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Multimodal imaging assessment of a very rare cause of heart failure in adults

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Introduction. Congenitally Corrected Transposition of the Great Arteries (CCTGA) is a rare defect consisting in the abnormal twisting of the heart during fetal development. As a result, the two ventricles and their valves are reversed. CCTGA is frequently associated with other cardiac abnormalities. 25% of patients are developing heart failure, related to perfusion mismatch (the morphological left ventricle is supplied by a single coronary artery), and to the progressive deterioration of the structural right ventricle situated on the systemic side of the circulation.

Case report. A 45-year-old male was referred to our hospital for fatigue and dyspnea, occuring in the last five months. Physical examination revealed tachypnea, a slightly intense systolic murmur at the apex, and pulmonary congestion, in the absence of cyanosis, peripheral edema or jugular venous distension. Heart rate and blood pressure were normal. Usual laboratory work-up indicated increased levels of NT-proBNP, without any other abnormalities. ECG presented signs of pressure overload of the systemic ventricle (Figure Ia). Transthoracic echocardiography (TTE) highly suggested the diagnosis of CCTGA, due to atrioventricular valve displacement, with the morphological tricuspid valve closer to the apex in 4-chamber view (Figure Ib). TTE showed also dilated and dysfunctional left ventricle, mild left atrioventricular regurgitation, and normally functional right ventricle. Cardiac computed tomography emphasized a specific feature of CCTGA: the parallel emergence of aorta and pulmonary trunk, with the aortic arch crossing over the left pulmonary artery (Figure Ic). Cardiac magnetic resonance imaging confirmed dilatation and low ejection fraction of the systemic ventricle (20%), and displayed presence of trabeculations and the moderator band in the systemic ventricle (Figure Id). None of these evaluations found additional cardiac structural anomalies. Thus, patient was diagnosed with heart failure due to isolated CCTGA.

Discussions and relevance of case report. This case emphasizes a very rare cause of heart failure in adults. CCTGA is reported in 0.5-1% of all congenital diseases, especially in males. Isolated CCTGA accounts for less than 10% of all cases, and represents the phenotype that is usually diagnosed in adulthood. In the absence of associated anomalies, the prognosis of these patients is particularly affected by the occurrence of heart failure in the 4th or 5th decade of life. Meanwhile, this case highlights the importance of a multimodal approach in CCTGA, and the specific contribution of each imaging method in the process of an accurate diagnosis.

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