Isolated subpulmonic fibrous ring, mirror-image dextrocardia and situs solitus in a young lady unreported and a near miss

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Abstract

Congenital diseases causing obstruction of the right ventricular outflow tract (RVOT) are common, but the isolated subpulmonary membrane/ring is extremely rare and can be difficult to diagnose precisely, especially in adults. We report a case of surgically resected isolated subpulmonic fibrous ring in a lady with mirror-image dextrocardia and abdominal situs solitus that was misdiagnosed by echocardiography as a subaortic membrane.

Keywords: Dextrocardia • Membrane • Congenital • Situs solitus

INTRODUCTION

Congenital heart diseases producing obstruction of the right ventricular outflow tract (RVOT) are relatively common and include abnormalities at the mid-right ventricle, the infundibulum, the pulmonary valve, supravalvular or the branch and/or peripheral pulmonary arteries. Subvalvular pulmonary stenosis commonly occurs as muscular hypertrophy associated with Tetralogy of Fallot or ventricular septal defect (VSD) [1]. Membranous subpulmonary stenosis is rare, and only few cases have been reported, mostly in association with other congenital defects like pulmonary valve stenosis and VSD [2]. Isolated subpulmonary membranes are extremely rare [3]. We encountered a young lady with this anomaly, associated with mirror-image dextrocardia and situs solitus where the diagnosis was missed by echocardiography and confirmed at the time of surgery.

CASE REPORT

A 27-year old lady was referred with increasing shortness of breath on moderate exertion. Examination revealed normal vital signs. The apex was on the right fifth intercostals’ space, of normal character. First and second heart sounds were audible, with harsh systolic murmurs at the left sternal border.

Electrocardiogram revealed right axis deviation and left ventricular hypertrophy. Chest X-ray showed dextrocardia with abdominal situs solitus and cardiomegaly. Transthoracic echocardiography (TTE) showed situs solitus, mirror-image dextrocardia (right ventricle on the left) with atrioventricular and ventriculo-arterial concordance. There was a discrete membrane that was reported to be below the aortic valve, causing severe stenosis with a peak gradient of 70 and mean of 40 mmHg. No other associated congenital defects were seen.

Transoesophageal echocardiography (TOE) was performed preoperatively and confirmed the TTE findings. After discussion with the surgeon, the operation was performed through a median sternotomy.

SURGICAL FINDINGS

The findings were Situs solitus, L-loop ventricles, L-malposition of great vessels with concordant atrioventricular and ventriculo-arterial connections. Mirror-image dextrocardia: the left ventricle was anterior, with the apex towards the right, right ventricle was to the right and posteriorly. The aorta was anterior to the pulmonary artery with the aortic arch to the right (Fig. 1). On bypass, the aorta was opened in the normal way, and there was no subaortic membrane.

TOE was repeated, and with direct anatomical guidance a discrete fibrous ring 2.5 cm below the pulmonary valve was found (Fig. 2). The right atrium was opened, and through the tricuspid valve which was retracted to get into the RVOT, the ring was excised. The pulmonary valve was normal. TOE confirmed the complete excision of the membrane and a wider RVOT with a Doppler peak gradient of 30 mmHg. She continued to do well on follow-up in the out-patient clinic, 2 years post surgery.

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DISCUSSION

Isolated subpulmonary fibrous ring is extremely rare, and no association with mirror-image dextrocardia and situs solitus, to our knowledge, has been previously reported. The pathology of the fibrous ring can be related to tricuspid valve tissue or fibrous tags from the inferior vena cava or coronary sinus [4].

The diagnosis of such a disease can be challenging, especially in adults, because of its rarity and the difficulty of assessing RVOT on TTE. Strict adherence to sequential analysis could have avoided the misdiagnosis on TTE; however, the presence of mirror-image dextrocardia with the anteriorly positioned aorta might have misled the echocardiographer to the diagnosis of subaortic membrane, which is a well-known entity. Mirror-image dextrocardia is usually associated with situs inversus, and its association with situs solitus is extremely rare [5]. More imaging using 3D echocardiography and cardiac CT/magnetic resonance imaging is highly recommended before surgical treatment.

This case is a near miss where a subpulmonic membrane could have been left behind if a TOE was not used routinely in our centre for such cases.

CONCLUSION

We described a case of situs solitus, mirror-image dextrocardia and subpulmonary fibrous ring, which to our knowledge has not been reported previously. TOE is a valuable tool intraoperatively.

Conflict of interest: none declared.

REFERENCES