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Original Research Paper

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COR-TRIATRIUM DEXTER, ATRIAL SEPTAL DEFECT AND PARTIAL ANOMALOUS PULMONARY VENOUS CONNECTION IN A 35-YEAR-OLD WOMAN

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ABSTRACT Cor triatriatum dexter is a rare congenital heart anomaly in which a membrane divides the right atrium into 2 chambers. It exists either in isolated classical form or may be associated with simple to complex congenital cardiac anomalies. We report the case of a 35-year-old female who had cor triatriatum dexter, partial anomalous pulmonary venous connection and a large atrial septal defect. In patients with concomitant heart anomalies, open-heart surgery remains the gold standard in the treatment of cor triatriatum dexter.

KEYWORDS : Cor triatrium dexter, sinus venosus atrial septal defect, partial anomalous pulmonary venous connection

INTRODUCTION

Cor triatriatum dexter (CTD), or partitioning of the right atrium (RA) to form a triatrial heart, is an extremely rare congenital anomaly which is caused by the persistence of the right valve of the sinus venosus. The incidence of cor triatriatum is approximately 0.1% of congenital heart diseases. The right atrial partition is due to exaggerated fetal eustachian and thebesian valves, which together form an incomplete septum across the lower part of the atrium. This septum may range from a reticulum to a substantial sheet of tissue [1]. We present a case of CTD in a young adult who underwent successful surgical excision of the membrane and closure of sinus venosus atrial septal defect with IVC blood directed to RA.

Case Report

A 35-year old female presented with complaints of shortness of breath and atypical chest pain restricting her daily activities for 12 months. On examination, her second heart sound was wide and fixed split and an ejection systolic murmur was heard in the pulmonary area.

Further evaluation by a trans-thoracic echocardiography (TTE) showed a membrane in the right atrium with ostium secundum atrial septal defect, dilated RA and right ventricle (RV) and normal biventricular function. Trans-esophageal echocardiography (TEE) showed similar findings. CT cardiac showed cor triatrium dexter with membrane running from the level of thebesian valve of coronary sinus and eustachian valve of inferior vena cava (IVC) inferiorly upto the lower part of crista terminalis laterally, the right upper and lower pulmonary veins draining into right atrium, large ostium secundum atrial septal defect and a small sinus venosus atrial septal defect. (Figure 1 a,b)

After routine pre-operative evaluation patient was taken for surgery. The approach was through a median sternotomy. CPB was established with aortic and bicaval cannulation. After clamping aorta, root cardioplegia was given and the patient was cooled to 28°Celsius. The intra-operative findings were similar to the CT findings with the IVC blood draining into left atrium (LA).(Figure 2 a)

We resected the membrane (Figure 2 b), closed the ostium secundum and sinus venosus atrial septal defect (ASD) with baffling of right upper and lower pulmonary veins into LA using autologous untreated pericardial patch.(Figure 2 c).

RESULTS

Post-operatively patient was hemodynamically stable and was discharged on post-operative day 5. Trans-thoracic echocardiography at discharge showed no residual membrane, ASD patch intact and all pulmonary veins draining into LA, IVC draining into RA and normal biventricular function.

DISCUSSION

Cor triatriatum sinister (CTS) is an uncommon congenital cardiac anomaly first described by Church in 1868, and the name was given by Borst in 1905. Cor triatriatum dexter (CTD) is rarer than the CTS and was first described by Rokitansky in 1875 [2].

It is generally believed that CTD results from persistence of the right valve of the sinus venosus. During embryogenesis, the right horn of the sinus venosus gradually incorporates into the right atrium to form the smooth posterior portion of the right atrium, whereas the original embryologic right atrium forms the trabeculated anterior portion. The connection between the right horn of the sinus venosus and the embryologic right atrium is the sinoatrial orifice, which is flanked on either side by two valvular folds, the right and left venous valves. At some point during this incorporation, the right valve of the right horn of the sinus venosus divides the right atrium in two. This right valve forms a sheet that serves to direct the oxygenated venous return from the inferior vena cava across the foramen ovale to the left side of the heart during fetal life. Normally, the valve regresses by approximately at 12-weeks of gestation and leaves behind the crista terminalis superiorly and the eustachian valve of the inferior vena cava and the thebesian valve of the coronary sinus inferiorly [3].

CTD is frequently associated with right-side defects caused by abnormal fetal circulation. Among these, the most frequent are stenosis or atresia of the pulmonary valve, tricuspid valve abnormalities andAtrial septal defect [4].One or more pulmonary veins may have an anomalous drainage pattern leading to the right atrium, superior vena cava, or the distal chamber of the left atrium [2].

Cor triatriatum sinister remains an uncommon form of congenital heart disease although it is being diagnosed with increasing frequency in adultrarely associated with mitral pathology which requires high suspicion for diagnosis. [5]

The clinical presentation of CTD depends on the degree of right atrial septation and the size of the sinoatrial orifice. Cor triatriatum dexter can present in isolation or as part of complex right-sided heart defects. Right ventricular inflow obstruction—and, in cases of associated ASD, signs of right-to-left shunting—can be so subtle that diagnosis is difficult and might even be missed during cardiac surgery for other conditions [6]. Symptomatic patients should undergo

correction of the underlying abnormalities. Surgical resection of the membrane is the treatment of choice for symptomatic patients with significant obstruction, although percutaneous catheter-based disruption of the membrane has been used as an alternative [7].

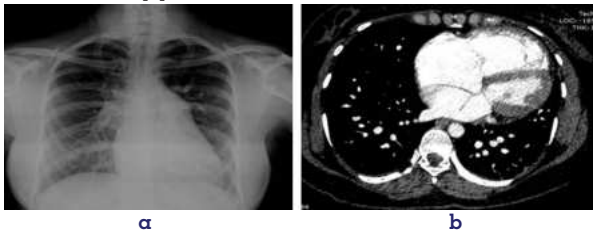

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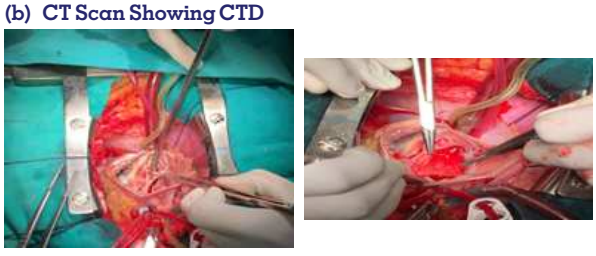
Figure 1
(a) Pre-operative Chest Roentgenogram
(b) CT Scan Showing CTD



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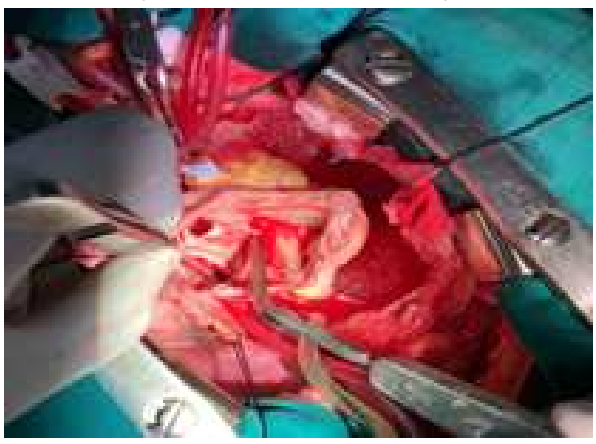

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Figure 2
(a) CTD Membrane Separating RA into Two chambers
(b) Excision of the Membrane
(c) Closure of ASD with Autologous Untreated Pericardial Patch

CONCLUSION

Cor triatrium dexter is rare in adults. Preoperative imaging with TTE, TEE, and CT scan is essential for making correct therapeutic decisions. In patients with concomitant heart anomalies, open-heart surgery remains the gold standard in the treatment of cor triatriatum dexter.

Abbreviations

CTD: Cor triatrium dexter
RA: Right atrium
CT: Computed tomography
RV: Right ventricle
IVC: Inferior vena cava
LA: Left atrium
CPB: Cardio-pulmonary bypass
ASD: Atrial septal defect
TTE: trans-thoracic echocardiography
TEE: trans-esophageal echocardiography

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