Subpulmonary membrane: A rare cause of right ventricular outflow tract obstruction

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ABSTRACT

Subpulmonary membrane is a rare cause of right ventricular outflow tract (RVOT) obstruction, and only a few case reports exist with or without associated ventricular septal defect. We report a series of three cases with subpulmonary membrane causing RVOT obstruction. Two of these have been operated (the first case operated after unsuccessful attempt at balloon dilatation), and the third case is on follow-up at present.

Keywords: Right ventricular outflow tract obstruction, subpulmonary membrane, ventricular septal defect

INTRODUCTION

Sub-aortic membrane is a well known cause of left ventricular outflow obstruction but discrete sub-pulmonary membrane causing right ventricular outflow obstruction is less well defined.[1] We present series of three cases of this rare entity.

CASE REPORTS

Case 1

A 3-year-old boy was referred to our hospital with the diagnosis of severe pulmonary stenosis with right ventricular dysfunction and severe tricuspid regurgitation. On further evaluation at our hospital, he was cyanosed with SpO_2 of 78%, echocardiography showed discrete subpulmonary membrane adherent to pulmonary valve, peak gradient across right ventricular outflow tract (RVOT) was 130 mmHg, RV systolic dysfunction was present, and patent foramen ovale (PFO) was shunting right to left. Emergency balloon dilatation was attempted under general anesthesia, two undersized balloons were used, (Tyshak II, NuMed, HopKinton, NY, USA), both the balloons got ruptured during inflation, and there was little effect on the severity of pulmonary stenosis [Figure 1]. The patient was then taken for surgery; excision of subpulmonary membrane was done along with PFO closure. On the 1st postoperative day, he developed a massive pulmonary hemorrhage for which he required prolonged ventilation. He was finally extubated on the 10th postoperative day. At 5-year follow-up, his echocardiography showed normal right ventricular systolic function and RVOT peak gradient of 20 mmHg.

Case 2

This is a 7-year-old boy, brought to the outpatient department with complaints of breathlessness and cyanosis. Echocardiography showed severe RVOT stenosis caused by subpulmonary membrane adherent to pulmonary valve, hypertrophied right ventricle, and small atrial septal defect (ASD) shunting right to left.

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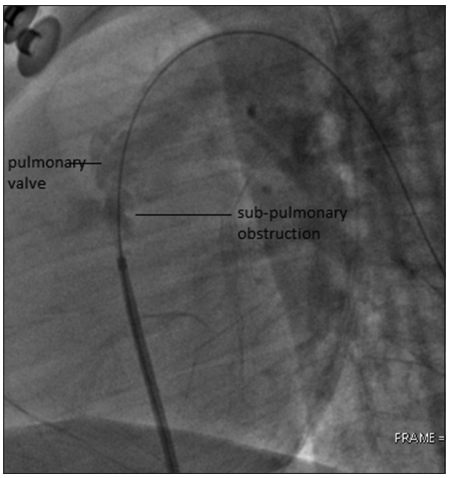

Figure 1 (Case 1): RVOT angiogram with end hole guide catheter with guidewire *in situ*. Severe subpulmonary stenosis is seen. Pulmonary valve is opening normally. RVOT: Right ventricular outflow tract

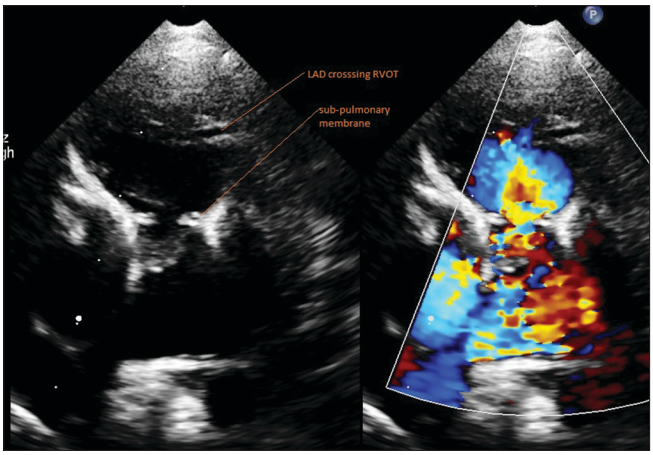

Figure 2 (Case 3): Echocardiogram in parasternal short-axis view with color comparing image showing discrete subpulmonary membrane. Turbulence in color flow starts at the level of subpulmonary membrane

His room air oxygen saturation by pulse oximetry was 70%. Surgical excision of subpulmonary membrane was done along with ASD closure. There was a commissural fusion of pulmonary valve leaflets for which pulmonary valvotomy was also done. The postoperative course was smooth for this patient, and he was discharged on the 5th postoperative day.

Case 3

This is a 3-year-old male child, who was asymptomatic and suspected to have a congenital heart defect because of incidental finding of a heart murmur. Echocardiography showed a small perimembranous ventricular septal defect (VSD) shunting left to right. Discrete sub-pulmonary membrane causing right ventricular outflow obstruction was also present [Figure 2 and Video 1]. The peak Doppler gradient across the RVOT was 51 mmHg in the presence of normal right ventricular function. Additional incidental finding was left anterior descending coronary artery arising from the right coronary sinus and crossing RVOT. This child has been advised for regular follow-up only as the stenosis is not severe.

DISCUSSION

Subpulmonary membrane as a cause of RVOT obstruction in concordant atrioventricular and ventriculoarterial connections is very rare.[1] Only a few isolated case reports are there in the literature.[1,2] A few cases are described with VSD.[3,4] Ours is the first case series on this rare congenital malformation, describing moderate to critical RVOT obstruction.

This entity is clearly different from the double-chambered right ventricle in which case obstruction is caused by muscular tissue and is much below the pulmonary valve whereas in subpulmonary membrane obstruction is caused by a fibrous tissue which is very close to the pulmonary valve. On echocardiography, this pathology more closely resembles subaortic membrane.

Balloon dilatation has been done in two of these case reports (with intact ventricular septum) and the result in both the cases was suboptimal and the need of future surgical intervention was felt.[2,5] Based on two previous case reports and one case from our series, it can be said that surgical excision of subpulmonary membrane is the preferred mode of treatment. Balloon dilatation in our first case was done in 2014; the literature on this rare entity was even scarce at that time. Re-growth of sub-pulmonary membrane after surgical excesion has not been reported so far.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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