Congenitally corrected transposition of great arteries with ischemic symptoms in middle age

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ABSTRACT

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital disease first described by Von Rokitansky in 1875. Transposition of the great arteries comprises 2.6 - 7.8% of all cases of congenital heart disease, and if uncorrected, is commonly fatal in the first year of life. Patients with corrected transposition of the great arteries without associated defects may remain undiagnosed until adult life. Symptoms occur rarely before the fourth and fifth decades, when rhythm disturbance, left atrioventricular valve regurgitation, and moderately impaired systemic ventricular function cause congestive cardiac failure. We report here a case of myocardial ischemia, due to drug overdose, in CCTGA without associated anomalies.

Case Report. A 40-year-old “drug trafficker” was presented with history of swallowing a large number of tablets, with the aim of concealing the evidence. He was uncooperative in volunteering any detailed history. Except for mild chest discomfort, there was no symptoms of abdominal pain, vomiting, diarrhea, shortness of breath, or loss of consciousness. No history of smoking, diabetes mellitus (DM), hypertension (HTN), dyslipidemia, or ischemic heart disease (IHD) in the family. No past history of hospitalization due to any cardiac problem. He was a family history of congenital heart disease, DM, HTN or chronic illnesses.

Physical examination, he is of normal build, conscious and oriented with agitated mood, but not in distress. He was not pale, cyanosed, or jaundiced. No lymph node was palpated. His temperature was normal, pulse rate 82/min, blood pressure 150/100 mm Hg, and normal jugular venous pressure. Cardiac auscultation revealed a single, second heart sound and pan-systolic murmur at the apex. The examinations of other systems were essentially normal. He was admitted with an impression of drug overdose, and treated with gastric lavage and charcoal. His hemoglobin, blood sugar, urea, creatinine, and electrolytes were within normal range. The toxicology screen was positive for amphetamine. His creatinine kinase was elevated (328 u/l, normal range: 25-195 iu/L), and the electrocardiogram showed...
Discussion. Transposition of the great arteries comprises 2.6 - 7.8% of all cases of congenital heart defects, and if uncorrected, is commonly fatal in the first year of life. The clinical course is complicated by associated intracardiac defects such as ventricular septal defect, subvalvar and valvar pulmonary stenosis, left AV valve regurgitation, and AV conduction disturbances. Patients with corrected transposition of the great arteries without associated defects may remain undiagnosed until adult life. Symptoms occur rarely before the fourth and fifth decades, when rhythm disturbance, left AV valve regurgitation, and moderately impaired systemic ventricular function cause congestive cardiac failure.

This is supported by another study that patients with CCTGA are increasingly subject to congestive heart failure (CHF) with advancing age; this complication is extremely common by the fourth and fifth decades. Tricuspid (systemic AV) valvular regurgitation is strongly associated with RV (systemic ventricle in CCTGA) dysfunction and CHF, whether it is causative or a secondary complication remains speculative.

Little attention has been given to myocardial perfusion and the possible role of myocardial ischemia, infarction, or both, in right ventricular dysfunction. Impaired myocardial blood flow reserve of the anatomic RV in the absence of ischemic symptoms may be associated with reduced ventricular function of the systemic chamber. As discussed previously, a patient with CCTGA usually presents with rhythm disturbance, left AV valve regurgitation, and moderately impaired systemic ventricular function causing congestive cardiac failure. Symptoms occur rarely before the fourth and fifth decades. We conclude that in our case, the 40-year-old man, in whom we discovered CCTGA without intracardiac anomalies, presented with symptoms of angina due to vasospasm, possibly due to drug overdose of amphetamine.

References


Antero-lateral ischemic changes (Figure 1). The thallium scan showed antero-septal perfusion defect at rest. The echocardiogram (Figure 2) showed CCTGA and mild systemic valve regurgitation without associated anomalies with mild anterior wall hypokinesia, with an overall ejection fraction of 45%. He was referred to the cardiac catheterization laboratory for coronary angiography that showed anomalous origin of coronaries, left circumflex and right coronary artery arising from left coronary sinus, and left anterior descending coronary artery from right coronary sinus, without any obstructive lesion. Hence this patient, who took over dose of amphetamine, developed acute coronary syndrome, non-ST elevation myocardial infarction. For this he was treated in the hospital, and was discovered to have CCTGA.
1. Zaidi et al.


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