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Case Report

Mitral and Tricuspid Valve Replacement in Uncommon Case of Situs Inversus with Dextrocardia

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Abstract

Background: **Incidence of congenital cardiac anomalies in dextrocardia with situs inversus combined with severe rheumatic mitral valve stenosis (MS) and tricuspid regurgitation is uncommon. Also, anatomic malposition can cause hindrance for surgeon while handling cannulation and diseased valves.**

Objective: **We reported a case of situs inversus and dextrocardia with rheumatic mitral stenosis and severe tricuspid regurgitation. Details of diagnosis, treatment and some technical operative techniques to cope with this condition were described and the literature review was performed.**

Case report: **The patient was a 48-year-old woman with a history of cardiomegaly, progressive dyspnea, easy fatigability, paroxysmal nocturnaldyspnea, anasarca, marked jaundice and had the New York Heart Association (NYHA) Class 3 with underlying hemoglobin H disease. She underwent mitral and tricuspid valve replacement due to severe rheumatic mitral stenosis and tricuspid regurgitation. She was improved to NYHA Class 1 within two months after the operation.**

Conclusions: **Based on this uncommon patient presenting with dextrocardia, the recommended primary operative approach is performed with the surgeon standing at the left side of the patient. A transeptal-atriotomy provided an excellent exposure without heart positioner.**

Keywords: **dextrocardia, hemoglobin H, rheumatic heart disease, situs inversus**

Introduction

Dextrocardia with situs inversus is an uncommon congenital heart defects, but the association of this condition with acquired valvular heart disease is even rarer. Herein, we reported a case of middle-aged Thai woman with dextrocardia, situs inversus, acquired rheumatic mitral stenosis and tricuspid valve regurgitation plus Hb H disease.

Case Report

A 48-year-old woman presented with a history of progressive dyspnea and palpitation on exertion, cardiomegaly, cardiac murmur, atrial fibrillation, jaundice, hepato-splenomegaly and pitting edema at both legs of 5 months. Physical examination revealed jaundice and edema with blood pressure of 110/70 mm Hg and atrial fibrillation at a rate of 78/min. The

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positive physical findings were marked jaundice, moderately pale conjunctiva and jugular venous distension. The point of maximal impulse was at the fifth right intercostal space one centimeter away from midclavicular line. The first heart sound was moderately accentuated as was the pulmonary component of the second heart sound. An opening snap was heard over the entire precordium plus a grade 3/6 diastolic rumbling murmur at the apex radiated to her axillar with grade 3/6 holosystolic murmur at the left parasternal border. The chest film revealed dextrocardia and situs inversus, cardiothoracic ratio of 0.81 and increased pulmonary vascular markings (Figure 1)

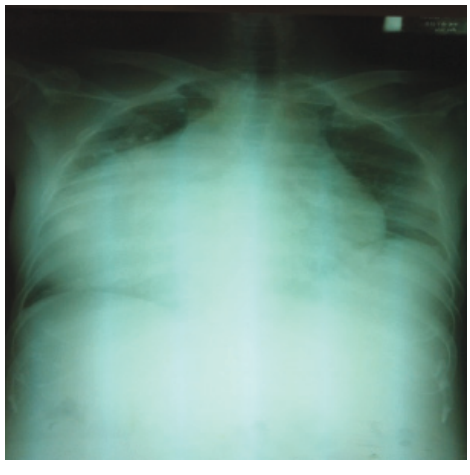

Figure 1 Noted dextrocardia, marked cardiomegaly with prominence of pulmonary vascularity

Routine laboratory studies showed mild to moderate degree of anemia from Hb H disease and cholestasis jaundice which was confirmed by abdominal ultrasonography. There were dextro position of abdominal organs, mild hepatomegaly with congestion of hepatic vein and inferior vena cava and no liver parenchymal disease. Biliary trees, spleen, pancreas and kidneys appeared normal and large amount of ascites was noted.

Two-D echocardiography showed dextrocardia, LAE, RAE, RVH and D-shape LV. Overall LV systolic functions were normal with an EF = 67%. The aortic valve was tri-leaflet and appeared structurally normal. MV mobility +2 valve thickening +3 subvalvular fusion +2 calcification+3 to +4 and sever MS MVA = 0.79 cm2by trace, MVA = 1.07cm2 by PHT , MPG = 6.11 mmHg. RAE, RVH with severe TR, RVSP = 40.02 mmHg. Pericardium was normal with minimal pericardial effusion.

The patient underwent open heart surgery through standard median sternotomy, cardiopulmonary bypass was established by cannulating ascending aorta and SVC/IVC, myocardial preservation using cold blood antegrade cardioplegia (Figure 3).

Mitral valve was replaced using C-E PERIMOUNT Plus Pericardial Bioprosth size 27 mm and tricuspid valve was replaced using C-E PERIMOUNT Plus Pericardial Bioprosth. Mitral size 25 mm subsequently. Rheumatic tricuspid valve was shown in Figure 4.

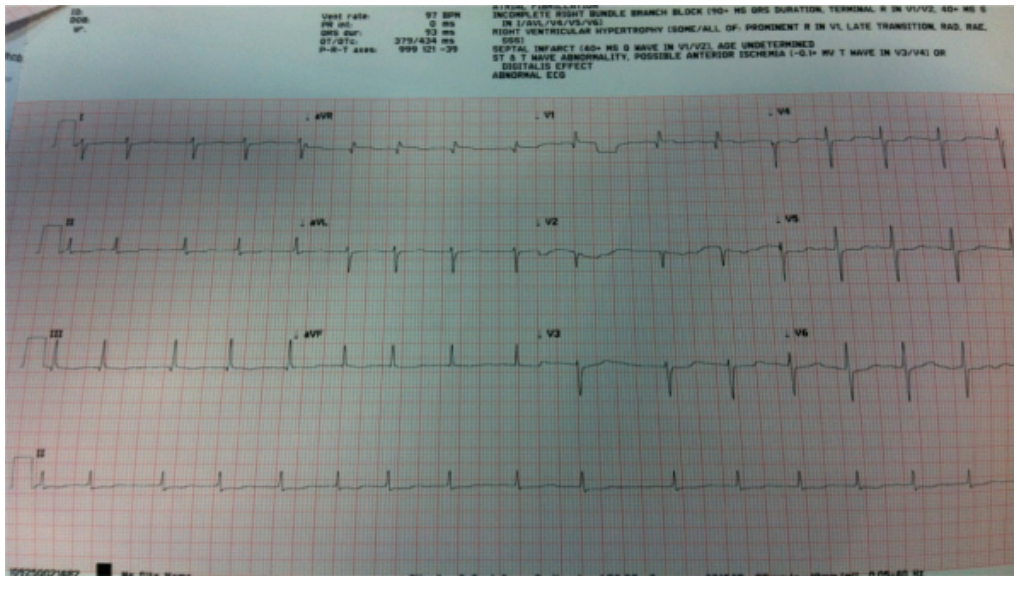

Figure 2 The electrocardiogram was consistent with dextrocardia and also showed atrial fibrillation, a mean QRS axis of +120 degree

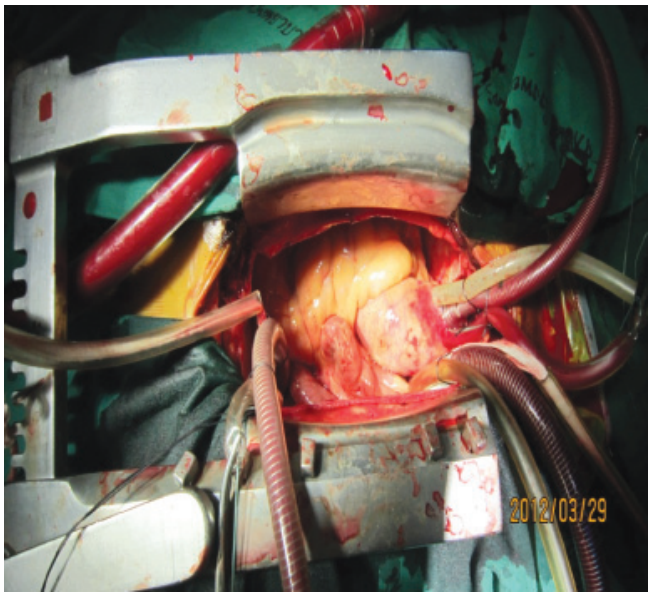

Figure 3 CPB Circuit was shown and the surgeon standing on the left sided

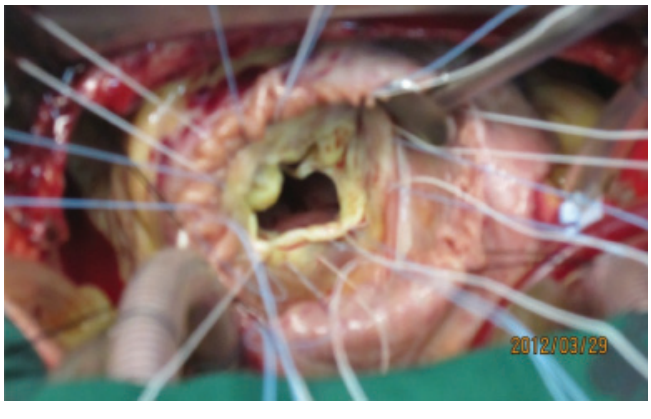

Figure 4 Severe rheumatic TR due to thickened, retracted all leaflets plus large cleft between anterior and posterior leaflet

The patient recovered uneventfully. She was extubated within 3 hours after the operation and stayed in ICU less than 24 hours. Her total hospital stay was 10 days. Until her last follow-up at 30 months, she was still leading her normal life.

Discussion

Dextrocardia is a cardiac positional anomaly in which the heart is located in the right hemithorax with its base-to-apex axis directed to the right and caudad. The malposition is intrinsic to the heart and not caused by extracardiac abnormalities. Dextrocardia should be differentiated from cardiac dextroposition, which is defined as displacement of the heart to the right secondary to extracardiac causes such as right lung hypoplasia, right pneumonectomy, or diaphragmatic hernia[1]. A number of congenital heart defects have been reported with dextrocardia, including ventricular septal defect, patent ductus arteriosus, atrial septal defect (secundum and primum), tetralogy of Fallot, pentalogy of Fallot, infundubular pulmonary stenosis, transposition and pseudotruncus, corbiloculare, atrial-ventricular canal with patent ductusarteriosus, and total anomalous pulmonary venous return[2,3]. Multiple skeletal defects have also been reported in association with dextrocardia, including multiple deformities of the spine and skull asymmetry[4].

Total situs inversus is uncommon, its incidence being one per 12,019 births[5], and the association of acquired valvular lesions with dextrocardia and situs inversus is unknown but must be very rare. The first case of dextrocardia and acquired valvular heart disease was reported by Owen in 1911; the electrocardiogram in this case was interpreted by Dr. Thomas Lewis[6]. Precise anatomical diagnosis is essential for successful surgery based on two-dimensional echocardiographic studies by using standard criteria for diagnosis of situs, while cardiac catheterization was used to confirm cardiac anatomy in few cases[7,8]. Open-heart in patients with a cardiac position anomaly is technically demanding, and surgeons are required to make certain modifications to their normal surgical techniques when operating on patients with dextrocardia. Some authors have recommended standing on the right side as usual while using a Starfish™ heart positioner to provide adequate exposure to the posteriorly placed right atrium, making venous cannulation relatively easy and safe[9]. In this case, we stood on the left side of the operating table, which provided excellent exposure and made the procedure relatively easy.

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