Congenitally corrected transposition of the great vessels associated with morphological right ventricular non-compaction presenting with supraventricular tachycardia

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DESCRIPTION
A 25-year-old man presented to the emergency department with acute onset palpitation. He had a history of four to five similar episodes in the past 2 months, which had resolved on their own. ECG showed supraventricular tachycardia (SVT) with a rate of 170/min (figure 1). He was treated with intravenous adenosine which reverted the tachycardia. Transthoracic echocardiogram was attempted but an adequate window could not be obtained as the patient was obese. Only the spatial relationship of the two great vessels in the form of malposition could be made out which confirmed the aorta was left and anterior and the pulmonary artery was right.

Figure 1 ECG showed supraventricular tachycardia.

Figure 2 Transthoracic echocardiogram showed the spatial relationship of the two great vessels where aorta on the left and anterior of the pulmonary artery.
and posterior (figure 2). The visceral situs was normal. Hence a transoesophageal echocardiogram was performed which suggested a diagnosis of corrected transposition of great vessels (c-TGV) with dynamic left ventricular outflow tract obstruction with a peak gradient of 20 mm Hg (figure 3, video 1). Dysfunction of the systemic ventricle (morphological right ventricle, MRV) was also noted. Cardiac MRI confirmed the diagnosis as c-TGV with small perimembranous ventricular septal defect and non-compaction of the MRV with a ratio of the non-compacted to compacted segment greater than 2.3 as per the present criteria1 (figure 4A,B).

There are only a few case reports in the literature where c-TGV has been associated with non-compaction of MRV2 and few cases have been reported with SVT3. In our case, there was presence of SVT and non-compaction of MRV in a case of c-TGV.

Figure 3 Transthoracic echocardiogram suggested a diagnosis of corrected transposition of great vessels with dynamic left ventricular outflow tract obstruction.

Video 1 TEE suggested a diagnosis of c-TGV with dynamic left ventricular outflow tract obstruction.

Figure 4 (A and B) CMRI showed the relationship suggesting corrected transposition of great vessels and confirmed the presence of non-compaction of morphological right ventricle.
Learning points

▸ Congenitally corrected transposition of great arteries (TGA) can be associated with non-compaction of morphological right ventricle.
▸ Supraventricular tachyarrhythmias are a rare occurrence with corrected transposition of great vessels.
▸ Cardiac MRI is most reliable investigation to diagnose non-compaction.

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REFERENCES