Congenitally corrected transposition of great arteries

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DESCRIPTION

A 60-year-old woman initially presented 8 years ago with atypical chest pain and palpitations. Her 12-lead ECG demonstrated complete heart block, following which she underwent dual-chamber pacemaker implantation. She also had transthoracic echocardiogram, which was felt to be technically difficult due to body habitus. She was diagnosed to have mild mitral valve regurgitation, due to mild mitral valve prolapse, and was kept under annual surveillance. There was no other significant medical history. Her last TTE was done a year ago and she had been symptom-free up to her recent clinic visit. Her repeat TTE showed apically displaced Ebstein-like left atrioventricular valve (AV) with tricuspid valve morphology (confirmed by three-dimensional (3D) echo) and heavily trabeculated morphologically right ventricle on the left side of the heart (figures 1, 2, 3 and 4, online supplementary video 1 and supplementary video 2). Her systemic ventricle was mild to moderately impaired, and the tricuspid, left AV was moderately regurgitant. There are no obvious shunt and no other significant valvular abnormalities identified on this recent study. The above findings (atrioventricular and ventriculoarterial discordance) were in keeping with congenitally corrected transposition of great arteries (CC-TGA).

CC-TGA is a rare congenital heart defect accounting for <1% of all congenital heart diseases.[1] They are usually associated with ventricular septal defect, pulmonary stenosis, Ebstein-like malformation of the systemic AV valve and high-degree AV block. Patients without coexistent shunts are usually asymptomatic and usually diagnosed from investigations like ECG, chest X-ray and TTE done for other reasons. This condition is often missed because of the failure to recognise the above anatomical variation, as in our case.[2] Patients with CC-TGA require regular long-term follow-up as

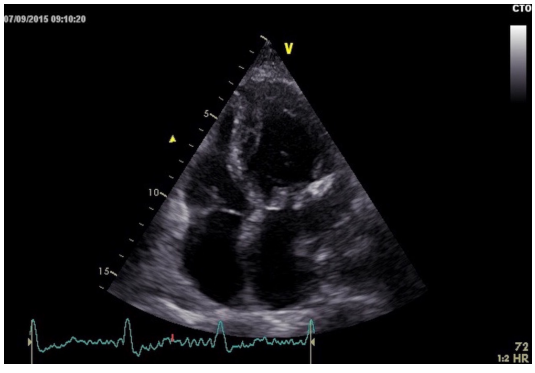

Figure 1 Transthoracic echocardiogram (atypical 4 chamber view) showing Ebstein-like malformation of the systemic atrioventricular valve and heavily trabeculated morphologically right ventricle on the left side.

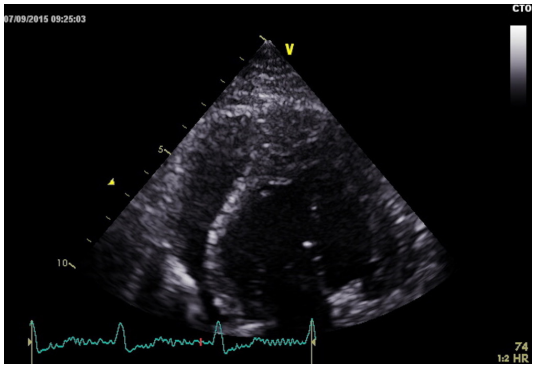

Figure 2 Transthoracic echocardiogram (A4C, zoom view) showing heavily trabeculated morphologically RV on the left side.

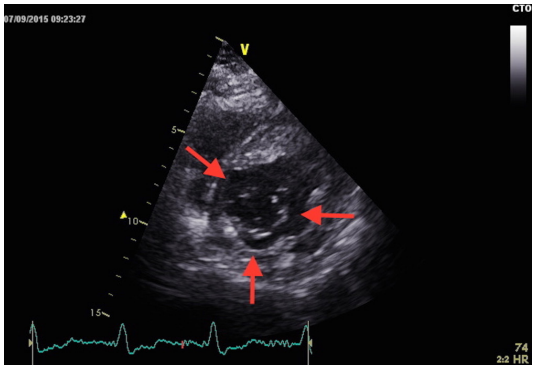

Figure 3 Transthoracic echocardiogram (parasternal short axis view) showing tricuspid valve morphology (arrows) of the systemic atrioventricular valve.

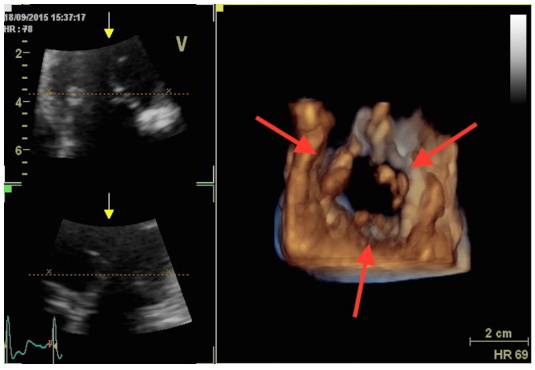

Figure 4 Live three-dimensional zoom view. Left hand panel shows the crop line placed on the systemic atrioventricular (AV) valve. Right hand panel shows the tricuspid systemic AV valve (arrows) viewed from the left atrium.

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they are prone to develop high-degree AV block, systemic AV valve regurgitation and systemic ventricular failure. This case also illustrates how newer modalities like 3D echo can be helpful in diagnosing this uncommon congenital heart disease.

Learning points

- Congenitally corrected transposition of great arteries (CC-TGA) is a rare congenital heart defect, mostly associated with ventricular septal defect, pulmonary stenosis, Ebstein-Like malformation of the systemic atrioventricular (AV) valve and high-degree AV block.
- Patients with CC-TGA entail long-term follow-up as they are at risk of developing high-degree AV block, systemic AV valve regurgitation and systemic ventricular failure.

Contributors BS was responsible for managing the patient along with cardiologist-in-charge and helped in manuscript preparation. HM performed three-dimensional echo and helped in manuscript preparation. MNP is the cardiologist-in-charge of the patient and critically revised the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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