across mitral valve (peak gradient=25 mm Hg, mean gradient=18 mm Hg) and calculation of mitral valve area (0.74 cm²) by pressure half time.

Figure 2 Calculation of mitral valve area (0.64 cm²) by two-dimensional planimetry.

Figure 3 Transthoracic echocardiogram, apical four-chamber view, showing the apical displacement of septal tricuspid leaflet by 31.9 mm. Note the atrialised portion (*) of the RV. Also note the doming of the anterior mitral leaflet. LA, left artery; LV, left ventricle; RA, right artery; RV, right ventricle.
recurrent paradoxical embolism. Ebstein’s anomaly is a rare congenital heart disorder occurring in $\approx 1/200\,000$ live births and accounting for $< 1\%$ of all cases of congenital heart disease. Acquired rheumatic mitral valve disease is an extremely rare association. Severe MS is likely to alter the natural history of Ebstein’s anomaly, leading to a rapid progression of symptoms and an earlier onset of atrial fibrillation, heart failure and pulmonary hypertension. BMV in such a complex clinical scenario that offers an excellent outcome.

Learning points

- Ebstein’s anomaly of tricuspid valve is a rare anomaly with a reported prevalence of $1/200\,000$ live births.
- Acquired rheumatic mitral stenosis in association with Ebstein’s anomaly of tricuspid valve is extremely rare.
- This can lead to earlier onset of adverse complications, which include atrial fibrillation, hypertension and heart failure, in a patient of Ebstein’s anomaly.
- Balloon mitral valvotomy offers an excellent treatment approach in the absence of contraindications. Relief of mitral stenosis decreases the pulmonary hypertension and severity of associated tricuspid regurgitation.

Competing interests None declared.

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REFERENCES