Case Report

Congenitally Corrected Transposition of the Great Arteries in a Seventy-Year-Old Woman

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Corrected transposition of the great arteries is a rare condition, and few patients with this abnormality survive past 50 years of age because of associated defects, or the subsequent development of atrioventricular valvular insufficiency or heart block or both. The case of our patient is of interest not only because she reached old age, but also because she lived a normal life, presenting with minor cardiac impairment and palpitations at the age of 70 years.

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Congenitally corrected transposition of the great arteries of the heart is a rare form of congenital heart disease that was first described by Von Rokitansky in 1875.[1] The atrioventricular dissociation means that the morphological right atrium discharges blood into the morphological left ventricle, while the morphological left atrium drains into the morphological right ventricle. Thus, the left ventricle supplies the pulmonary circulation while the right supports the systemic circulation. Few patients with this anomaly survive beyond 50 years of age.[2]

Case description

A woman aged 70 years, a farmer who had lived all her life in the countryside, an uneducated mother of three, came to our hospital for a cardiological evaluation because of palpitations she had felt for a year. She was referred by a private general practitioner because of the above problem and the auscultation findings. The patient reported no fainting episodes, dyspnoea, chest pain, or lower limb oedema. Her family history contained no reports of heart disease (congenital or acquired), nor any sudden death at a young age. The clinical examination revealed a single, loud second sound, a 2-3/6 holosystolic murmur at the apex and a 2/6 left parasternal diastolic murmur.

The ECG showed a QS morphology in leads V_{1-2}, a missing Q on the left precordial leads with ST-segment depression and an asymmetric T wave on the same leads, and an rS morphology in lead III. There was also first degree atrioventricular block with a PR interval of 280 ms (Figure 1).

The chest X-ray showed a slightly elevated cardiac index (17/30) and a narrow aorticopulmonary trunk. More specifically, there was no opacification of the ascending aortic arch and the main pulmonary arterial trunk, with straightening of the left upper cardiac margin and a round lower left cardiac border (Figure 2).

Blood pressure was within normal limits. Blood chemistry and biochemical examinations were also normal.

Echocardiography revealed the following:

- Normal position of the atria.
- Connection of the morphological right atrium via the right atrioventricular valve,

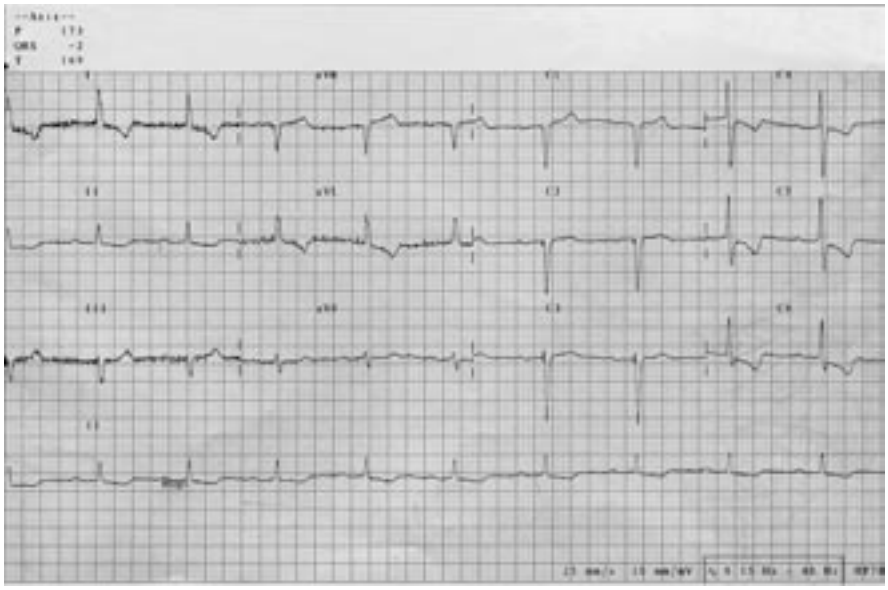

Figure 1. ECG showing normal sinus rhythm with a left axis deviation and absence of Q waves in leads V_4-V_6. The PR interval is prolonged to 280 ms.

which had mitral valve morphology, to the morphological left ventricle, which was on the right. This ventricle (venous ventricle) was connected via the right semilunar valve (pulmonary valve) to the pulmonary artery. There was no stenosis at any level, apart from negligible (1+) valvular insufficiency.

- The morphological left atrium received the pulmonary veins and was connected via the left atrioventricular valve, which had tricuspid morphology, to the morphological right ventricle, which was on the left. The left atrioventricular valve had an Ebstein morphology with a mild to moderate degree of insufficiency (2/3+). The systemic (arterial) ventricle was not appreciably distended and sustained satisfactory systolic function (ejection fraction 50%). This ventricle was connected via the left semilunar valve (aortic valve) to the aorta. The aortic valve was thickened and showed mild (2+) insufficiency.
- The course of the two great vessels was parallel (Figure 3a, b).

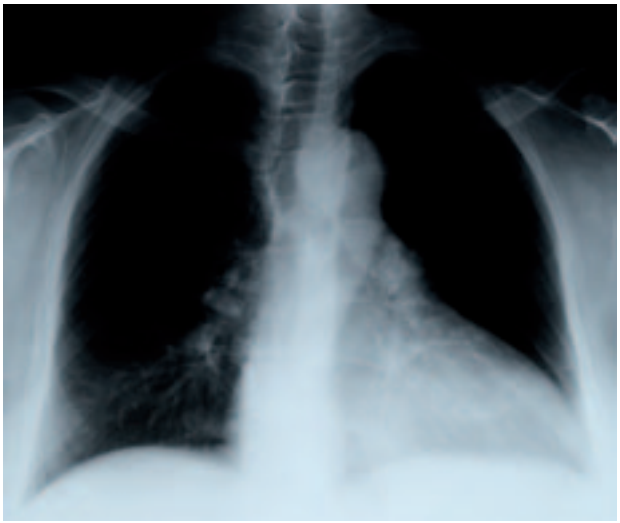

Figure 2. Posteroanterior chest X-ray showing a mildly enlarged cardiac silhouette. The ascending aortic shadow and the main pulmonary artery segment are absent. A round left lower cardiac border may be seen.

On the 24-hour Holter ECG the basic rhythm was sinus rhythm. There were occasional atrial and unifocal ventricular extrasystoles. The maximum heart rate was 118 and the minimum 45 beats/min. There were no atrioventricular conduction disturbances apart from the first degree atrioventricular block. There were no complexes indicative of preexcitation syndrome.

We recommended that the patient should undergo cardiac catheterisation, but she refused. She also refused a thallium^{201} myocardial perfusion study for the evaluation of her susceptibility to ischaemia and her coronary arteries.

She was put on medication with converting enzyme inhibitors and was advised to have an annual cardiac check up with echocardiogram and Holter. On re-examination a year later her clinical and laboratory findings remained satisfactory. Her daily physical activity continues to be entirely satisfactory for her age.

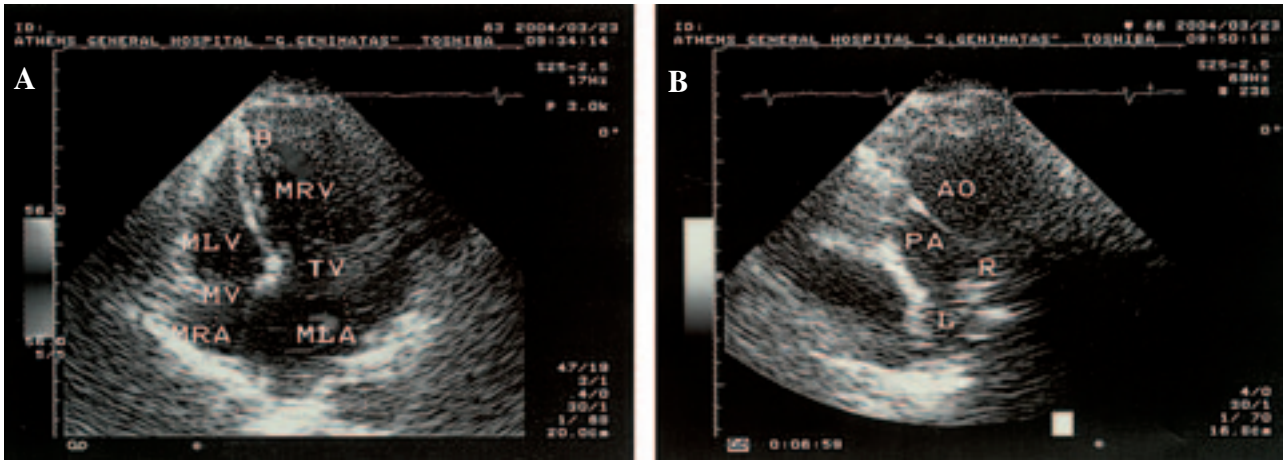

Figure 3. Transthoracic echocardiography. A: Apical four-chamber view. MB – moderator band; MLA – morphological left atrium; MRA – morphological right atrium; MLV – morphological left ventricle; MRV – morphological right ventricle; MV – mitral valve; TV – tricuspid valve. B: Modified left parasternal short axis view. The aorta arises in front of the pulmonary trunk, which is identified by its bifurcation.

Discussion

Congenitally corrected transposition of the great arteries is a rare entity that represents less than 1% of all clinically diagnosed congenital heart diseases. Recognisable associated defects are seen in 98% of cases.[3] These include the following: ventricular septal defect (74%), pulmonary valve stenosis (74%), Ebstein-type anomalies of the systemic (tricuspid) valve (>75%), and complete atrioventricular block (5%).[4] Rarer abnormalities are straddling of the interventricular defect by the right atrioventricular valve and a lone coronary artery.[3] Only 1-10% of individuals with congenitally corrected transposition of the great vessels have no associated defects.[5]

Because of the reversal of the systemic conduction, the ECG contributes significantly to the diagnosis. The abnormal direction of the initial depolarisation of the septum from right to left leads to the appearance of Q waves in the right precordial leads that are absent from the left precordial leads. Also, the His bundle is longer as a result of the greater distance between the atrioventricular node and the base of the interventricular septum. This explains the disturbances of atrioventricular conduction that are frequently seen.[6] First degree atrioventricular block is found in around 50% of cases, while its progression to complete heart block occurs at a rate of 2% per year.[7] In some patients there is also an accessory pathway that provides the anatomical substrate for pre-excitation.[6]

In corrected transposition, the coronary arteries are distributed in a similar way to the ventricles. The anterior descending branch and the circumflex artery supply the morphological left ventricle while the right coronary artery supplies the morphological right ventricle.[6]

Patients with congenitally corrected transposition very often exhibit atrial tachyarrhythmias, such as atrial fibrillation, atrial flutter, or supraventricular tachycardia. If there are no associated defects, the appearance of these arrhythmias is delayed until the fifth decade of life, much later than in patients with other abnormalities. In every case they are potentially damaging and should be treated.[2]

The clinical course of corrected transposition of the great vessels depends on the presence and severity of the associated defects. Even in the absence of such anomalies, or after their surgical repair, the question remains whether the anatomical right ventricle is capable of maintaining an adequate cardiac output over a long period.[8] There are many anatomical reasons why the right ventricle and the tricuspid valve, even when morphologically normal, are inferior to the left ventricle and the mitral valve as far as the long term maintenance of systemic circulation is concerned. The particular morphology of the tricuspid valve, the smaller papillary muscles, the conduction system and the single-vessel perfusion of the systemic ventricle by the right coronary artery, all predispose against it.[3]

Insufficiency of the systemic ventricle is the cause of death in more than 50% of patients.[7] In most cases it is accompanied by severe insufficiency of the systemic atrioventricular valve. It is not clear whether the latter is the cause of the ventricular dysfunction or the result, although in patients without associated defects systemic

atrioventricular valve insufficiency has been found to precede the deterioration in ventricular function.[9]

Graham et al[10] observed progressive dysfunction of the anatomical right ventricle in adult life. Cumming et al,[11] studying patients without associated defects, reached a different conclusion. If the insufficiency of the systemic (tricuspid) atrioventricular valve is severe it must be replaced, and this should be done before the appearance of dysfunction of the systemic right ventricle (ejection fraction >45%).[12] When the tricuspid insufficiency is combined with dysfunction of the systemic right ventricle, the double switch operation (arterial and atrial) is considered appropriate.[13,14]

The international literature contains very few cases of adult patients with congenitally corrected transposition of the great arteries without associated defects or surgical intervention. Specifically, only about 10 cases of patients aged over 65 years have been reported.[15-21] The oldest patient was a woman aged 84 years in whom the condition was discovered as a chance post mortem finding.[22] Our patient adds one more case to this short list. As far as we know, it is the first such case to be described in Greece.

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