Cor triatriatum dexter: A rare cause of childhood cyanosis

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ABSTRACT

Cor triatriatum dexter is a rare congenital heart anomaly where the right atrium is divided into two chambers by a membrane. We report a boy who had persistent mild cyanosis and diagnosed to have cor triatriatum dexter with secundum atrial septal defect by transoesophageal echocardiography. Interestingly, he had persistent mild cyanosis despite insignificant obstruction to the right ventricular inflow and normal pulmonary artery pressure. The pathophysiology, approach to the diagnosis, and mode of treatment are also discussed.

Keywords: Atrial septal defect, cor triatriatum dexter, cyanosis, transoesophageal echocardiography

INTRODUCTION

Cor triatriatum dexter, or partitioning of the right atrium (RA) to form a triatrial heart, is an extremely rare congenital anomaly that is caused by the persistence of the right valve of the sinus venosus.[1] The incidence of cor triatriatum is approximately 0.1% of congenital heart malformation.[2] Typically, 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]

CASE REPORT

A 9-year-old boy was admitted to a local district hospital for pneumonia. After one week of treatment with antibiotics, he was asymptomatic and there was no more sign of respiratory distress. However, his oxygen saturation persistently ranged from 88 to 92%. Initial transthoracic echocardiography (TTE) showed a structurally normal heart. He was referred to our hospital for further assessment. Clinically, he had mild cyanosis. Other clinical examination including the cardiovascular system examination was unremarkable. Evaluation with TTE showed a normal heart anatomy. However, contrast echocardiography demonstrated a right-to-left shunting at the atrial level. Subsequently, a transesophageal echocardiography (TEE) showed a membrane across right atrium [Figure 1] without causing obstruction to right ventricular inflow [Figure 2]. There was also an anterior secundum atrial septal defect (ASD) and the bubble study showed an immediate right-to-left shunting through the ASD [Figure 3]. His cardiac catheterization showed a normal hemodynamic study. Two months later, surgical resection of the cor triatriatum dexter membrane and patch closure of ASD were performed. Postoperative echocardiography showed the minimal residual membrane of the cor triatriatum dexter with no residual ASD shunting. Clinically, he remained asymptomatic and his saturation improved to >95%.

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DISCUSSION

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 into 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 the fetal life.[1] Normally, the valve regresses by approximately 12 weeks gestation and leaves behind the crista terminalis superiorly and the eustachian valve of the inferior

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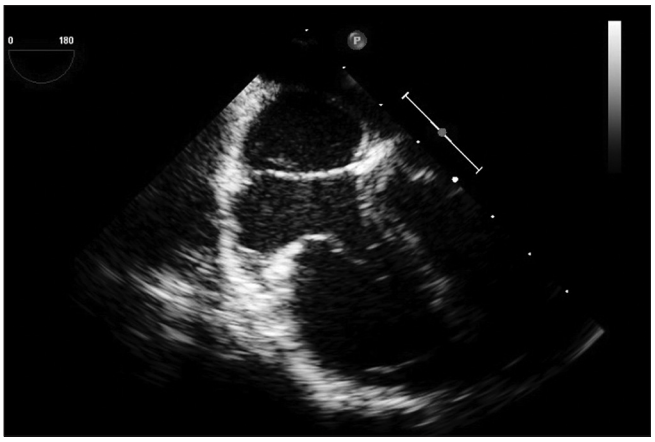

Figure 1: 2D transoesophageal echocardiography apical four chamber view showing presence of a membrane separating the right atrium into two parts

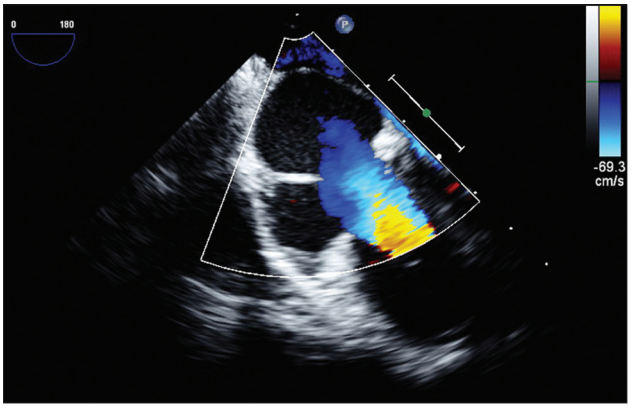

Figure 2: The blood flow to the right ventricle was not obstructed by the membrane

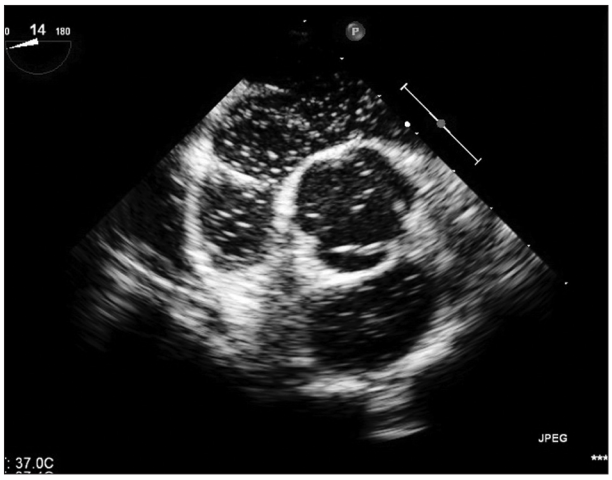

Figure 3: Contrast echocardiography showed an immediate right-to-left shunting through the secundum atrial septal defect

vena cava and the thebesian valve of the coronary sinus inferiorly. Complete persistence of the right sinus valve results in a separation between the smooth and trabeculated portions of the right atrium, constituting cor triatriatum dexter.[3]

Unlike cor triatriatum sinister, which carries a high mortality rate if not repaired, cor triatriatum dexter has varying clinical manifestations depending on the degree of partitioning or septation of the right atrium. When the septation is mild, the condition is often asymptomatic and is an incidental finding during surgery to correct other cardiac abnormalities or during echocardiography. More severe septation can cause right-sided heart failure and elevated central venous pressures due to obstruction of the tricuspid valve, the right ventricular outflow tract, or the inferior vena cava.[1] Cor triatriatum dexter can occur as an isolated cardiac anomaly or associated with other malformation of right heart structures such as pulmonary artery stenosis or atresia, tricuspid valve abnormality, ASD, and Ebstein anomaly.[1,4] Cyanosis is a very rare presentation of this condition. Hoye and colleagues reported a neonate with cor triatriatum dexter with cyanosis and assessment showed significant obstruction to right ventricular inflow. Most of the flow was directed to the left atrium through a patent foramen ovale.[5] Our patient has an ASD that allows the deoxygenated blood from right atrium to flow across it and mix with oxygenated blood in the left atrium that led to mild degree of cyanosis. Interestingly, this happened despite insignificant flow obstruction to right ventricle and normal pulmonary artery pressure. One possible explanation is that the membrane acts as a venous valve that directs the blood flow from right to left atrium.

Since many patients are asymptomatic, the diagnosis of cor triatriatum dexter often is determined at postmortem examination. Antemortem diagnosis can be determined by using angiography, echocardiography, contrast echocardiography, or MRI. There have been numerous reports about the use of echocardiography to determine the diagnosis noninvasively that mainly date from the mid-1980s to early 1990s.[6,7] Baweja *et al*., reported a case in which 3D transesophageal echocardiography was able to detect cor triatriatum dexter in partial atrioventricularseptal defect (AVSD) patient that was missed by 2D echocardiography.[8] In a study in which MRI was compared with echocardiography and cardiac angiography in the evaluation of pulmonary venous anomalies, which included cases of cor triatriatum, MRI had a higher detection rate (95%) than the other modalities (69% for angiography and 38% for echocardiography).[9]

Asymptomatic patients are generally not treated unless they are undergoing cardiac surgery for other reasons. In the past, the mainstay of treatment for symptomatic patients has been surgical resection of the dividing membrane.[2-4] Our patient underwent surgery to close the ASD and the opportunity was taken to do a resection of the cortriatriatum membrane as well. Recently, percutaneous catheter disruption of the membrane has been reported and has been suggested as a preferred alternative to open heart surgery.[10]

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