Ebstein's anomaly of tricuspid valve with rheumatic mitral stenosis: a rare association

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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 polyarthritides 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

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

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 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 atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

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Video 2 Transthoracic echocardiogram, apical four-chamber view, showing the apical displacement of the septal tricuspid leaflet and a sail-like elongated anterior tricuspid leaflet. Thickened mitral valve leaflets with doming of anterior mitral leaflet and fixed posterior mitral valve leaflet are also seen suggestive of rheumatic mitral valve disease.

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.¹ 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.² BMV in such a complex clinical scenario that offers an excellent outcome.³

Video 3 Transthoracic echocardiogram, apical four-chamber view, showing eccentric jet of severe tricuspid regurgitation.

Figure 4 Continuous wave Doppler estimation of gradient across the tricuspid regurgitation jet.

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.

Patient consent Obtained.

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