Case Reports

Cor Triatriatum Dexter and Atrial Septal Defect in a 43-Year-Old Woman

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Cor triatriatum dexter is a rare congenital heart anomaly in which a membrane divides the right atrium into 2 chambers. We report the case of a 43-year-old woman who had cor triatriatum dexter and a large atrial septal defect. During attempted percutaneous closure, the balloon disrupted the membrane and revealed that the defect had no inferior rim, precluding secure placement of an AMPLATZER Septal Occluder. Surgical treatment subsequently proved to be successful.

In patients with an incomplete membrane and a septal defect with well-defined rims, percutaneous treatment can be the first choice. In patients who have cor triatriatum dexter and unfavorable anatomic features or concomitant complex heart anomalies, open-heart surgery remains the gold standard for treatment. ***(Tex Heart Inst J 2014;41(4):418-20)***

Cor triatriatum dexter (CTD) is a congenital anomaly in which the right atrium is divided into 2 parts by a membrane or fibromuscular band. This condition is extremely rare: in high-volume echocardiographic laboratories, the prevalence of CTD is less than 0.01%.[1] Cor triatriatum dexter is often associated with other congenital anomalies, such as pulmonary artery stenosis or atresia, hypoplastic right ventricle, Ebstein anomaly, and atrial septal defect (ASD).[2,3] Symptomatic patients with CTD and ASD are typically candidates for surgery, although successful percutaneous procedures have been reported.[4,5] We report our experience with a CTD patient when we attempted percutaneous treatment of the anomaly.

Case Report

In January 2013, a 43-year-old woman was examined at a regional hospital because of dyspnea on exertion (New York Heart Association [NYHA] functional class II–III). She had no signs of chronic heart failure. A continuous murmur was heard. A transthoracic echocardiogram (TTE) showed a 2.5 × 3.5-cm ASD (pulmonary-to-systemic flow ratio, 3:1) and a membrane that incompletely divided the right atrium into 2 chambers. Cor triatriatum dexter or a giant Eustachian valve was suspected, and the patient was referred to our institution.

We used transesophageal echocardiography (TEE) and catheterization in an attempt to correct the anomaly percutaneously. The TEE showed that a membrane divided the right atrium into upper and lower chambers, thus confirming a diagnosis of CTD (Fig. 1). Percutaneous balloon dilation disrupted the membrane and enlarged the communication between the chambers. This exposed the complete absence of an inferior rim on the patient's ASD: no tissue was available for anchoring an AMPLATZER™ Septal Occluder (St. Jude Medical, Inc.; St. Paul, Minn). The risks of device malpositioning, device embolization, and residual shunting were high, so the patient was scheduled for surgery.

Median sternotomy was performed and cardiopulmonary bypass was established. Access to the anomaly was gained through a right atriotomy. The large, fibrous congenital membrane, partially disrupted by the earlier percutaneous dilation, ran diagonally through the right atrium, reaching the interatrial septum on one end and the orifice of the inferior vena cava (IVC) on the other (Fig. 2A). A free margin and multiple small fenestrations enabled communication between the 2 atrial compartments. The ASD had a well-defined rim except in its inferior portion, where there was none. We resected the membrane, preserved its basal section for reconstructing the interatrial septum (Fig. 2B), and closed the residual ASD with use of a 3 × 2.5-cm pericardial

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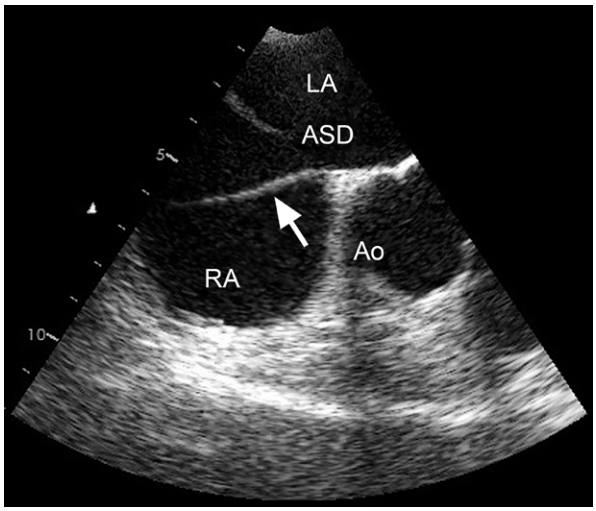

Fig. 1 *Transesophageal echocardiogram shows a membrane (arrow) dividing the right atrium.*

Ao = aorta; ASD = atrial septal defect; LA = left atrium; RA = right atrium

patch (Fig. 2C). The patient's postoperative recovery was uncomplicated, and she was discharged from the hospital 6 days after surgery. No residual shunt was seen on TEE 3 months later, and the patient's functional status improved to NYHA class I.

Discussion

The main pathologic and echocardiographic finding in CTD is a membrane that separates the right atrium into 2 chambers. The upper chamber receives the venous blood from the venae cavae, and the lower chamber is in contact with the tricuspid valve and the right atrial appendage. The membrane varies substantially in size and shape. It can resemble a diaphragm or be funnel-shaped or bandlike; and it can be intact or contain fenestrations, ranging from small and restrictive to large and open.

Cor triatriatum dexter results from the complete persistence of the right sinus valve of the embryonic heart. This structure is normally reabsorbed in early gestation, leaving the Eustachian valve and the valve of the coronary sinus (Thebesian valve). The right sinus valve might persist to a lesser extent, forming a prominent Eustachian valve or the Chiari network. These anomalies have the same embryogenesis as CTD and can mimic it.[6] Echocardiographic evidence of ASD and eventually the presence of cyanosis suggest CTD.[7] In addition, CTD can be mistaken for a right atrial mass.[8] 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.[2,3] 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. Symptomatic patients should undergo correction of the underlying abnormalities. Our patient had dyspnea on exertion, and the large ASD was considered to be responsible for those symptoms.

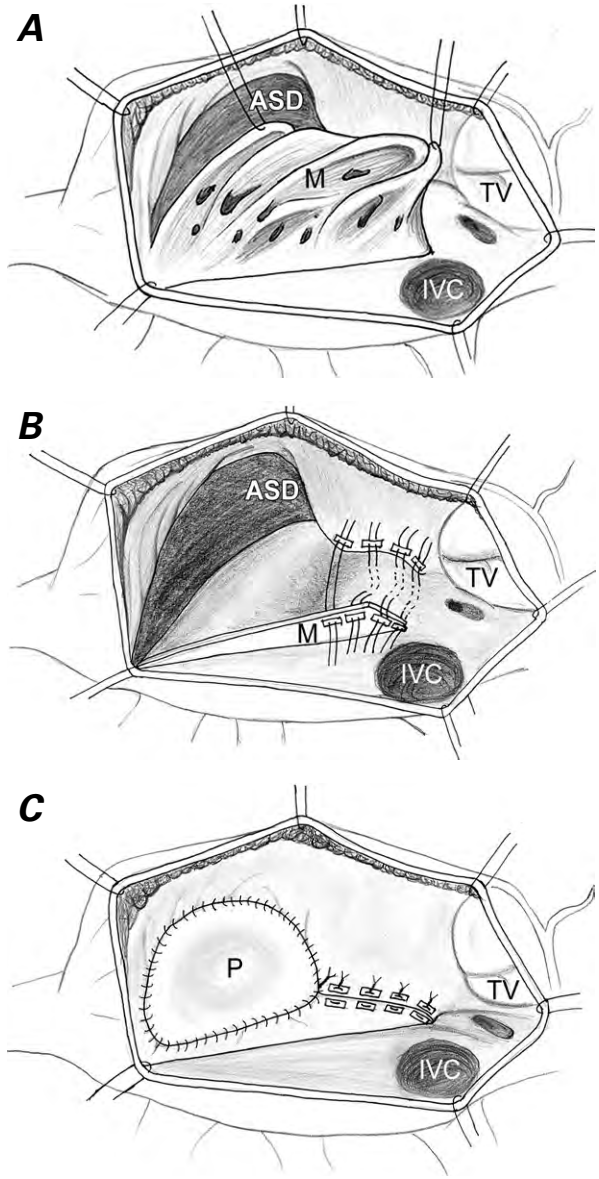

Fig. 2 *Diagrams depict surgical correction of the anomaly. **A**) Right atriotomy reveals a large, fibrous membrane, with small fenestrations, that divides the right atrial cavity into 2 compartments. **B**) All but the basal part of the membrane is removed, and the remnant is joined to the adjacent rim of the atrial septal defect (ASD) with a few stitches. The base of the lower chamber is plicated, with care taken not to obstruct coronary sinus inflow. **C**) The residual ASD is closed with use of a pericardial patch.*

IVC = inferior vena cava; M = membrane; P = patch; TV = tricuspid valve

Successful percutaneous ASD closure has been reported in patients with CTD.[4] Percutaneous catheter disruption of the membrane has been proposed as an alternative to surgery for CTD.[5] However, the CTD membrane can obstruct the view and preclude adequate definition of the margins of an associated ASD. Consequently, as we found, it can be difficult to determine whether percutaneous or surgical ASD closure is optimal.

The criteria for successful percutaneous ASD closure include a maximal defect diameter of 40 mm and rims of at least 5 mm toward the caval orifices, right upper pulmonary vein, and mitral valve.[9,10] Because percutaneous correction was the less invasive option, we attempted it first. However, after the membrane was disrupted, it became evident that our patient's ASD had an IVC rim insufficient for catheter-based closure. Only surgical treatment afforded full insight into the anatomy and enabled correction of the anomaly.

Cor triatriatum dexter is unexpected in adults, and it can pose therapeutic dilemmas chiefly because of its rarity. Methodical preoperative imaging with TTE, TEE, and nuclear magnetic resonance spectroscopy is essential for making correct therapeutic decisions. In patients with nonobstructive membranes and ASDs with well-defined rims, the percutaneous approach is probably the treatment of choice. In patients with unfavorable anatomic features or concomitant complex heart anomalies, open-heart surgery remains the gold standard in the treatment of cor triatriatum dexter.

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