Discrete subpulmonic membrane in transposition of great arteries with an intact ventricular septum

Trushar Gajjar, Nageswar Rao, Neelam Desai

Received: November 23, 2014 Accepted: January 06, 2015 Published online: February 10, 2015

The left ventricular outflow tract obstruction is rare in transposition of the great arteries with an intact ventricular septum. The obstruction may be dynamic or anatomic. Herein, we present a four-year-old boy who was diagnosed with dextro-transposition of the great arteries, patent ductus arteriosus with an intact ventricular septum, and left ventricular outflow tract obstruction due to a discrete subpulmonic membrane.

Keywords: Left ventricular outflow tract obstruction; subpulmonic membrane; transposition of great arteries.

The patient was taken for surgery and a conventional median sternotomy approach was used. Preoperative findings revealed d-TGA, and the aorta to the right and anterior to the pulmonary artery. The coronary arteries were normal for d-TGA with sinus-1 giving the left coronary artery and the sinus-2 giving the right coronary artery. Both the atria were of good size with an absent atrial isomerism. There was a large secundum type of atrial septal defect and a complete diaphragm like subpulmonic membrane with a central opening (Figure 2a). Pulmonary valve was tricuspid with thickened leaflets and a small PDA. The subpulmonic membrane was excised, the PDA was ligated and a physiological correction was done by a modified Sennings operation. The weaning from cardiopulmonary bypass was done in a usual way.

The postoperative course was uneventful and an immediate postoperative echocardiography revealed nonobstructed systemic and pulmonary venous return, no residual shunt, mild tricuspid regurgitation, trivial pulmonary regurgitation; gradient across the LVOT was 6 mmHg with mild right ventricular dysfunction and a normal left ventricular function. A three month and two year follow-up showed asymptomatic patient and echocardiography revealed normal systemic and
pulmonary venous drainage, insignificant gradient across the LVOT and a good ventricular function.

**DISCUSSION**

Left ventricular outflow tract obstruction is well-known in overall 30-35% of patients in association with TGA.\(^1\) However, only 0.7% of patients with TGA and intact ventricular septum develop a critical LVOT obstruction.\(^3\) Very few cases of discrete subpulmonic membrane have been reported in d-TGA/IVS in the literature.\(^1\) The obstruction may be dynamic or anatomic with anatomic causes ranging from the subvalvar fibrous ridge to fibrous tags, hypertrophied bands and valvar stenosis.\(^1\) A discrete subpulmonic membrane causing LVOT obstruction in patients with d-TGA is rare, although it has been reported with doubly committed ventricular septal defect in association with subpulmonary fibrous ridge.\(^2\) Ozkutlu et al.\(^2\) reported a discrete subaortic and subpulmonic stenosis with doubly committed ventricular septal defect in a series of nine patients. Raff et al.\(^3\) reported a subpulmonic membrane in association with a ventricular septal defect and aortic insufficiency. Duggal et al.\(^4\) showed a subpulmonic membrane in association with a supracristal ventricular septal defect. Tomar et al.\(^5\) recommend a careful echocardiography of all similar presentations which may enable us to learn more of this rare lesion. The pathophysiology of LVOT obstruction in TGA/IVS shows that a dynamic type of obstruction is common.\(^1\) A fixed type of obstruction is very rare and of two

**Figure 1.** (a) Transthoracic echocardiogram in a subcostal view showing a discrete subpulmonic membrane. (b) A left ventriculogram through the femoral artery to descending aorta to patent ductus arteriosus to main pulmonary artery to left ventricle showing intact interventricular septum and discrete subpulmonic membrane. SPM: Subpulmonic membrane.

**Figure 2.** Intraoperative pictures taken from the head end of the table showing, (a) a complete fibrous ring just below the pulmonary valve, (b) excised subpulmonic ring. SPM: Subpulmonic membrane.
Discrete subpulmonic membrane in transposition of great arteries with an intact ventricular septum

types, discrete or tunnel type. The discrete variety initially appears as a patch of endocardial thickening on the septal bulge, but later may evolve into sharp fibrous ridge or a discrete membrane which is analogous to the subaortic membrane. The less common type of fixed subpulmonary obstruction is caused by a tunnel like subpulmonary fibromuscular ridge that extends across the outflow tract. A careful transthoracic echocardiography in subcostal and apical five-chamber view can be useful in the diagnosis of this rare anomaly. Although percutaneous dilatation of subpulmonic membrane has been reported in the literature, the surgical excision is the treatment of choice along with the definitive surgical procedure. If this patient would have presented early with normal, non-regressed left ventricle, Arterial switch would be the preferred operation. Unfortunately, our patient presented late with regressed left ventricle. In case of a significant and borderline left ventricle, the left ventricle can be trained in the late arterial switch operation. On the other hand, in case that corrected transposition (Corrected TGA) subpulmonic membrane can produce pulmonary insufficiency like subaortic membrane, but with lower pulmonary artery pressure, the chances of pulmonary insufficiency are rare. In the absence of a non-significant obstruction, probably gradient less than 40 mmHg without associated with cardiac anomaly requiring surgical correction, the subpulmonic membrane can be left alone, however, there is a possibility of progression of gradient later in life due to the development of fibrosis secondary to turbulence. However, if the gradient is more than 40 mmHg without associated with cardiac anomaly or associated cardiac anomaly is present in spite of gradient less than 40 mmHg in, corrected TGA resection of subpulmonic membrane is advisable.

In conclusion, a discrete subpulmonic membrane in d-TGA with IVS is a rare anomaly, careful transthoracic echocardiography is able to diagnose this rare entity and surgical excision is the treatment of choice along with the definitive surgical procedure.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES