Late presentation of congenitally corrected transposition of the great arteries

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DESCRIPTION

A woman in her 70s with no significant medical history presented with a 2-week history of dyspnoea. She underwent no annual health check-up and was not on any medication. On physical examination, there was jugular venous distension and peripheral oedema. An ECG revealed a sinus rhythm with left bundle branch block and left axis deviation; however, no arrhythmia was noted. Chest radiograph revealed cardiomegaly and bilateral pleural effusion. Chest CT revealed an anatomical transposition of the ascending aorta and pulmonary artery. Subsequent coronary CT angiography revealed anterior and posterior positioning of the ascending aorta and the pulmonary artery, respectively, and the ventriculoarterial discordance and the parallel course of the great arteries (**figure 1A**). The aorta and the pulmonary artery could be seen arising from the morphological right ventricle (RV) with trabeculation and the morphological left ventricle (LV), respectively (**figure 1B,C**). Notably, the right coronary artery perfused in the atrioventricular groove and the anterior interventricular sulcus. The left

coronary artery bifurcated into the diagonal branch and left circumflex artery (**figure 1D**). Transthoracic echocardiogram demonstrated trabeculation in the morphological RV with preserved ejection fraction, left atrium dilation and moderate anatomical tricuspid regurgitation; however, we objectified no intracardiac shunts, such as atrial or ventricular septal defect (VSD) and pulmonary stenosis (PS). Although the patient was in her eighth decade, she was diagnosed with congenitally corrected transposition of the great arteries (CCTGA) (situs solitus, L-loop and L-transposition) as an adult congenital heart disease (ACHD). We placed her on diuretics, and the symptom promptly subsided.

ACHD has become important in adult cardiology. CCTGA is a rare cardiovascular anomaly comprising <1% of all congenital cardiac diseases.¹ In CCTGA, the atria are at their normal location; however, there is atrioventricular and ventriculoarterial discordance whereby the right atrium is connected to the LV while the LV directly connects to the pulmonary artery. On the left side, the left atrium is connected to the RV which is connected to the aorta.^{2,3} Only 1% of patients with CCTGA have uncomplicated disease without associated lesions, such as VSD, PS and Ebstein's anomaly.⁴ In the absence of other associated anomalies, CCTGA causes no immediate haemodynamic complications, and the patient will be asymptomatic until advanced age as in this case.^{2,4} The mechanism of late heart failure onset in CCTGA is long-standing pressure and volume overload on the RV trying to meet the systemic demand. To date, only nine patients with congenitally corrected transposition of the great vessels older than 70 years have been reported in the literature.^{5,6}

The key features of CCTGA are transposition of the ascending aorta and pulmonary artery. Normally, the pulmonary artery is anterior while the ascending aorta is posterior, and the right pulmonary artery crosses the aortic arch, while the positioning of CCTGA shows transposition of the

Figure 1 (A) Contrast CT scans revealing anterior positioning of the ascending aorta and posterior positioning of the pulmonary artery. (B) Volume rendered imaging revealing atrioventricular and ventriculoarterial discordance. (C) Coronal view revealing the aorta arising from the morphological right ventricle with trabeculation. (AO, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle). (D) The right coronary artery (RCA) perfusing into the atrioventricular groove and the anterior interventricular sulcus, and the left coronary artery bifurcates into the diagonal branch (DB) and left circumflex artery (LCX).

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Learning points

- ▶ The key features of congenitally corrected transposition of the great arteries (CCTGA) are transposition of the ascending aorta and pulmonary artery.
- ▶ The patient with CCTGA in the absence of other associated anomalies can be asymptomatic until advanced age.

arteries, and the left pulmonary artery crosses the aortic arch.² Surgery is the care standard for CCTGA if it is complicated with structural abnormalities; however, general treatment for heart failure is also indicated for uncomplicated cases.³

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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