Case Report

COR TRIATRIATUM ASSOCIATED WITH EBSTEIN MALFORMATION OF ATRETIC MITRAL VALVE AND DOUBLE OUTLET RIGHT VENTRICLE

Funda Öztunç, M.D.* / Figen Akalın, M.D. / Resmiye Beşikçi, M.D.*****

* *Institute of Cardiology, Istanbul University, Istanbul, Turkey.*
** *Sub-department of Pediatric Cardiology, Department of Pediatrics, School of Medicine, Marmara University, Istanbul, Turkey.*
*** *Clinic of Pediatric Cardiology, Bakirköy Maternity Hospital, Istanbul, Turkey.*

ABSTRACT

Cor triatriatum is a rare congenital cardiac anomaly and association of this pathology with Ebstein's malformation of atretic mitral valve and double outlet right ventricle has not been previously reported. We present an 11-day-old infant with this unique anomaly diagnosed by echocardiographic examination. The infant died before surgical intervention.

Key Words: Cor triatriatum, Ebstein anomaly, Mitral valve atresia, Double outlet right ventricle

INTRODUCTION

Cor triatriatum is an uncommon congenital cardiac anomaly. Ebstein's malformation of the mitral valve is also very rare. We describe herein a unique case with cor triatriatum sinistrum associated with Ebstein's anomaly of atretic mitral valve and double outlet right ventricle.

CASE REPORT

An 11-day-old male baby was referred to our hospital for cyanosis and heart murmur. He was a term baby with 3000g birth weight and born to a healthy mother following an uneventful pregnancy. His parents were consanguineous but the family history was negative for congenital heart disease. On physical examination, he was tachypneic and cyanotic. Heart rate was 130 beats/minute. Femoral pulses were weak. Blood pressure measured on the right arm was 80 mm Hg. An ejection murmur of 3/6rd grade was audible at the left sternal border. The liver was palpable 3 cm below the costal margin. The chest x-ray showed en enlarged cardiac silhouette and increased pulmonary vascular markings. Right axis deviation and significant right ventricular hypertrophy was present on the electrocardiogram. Echocardiographic examination revealed that the apex was located on the left hemithorax; atrial situs was solitus; pulmonary and systemic venous connections were normal; a large secundum type atrial septal defect was present. The direction of the shunt was from left atrium to right atrium. Tricuspid valve had a normal anatomy; right ventricular pressure estimation using the mild regurgitation of the tricuspid valve was at systemic level. There was no true mitral valve. A membrane-like structure was present below the left atrium. At the septal edge of the membrane an orifice

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measured 3 mm in diameter was present. CW Doppler showed bi-directional shunt with maximum velocity of 1 m/s through this orifice. The chamber below this membrane is thought to be an atrialized chamber because the posterior wall of this chamber was very thin, and there was no true gradient between this chamber and the left atrium. This chamber was separated from the hypoplastic ventricle with thick muscular tissue. (Figs. 1,2) This tissue was located more than 2 cm apical than tricuspid valve. Left circumflex coronary artery passed through this region. It is thought that the muscular partition supported the displaced and atretic leaflet of the mitral valve and divided the left ventricle. Apical component of the left ventricle was in free communication with the large right ventricle through a subpulmonary ventricular septal defect. In addition to the large subpulmonic ventricular septal defect there was multiple small trabecular defects between the two ventricles. The great arteries originated from the large right ventricle. The pulmonary artery was enlarged. The origin of the right pulmonary artery was mildly stenotic. A gradient of 22 mmHg was measured between the main and right pulmonary artery. Patent ductus arteriosus was present. Aortic isthmus was narrow. Arcus aorta was measured 6.5 mm, and isthmus was measured 4 mm. A gradient of 26 mm Hg was measured in the descending aorta.

Pulmonary banding and relieving the aortic coarctation was planned for the patient, but the family did not contact us after the first visit. We acknowledged that he had died 6 weeks after the initial examination.

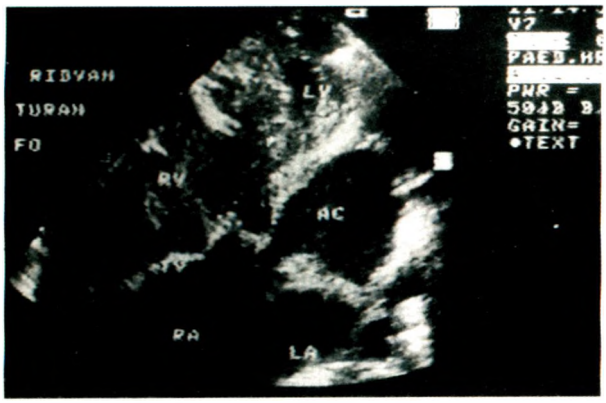

Fig.1: Subcostal view demonstrating the pulmonary venous chamber, atrialized chamber, apically displaced atretic mitral valve and ventricular septal defect.

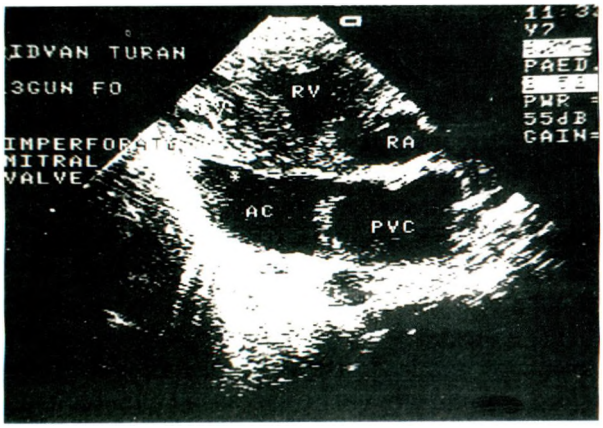

Fig.2: Parasternal long axis view showing the membrane in the place of mitral valve between pulmonary venous chamber and atrialized chamber.

DISCUSSION

The term cor triatriatum sinistrum is used for the anomaly in which the left atrium is subdivided into two chambers by a membrane. It occurs in only 0.1% of children with congenital heart disease (1). Although the mechanisms causing the development of this rare cardiac anomaly are not clear, Van Praag has suggested the reason was malincorporation of pulmonary venous and left atrial chambers rather than malseptation of the left atrium (2).

Thilenius et al (3) has described three morhologic subtypes of cor triatriatum sinistrum. The first type is the classical form in which the pulmonary veins drain into the proximal chamber. The second type also can be considered as a type of total anomalous pulmonary venous return with pulmonary venous drainage into the coronary sinus. In the third type there is a medially located chamber between the two atria and the pulmonary venous drainage is normal. Our case in compatible with the first subtype.

Various types of complex congenital cardiac anomalies are found to be associated with cor triatriatum which indicates a profound and diffuse disturbance in the development of the fetal heart. Partial and total anomalous pulmonary venous drainage, left superior vena cava, atrial septal defect, ventricular septal defect, patent ductus arteriosus, coarctation of the aorta,

atrioventricular canal, tetralogy of Fallot, double outlet right ventricle, single ventricle, mitral atresia, aortic atresia, Ebstein's anomaly of the tricuspid valve are reported as associated cardiovascular defects with cor triatriatum (4-7).

One of the type C cases described by Thilenius et al has similar associated anomalies such as double outlet right ventricle and mitral atresia but the type of atrial septation is different and Ebstein anomaly of the atretic mitral valve was not present in that case (3).

Nine cases of Ebstein's malformation of the mitral valve have been described previously (8). Three of them were associated with aortic obstruction: aortic atresia, hypoplasia, and coarctation. In three cases the tricuspid valve was also affected by Ebstein's malformation. In one case the lesion was associated with double outlet right ventricle and a subaortic ventricular septal defect (8). There was no case with cor triatriatum and Ebstein's malformation of the mitral valve.

Ebstein's malformation of the tricuspid valve with atresia, and right atrioventricular orifice atresia has been described before (9,10).

We found an interesting case of Gerlis and Anderson (11) with a diagnosis of cor triatriatum dexter with imperforate Ebstein's anomaly of the tricuspid valve. It was very similar to our case but in that case the right side of the heart was involved.

Since no other similar case combining cor triatriatum sinistrum, Ebstein's anomaly of the atretic mitral valve and double outlet right ventricle was found in our survey of the literature, this case is supposed to be a unique one. Unfortunately pathologic postmortem examination of the heart was not possible because the patient died out of hospital, but echocardiographic findings were descriptive.

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