CASE REPORT

Cor triatriatum sinister presenting in the adult as mitral stenosis

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Cor triatriatum sinister is a rare congenital defect in which the left atrium is divided by a fibromuscular membrane into two distinct chambers. Classically, patients present in infancy although in some cases they remain asymptomatic until adulthood. The clinical features on presentation can mimic those of mitral stenosis due to the obstructive properties of the membrane. Cor triatriatum sinister presented in this case in an adult as mitral stenosis. Factors that may be relevant in determining late presentation are also discussed.

Cor triatriatum sinister is a rare congenital defect first described by Church in 1868.[1] It is found in only 0.1% of cases of congenital heart disease.[2] In the classical form of cor triatriatum the left atrium is divided by a fibromuscular membrane into two chambers: a posterosuperior chamber receiving the pulmonary veins, and an anteroinferior chamber (true left atrium) communicating with the mitral orifice.[3] Embryologically this condition is thought to arise from the failure of inclusion of the pulmonary veins into the left posterior atrial wall. The clinical features on presentation can mimic those of mitral stenosis. Classically, patients present during the neonatal period or early infancy although in some instances cases remain undetected until the patient is an adult.[4]

We present a case of cor triatriatum sinister presenting in an adult as mitral stenosis. A comment follows on factors that may be relevant in determining late presentation.

CASE REPORT

A normally well 46 year old man presented to his general practitioner with a six week history of flu-like symptoms and right sided chest discomfort. On examination the patient was found to be normotensive with an irregular pulse of 90 beats/min. ECG confirmed the presence of atrial fibrillation. Chest radiography displayed significant cardiomegaly with pulmonary venous congestion. A transoesophageal echocardiogram was performed, which showed a thin mobile septum in the left atrium attached medially to the fossa ovalis. A 2 × 1 cm perforation was noted at the superior margin of the membrane. Doppler echocardiography estimated the mean transmembrane pressure gradient at 2 mm Hg. Treatment with digoxin and warfarin was commenced, after which all symptoms resolved. It was felt because of prognostic implications that surgical excision of the membrane would confer the best long term outcome. Coronary angiography performed before surgery showed a right dominant system with no evidence of coronary artery disease.

At surgery the heart was approached through a median sternotomy with standard aortobicaval cannulation to establish cardiopulmonary bypass. The findings of the transoesophageal echocardiogram were confirmed and the atrial membrane was excised around its periphery (fig 1). Postoperative recovery was uneventful and the patient was discharged home on the fifth postoperative day.

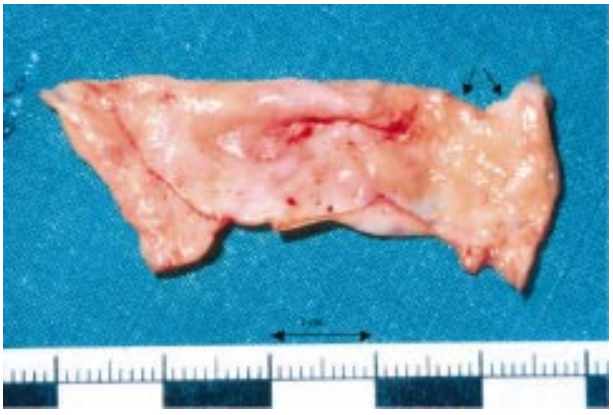

Figure 1 Left atrial membrane following surgical resection. Arrows show the site of the communicating fenestration.

DISCUSSION

Cor triatriatum sinister is a rare condition in which the clinical features can mimic those of mitral stenosis. Pathophysiologically the obstructive nature of the membrane leads to the creation of a pressure gradient with an associated rise in pulmonary arterial and venous pressures.[3] Classically, patients present in infancy although in some cases they remain asymptomatic until adulthood. It has been previously suggested that progressive narrowing of the membrane fenestration may be responsible.[4]

We recently conducted a preliminary review of 40 published cases into factors determining late presentation in cases of adult cor triatriatum sinister presenting as mitral stenosis. The membrane fenestration area was estimated directly by measuring the membrane fenestration or indirectly through pressure recordings. Our analysis suggests that the membrane fenestration area does not alter with age. We postulate that late clinical presentation in such instances may be caused by the development of atrial fibrillation or mitral regurgitation. Only one case report that we reviewed commented on the nature of the associated mitral valve disease. In this instance histological examination showed myxomatous degeneration.[5]

Cor triatriatum sinister remains a rare form of congenital heart disease although it is being diagnosed with increasing frequency in the adult population. This is likely to be a result of improvement in diagnostic imaging. The natural history of this condition has been poorly documented, which relates primarily to the small case numbers available for review. Our work adds further to the published literature available on this rare congenital anomaly.

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