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Dextrocardia with Cor Biloculare in a 44 Year Old Women

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Abstract:

Cor biloculare, a two-chambered heart, is a rare congenital anomaly of the heart. It's characterized by a single or common atrium communicating with a single or common ventricle by a single atrio-ventricular valve. Most patients with cor biloculare die within neonatal period or even in fetal period, and very few survivors could grow up to adult. In this article, we report an adult case with dextrocardia and cor biloculare, who survived to the 44 years of life.

Key words: Dextrocardia, Echocardiography, Heart, Heart Ventricles, Humans.

Introduction

The cor biloculare is a rare congenital anomaly of the heart, which is characterized by a single or common atrium communicating with a single or common ventricle by a single atrioventricular valve. Hitoko OGATA has reported a 42-year-old female who is the oldest survivor with cor biloculare [1]. However, there is no case report about dextrocardia with cor biloculare in adult. In this report we present the case of patient with dextrocardia and cor biloculare who survived to the 44 years of life.

Case Report

A 44 year old female patient was admitted to our hospital complaining of history of cyanosis since birth, and increasing dyspnea for past 7 days. She was diagnosed with "congenital heart disease" at birth for which she had not received any medical or surgical treatment. Her mental and physical development was normal, but her exercise tolerance was a grade 1/4 of the NYHA classification. During the past 7 days, she has been experiencing dyspnea, cough and expectoration, which she attributed to a cold. She denied fever, hemoptysis and orthopnea. She was the second child of healthy parents with no history of heart disease in her two siblings or family members. The mother's pregnancy was normal with a spontaneous vertex delivery.

On physical examination, her temperature was 36.2^{0}C, with a heart rate of 59 bpm, blood pressure of 111/54 mmHg, and her respiratory rate was 19 breaths per minute. There was pronounced clubbing of the fingers and toes with purple coloration of skin and mucus membrane of head and extremities and had distension of neck veins. Edema was

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absent in feet, hand or face. The respiratory system examination revealed moist rales in lung bases. Cardiovascular system examination was suggestive of maximum intensity of apex beat located in the right fifth intercostal space in the midclavicular line with regular rate and rhythm, without rub and or gallop. There was IV/VI systolic ejection murmur heard best in the apex. Abdominal organs were essentially normal to palpation.

The electrocardiography (ECG) appeared to show dextrocardia, and this was confirmed by chest X ray. In her ECG, P waves in leads I, II, III, avF were inverted, and QRS complexes were shown as qR, rs and rS in leads avF, avL and avR, respectively [Fig.1]. Chest X ray was read as dextrocardia with enlarged cardiac silhouette, and pulmonary infiltrates [Fig.2].

Echocardiography showed dextrocardia, single atrium and single ventricle, with stenosis in pulmonary artery and pulmonary valve, and mild regurgitation of atrium-ventricle valve [Fig.3,4]. The aorta arched abnormally to the right. The diameters of pulmonary truck, left and right pulmonary arteries were 19 mm, 11 mm and 12 mm respectively.

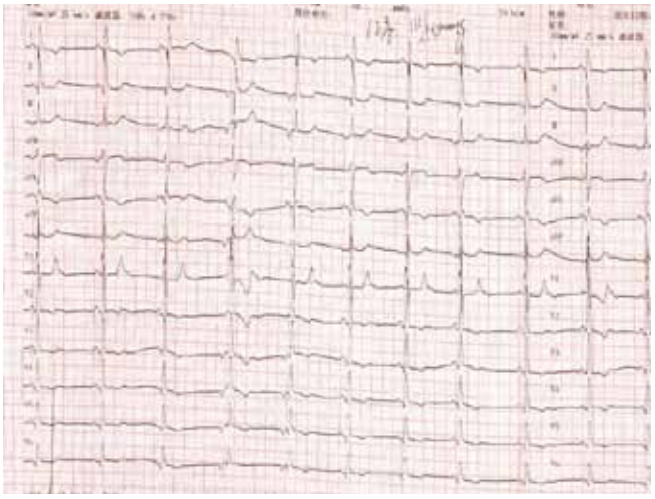

Fig.1: ECG: P waves in leads I, II, III, avF were inverted, and QRS complexes were shown as qR, rs and rS in leads avF, avL and avR, respectively.

Complete blood count showed WBC: 6.45×10^{9}/L, NE: 62.8%, LY: 27.9%, RBC 6.45×10^{12}/L, Hb 205 g/L. Arterial blood gas analysis: pH 7.405, PO_2 43.3 mmHg, PCO_2 31.7 mmHg, HCT 51.8%, SO_2 78.0%. Urinalysis, renal function, and serum lipids were normal on admission.

She was diagnosed as congenital heart disease (dextrocardia and cor biloculare), pneumonia, and secondary polycythemia. After anti-infection therapies for 13 days, she was discharged with stable conditions and was alive during 6 month follow-up.

Discussion

Cor biloculare, a two-chambered heart is probably the rarest congenital heart disease [2]. Most babies with cor biloculare die within weeks after birth. We reported a case of dextrocardia with cor biloculare surviving to the 44 years of life, whose survival time is the longest currently.

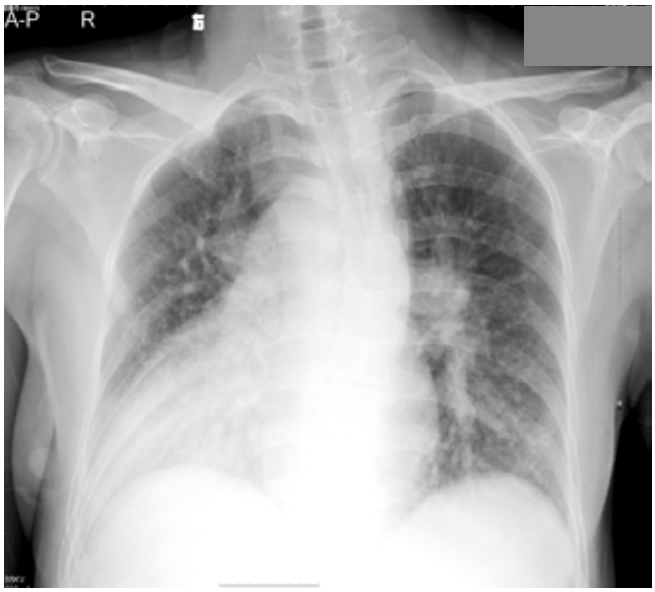

Fig.2: Chest X ray: dextrocardia with enlarged cardiac silhouette, and pulmonary infiltrates.

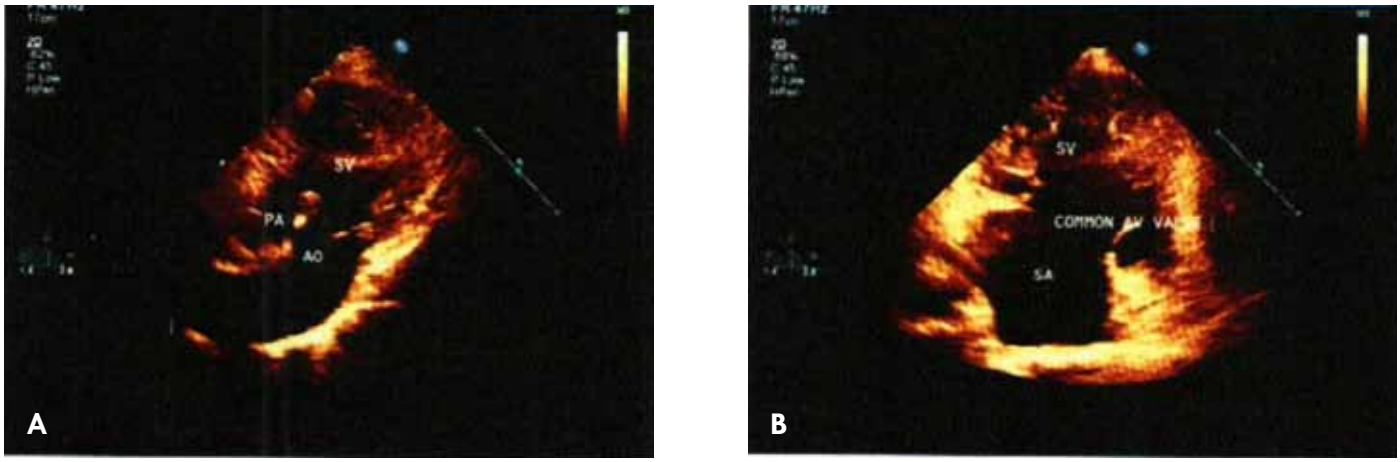

Fig.3: Echocardiography: dextrocardia, single atrium and single ventricle, with stenosis in pulmonary artery and pulmonary valve, and mild regurgitation of atrium-ventricle valve.

Cor biloculare is a two-chambered heart with one atrium and one ventricle, and a common atrioventricular valve, due to failure of formation of the interatrial and interventricular septa. In 1950, Brown divided the condition into three groups: (i) with an undivided truncus arteriosus, (ii) with normal division into aorta and pulmonary artery, and (iii) incomplete forms with some septal formation but a persistent single A-V valve [2]. Our case appears to belong to the second group. Cor biloculare is usually classified with the cyanotic group of congenital heart diseases. Blood enters the atrium from both systemic and pulmonary venous, and passes through a single atrioventricular valve into an enlarged single ventricle. The severity of cyanosis is dependent on the both diameters and pressures in aorta and pulmonary arteries. Echocardiography was suggestive of stenosis in pulmonary artery and pulmonary valve, which may increase the afterload of pulmonary circulation and decrease pulmonary blood volume. The hemodynamic effects could lead to the low level of oxygen in systemic circulation, and early occurrence of cyanosis.

Cor biloculare hardly ever occurs without other anomalies being present. It is usually associated with various other malformations such as dextrocardia.

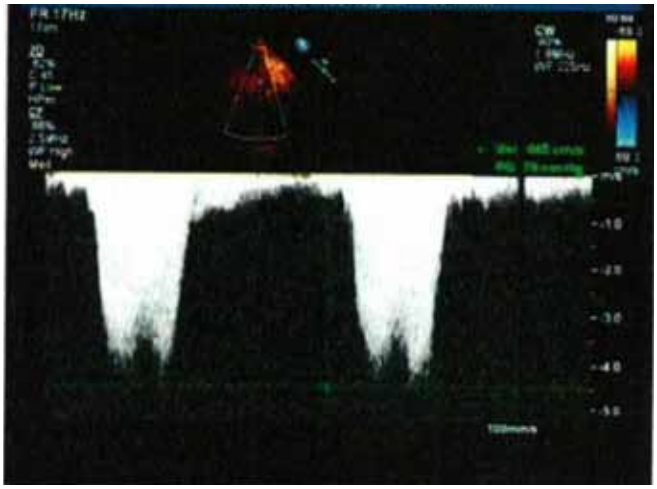

Fig.4: Doppler ultrasound detection.

Medical diagnosis of the two forms of congenital dextrocardia can be made by ECG or X-ray imaging. Dextrocardia is a congenital defect in which the heart is situated on the right side of the body. It is believed to occur in approximately 1 in 12,000 people [3]. There are two main types of dextrocardia: dextrocardia of embryonic arrest (also known as isolated dextrocardia) and dextrocardia situs inversus. Dextrocardia situs inversus is further divided into dextrocardia situs inversus solitus and dextrocardia situs inversus

totalis. The feature of X-ray image in this case is mirror-image dextrocardia. However, we didn't have further examinations of abdomen ultrasound to confirm whether it is a dextrocardia situs inversus or not.

The most common differential diagnosis of cor biloculare include tetralogy of fallot, tricuspid atresia and complete atrio-ventricular canal defect [4]. Echocardiography is a diagnostic test that uses ultrasound waves to create an image of the heart muscle. In fact, it is one of the most widely used diagnostic tests for congenital heart disease. This case was diagnosed by echocardiography, but not cardiac angiography. We conclude that routine obstetric ultrasound examinations and detailed fetal echocardiography focusing on four-chamber view of heart may be a useful screening modality for the presence of congenital heart disease.

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