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A Case of Idd-Type Corrected Transposition of the Great Arteries Combined with Inversion of the Viscera and Patent Foramen Ovale Complicated with Stroke

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Keywords:

Corrected Transposition of the Great Arteries; Inversion of the viscera; Patent Foramen Ovale; Stroke

Abbreviations:

LA: Left Atrium; RA: Right Atrium; LV: Left Ventricle; RV: Right Ventricle; AO: Aorta; PA: Main Pulmonary Artery; LPA: Left Pulmonary Artery; RPA: Right Pulmonary Artery; TR: Tricuspid Regurgitation; PFO: Patent Foramen Ovale; LAA: Left Atrial Appendage; TTE: Transthoracic Echocardiography.

1. Abstract

Corrected transposition of the great arteries is a rare complex congenital heart disease. Its clinical manifestations and changes in the natural history of the disease depend on its associated deformities. This article summarizes the diagnosis and treatment of a patient with IDD-type corrected transposition of the great arteries combined with intracardiac malformation sudden stroke. By summarizing the literature analysis, we hope to improve clinical understanding of the disease.

2. History of Presentation

Patient, male, 48 years old who was admitted to hospital for "recurrent episodes of palpitation that had lasted for a week". In the ward, ECG monitor was performed. Meanwhile beta blockers were administered orally to control the heart rate as well as furosemide and spironolactone to lighten the capacity load. In the next morning at 10:30 the patient felt paroxysmal palpitation. According to the data from the ECG monitor, paroxysmal atrial flutter was observed, and the heart rate was up to 171 beats/min. After about two minutes, the patient spontaneously converted to sinus rhythm. Then three hours later, the patient became unconscious, fell to the ground, had nausea and vomiting, was in a light coma, and was unable to answer.

3. Past Medical History

He denied any history of hypertension or diabetes.

4. Physical Examination

On admission: body temperature 36.6°C, pulse 78 times/min, respiration 17 times/min, blood pressure 100/72mmHg (1mmHg=0.1333kPa). The apex beat is located 0.5cm outside the fifth intercostal space on the right midclavicular line, with a heart rate of 78 beats/min, irregular rhythm, and 3-5 premature beats per minute. A grade 3/6 systolic murmur were heard in the third intercostal space on the right sternal border. When stroke occurred: The ECG monitor showed that the heart rate was 70-80 beats/min. Blood pressure was 105/60mmHg, oxygen saturation was 95-98%. Physical examination: orbital pressure reaction existed, bilateral pupils were equal, round, and 3mm in diameter, light reflex was sluggish, neck was soft; muscle strength (grade 0-5): muscle strength of left upper and lower limbs was grade 0, pathological signs were positive, the right upper and lower extremity muscle strength was grade 5, and the pathological signs were negative.

5. Investigations

1. Electrocardiogram (ECG): sinus rhythm, premature atrial beats, dextrocardia.

2. 24-hour Holter: Sinus rhythm, frequent premature atrial beats, part of the doublet, triplet, short paroxysmal atrial tachycardia, and occasional ventricular premature beats (Figure 1).

3. CT scan of chest and abdomen: (1)a small amount of pleural effusion on the left side. (2) dextrocardia, enlarged cardiac shadow, a small amount of pericardial effusion. (3) total inversion of the internal organs.

4. Transthoracic echocardiography (TTE): The liver located in the upper left abdomen, the heart located in the right thoracic cavity, the atrium was reversed, the ventricle was in the right loop, and the aorta and the pulmonary artery were arranged in the right front and left back. The aorta was connected to the anatomical right ventricle (functional left ventricle) and the pulmonary artery was connected to the anatomical left ventricle (functional right ventricle) (Figure 2). Left atrium and right atrium, functional left ventricle (anatomical right ventricle) enlarged, wall thickening, mild mitral regurgitation, and mild tricuspid regurgitation (Figure 3). Consider ccTGA (type IDD), a small amount of pericardial effusion.

5. Laboratory examination: Plasma D-dimer determination: 0.99mg/L, N-terminal pro-brain natriuretic peptide (NT-pro-BNP): 789.02pg/mL, other indicators were normal.

6. CT scan of brain after stroke showed infarction in the right frontotemporal parietal lobe and right basal ganglia. (Figure 4).

7.(1) Transesophageal echocardiography (TEE) after thrombectomy: patent foramen ovale. No obvious spontaneous imaging and definite signs of mural thrombus in the left atrium and left atrial appendage (Figure 5). (2) Transthoracic echocardiography with contrast echocardiography (cTTE): The foaming test was negative, and right-to-left shunt was not supported.

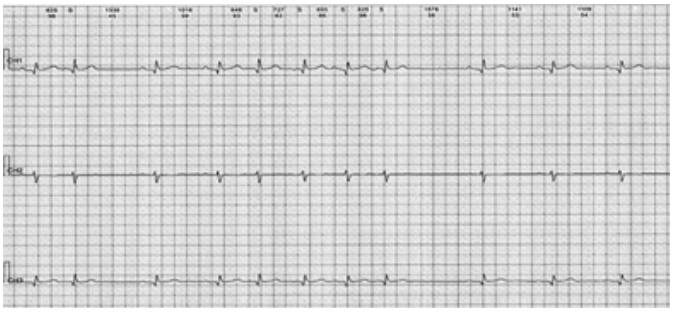

Figure 1: Atrial premature beat, atrial tachycardia captured by Holter monitor

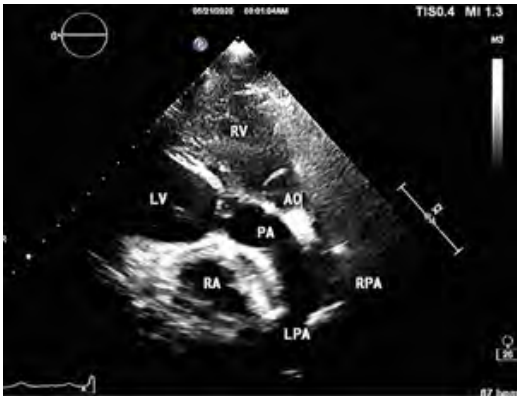

Figure 2: TTE: Atrial inversion, right loop of ventricle, aorta and pulmonary artery are arranged right anterior and left posterior, the aorta is connected to the anatomical right ventricle, and the pulmonary artery is connected to the anatomical left ventricle.

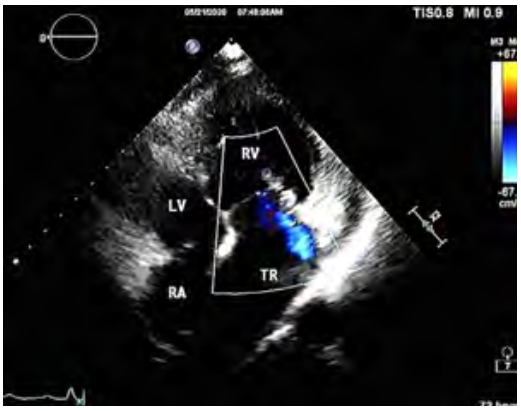

Figure 3: TTE: Small amount of regurgitation beam detected at tricuspid valve.

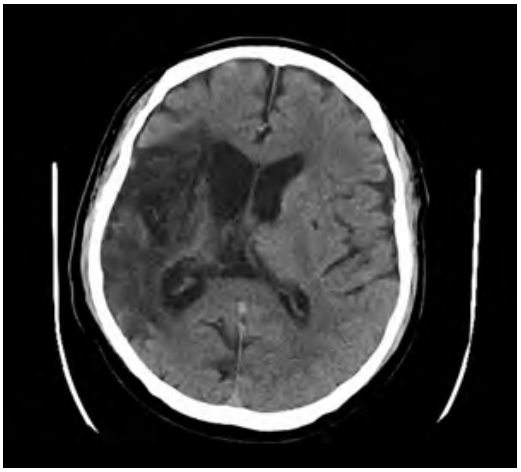

Figure 4: Brain CT showed cerebral infarction in right frontotemporal parietal lobe and right basal ganglia.

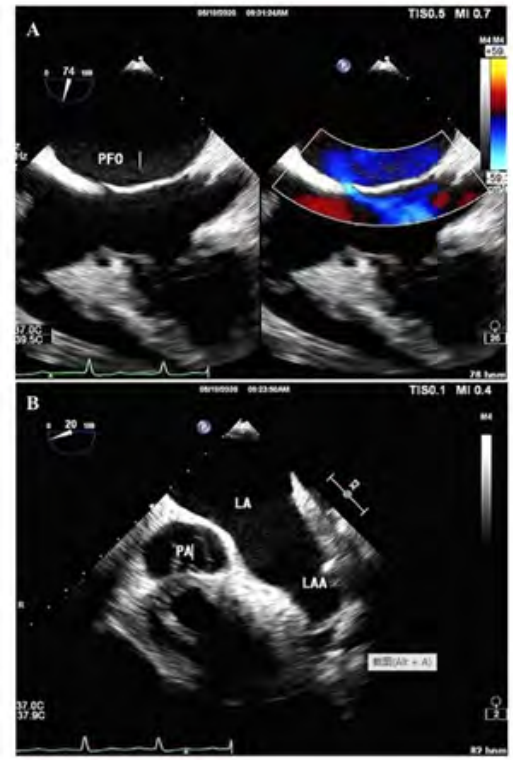

Figure 5: (A) TEE: Patent foramen ovale. (B) Spontaneous imaging and mural thrombosis were not seen in the left atrium and left atrial appendage

6. Differential Diagnosis

According to the medical history, three major causes for stroke shall be considered: 1. rupture of cerebral atherosclerotic plaque. 2. paradoxical embolism caused by PFO. 3. atrial thromboembolism sheds into the systemic circulation. Since the patient had no history of hypertension nor diabetes, and his cerebral angiography before thrombectomy showed that his entire head and neck blood vessels was smooth, the rupture of atherosclerotic plaque was not considered. As for this patient, although he had PFO intracardiac deformity, such defect was too small to cause hemodynamic changes. Therefore, he had normal growth and development before treatment, and had no clinical symptoms such as cyanosis of the lips and fingers. Although the patient had vomiting before stroke, which was a stimulus for paradoxical embolism, his cTTE test result was negative, and right-to-left shunt was not supported. This possibility could also be excluded. Thus, we have one last guess left, which is anatomical right ventricular insufficiency. The patient's echocardiography showed enlargement of the bi-atrial and functional left ventricle (anatomical right ventricle), mild tricuspid regurgitation. His chest and abdomen CT scan showed a small amount of pleural and pericardial effusion, and his NT-pro-BNP had already elevated. All these evidences indicated that the patient had such symptom. His dynamic electrocardiogram already captured the onset of short bursts of atrial tachycardia and atrial flutter, and his plasma D-dimer was elevated. Take these into consideration, we speculated that the patient most likely had thrombosis in the left atrium. After the atrial flutter was terminated and converted to sinus rhythm, Atrial contractility gradually increased, leading to thrombus shedding and stroke.

7. Management

In the emergency department, the patient was admitted to the DSA room for "cerebral angiography and mechanical thrombectomy" treatment. The patient recovered well after the operation and showed stable vital signs.

8. Discussion

Corrected transposition of the great arteries (congenitally corrected transposition of the great arteries, ccTGA) accounts for about 0.4%-1% of all congenital heart diseases [1-2]. It refers to the abnormal curvature of the original heart tube to the left during embryonic development. In this case, the anatomical left ventricle is connected to the right atrium to send out the pulmonary artery and acts as a functional right ventricle to undertake pulmonary circulation, while the anatomical right ventricle is connected to the left atrium to give off the aorta and acts as a functional left ventricle to undertake the systemic circulation [3]. Although the atrium-ventricle and ventricle-aorta connections of the patient were inconsistent, the hypoxic blood still entered the lungs through the pulmonary artery, and was transported to the systemic circulation by the aorta, thereby obtaining a "physiological correction" in the blood circulation [4]. This disease is often accompanied by abnormal visceral position. Studies have shown that 80%-95% of CTGA patients have orthotopic viscera, and 5%-20% have visceral inversion [5]. In CTGA patients, approximately 90% of patients have intracardiac malformations, such as atrioventricular septal defect, pulmonary stenosis, patent ductus arteriosus, patent foramen ovale, abnormal or insufficiency of atrioventricular valve, and so on [6,7]. It has also been reported that ccTGA without other malformations might be asymptomatic, but 25% of patients in age group between 40 and 50 years old have arrhythmia or morphological tricuspid regurgitation and right ventricular insufficiency concern during their doctor's appointments [8]. And anatomical right ventricle and tricuspid valve are not suitable for long-term burden of the systemic pressure load on the functional left ventricle and mitral valve. Septal deviation can be caused under long term elevated right ventricular pressure load [9]. As a result, the tricuspid valve annulus will gradually expand, resulting in insufficiency of the morphological tricuspid valve, followed by progressive enlargement of the left atrium and right ventricle, gradual fibrosis of the atrial muscle, reduced coupling between the myocardium. As a result, atrioventricular conduction delay or block will occur, and eventually atrioventricular reentry will occur, resulting in arrhythmias such as atrial flutter and atrial fibrillation. These arrhythmias gradually lead to congestive heart failure, which will eventually shorten the patient's life expectancy [10]. Morphological tricuspid regurgitation and right ventricular insufficiency are mutually causal. Morphological tricuspid regurgitation has a greater impact on the prognosis of ccTGA patients and which is one of the pri-

mary causes of poor prognosis [11]. Therefore, the long-term and dynamic observation on the degree of tricuspid regurgitation and the functional changes of the right ventricle by echocardiography are very important for the prognosis and life quality of the patients.

In terms of treatment, in addition to symptomatic treatment, CTGA patients currently have two types of surgical treatments [12]: One is physiological repair, including ventricular defect repair, left ventricular outflow tract obstruction dredging and tricuspid valve repair or replacement. The other is anatomical repair, whose purpose is to make the blood of the systemic vein flow back into the right atrium, reach the pulmonary circulation through the dissection of the right ventricle, and to make the blood of the pulmonary vein flow into the left atrium, through the dissection of the left ventricle to the systemic circulation. By this way, it reduces the incidences of tricuspid valve insufficiency and the anatomic right ventricular failure. Nevertheless, anatomical repair is complex and difficult, and there are many complications in patients too. This type of surgical treatment still requires more experiences. For patients with CTGA and inversion of the heart with anatomical right ventricular failure and malignant arrhythmia, biventricular pacing and implantable defibrillator therapy could also be a good choice [13].

9. Conclusion

In conclusion, for ccTGA patients, two aspects of situations need to be clarified [14]. On one hand, the positions and connections of the internal organs, atria, ventricles, and aortas, as well as intracardiac malformations should be clarified. If there is no concomitant intracardiac deformity, the hemodynamics are completely normal and no treatment is required; if there is an intracardiac malformation, surgical correction is required, so it is more important to find the concomitant deformity. On the other hand, cardiac structure, function, valve damage and regurgitation, and hemodynamics should be assessed. Proper usage of echocardiography could help to observing the shape of the tricuspid valve, the degree of disease and the size of the right ventricle, and assess the regurgitation and cardiac function changes accurately, and it also provide a comprehensive and important basis for clinical treatment planning surgical timing determination and prognosis evaluation.

10. Author's Statement

We confirmed that permission was granted by the subject patient to publish the case report.

11. Authorship

The authors confirm contribution to the paper as follows: study conception and design: Caixia Yin; data collection: Mengzhu Fu; analysis and interpretation of results: Xing Li. All authors reviewed the results and approved the final version of the manuscript.

12. Conflicts of Interest

The authors declare that they have no conflicts of interest to report regarding the present study.

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