CASE REPORT

Cor-triatriatum –A Rare Congenital Heart Disease Presented in Adulthood-surgical treatment in Apollo Hospitals Dhaka

N M Zahangir[1], A H M L Kabir[2], F Ahmed[3], S T Ahmad[3], M Hossain[4], N Hossain[5], M S Rahman[5], M M H Khan[6], M Q I Talukder[7]

Introduction

Cor-Triatriatum Sinstrum is a rare congenital defect in which the left atrium is divided by a fibro muscular membrane into two distinct chambers. Classically, patients present in infancy although in some cases they remain asymptomatic until adulthood.[1] Diagnosis is usually achieved by echocardiography, treatment of choice is excision of the membrane.

Case report

A 25 yrs old non diabetic, normotensive lady had complaints of exertional breathlessness for last 3 years. She had also complaints of recurrent fever. On 01.02.12 she was diagnosed as having Cor-Triatriatum (with dilatation of both 3rd atrium & left atrium proper) and recommended for resection of membrane. Echo screening showed whole left atrium is dilated (44.8 mm). A fenestrated partition membrane made it two chambers. Pulmonary veins opened to upper chamber (3rd atrium) which was hugely dilated. Lower chamber (left atrium proper) was mildly dilated. Surgical removal of fibro muscular membrane between two left atrial chambers was done on 02/02/2012. Following a full median sternotomy, thymus was dissected out. Pericardiotomy was done. Patient was heparinized and Cardio Pulmonary Bypass was established by aortic and selective SVC and IVC cannula. After reducing the temperature to 32 degree Celsius aorta was cross clamped. Right atriotomy was done by a longitudinal incision. Left atrium was opened and inspected through inter atrial septum. There was a fibro muscular membrane separating large upper left atrial chamber from lower small left atrial chamber proper. Through a small opening in the fibro muscular membrane the two chambers communicated. Pulmonary venous drainage and mitral valve were inspected and found to be normal. The membrane was excised out. Inter atrial opening was closed. Right atrium was closed by 5-0 prolene suture. After deairation aortic cross clamp was released. Patient returned to sinus rhythm spontaneously. Patient slowly weaned from CPB. The post operative period was uneventful. Post operative echo screening on 5th post operative day showed no residual membrane in the left atrial chamber. Patient was discharged on 8th post operative day in a haemodynamically stable condition.

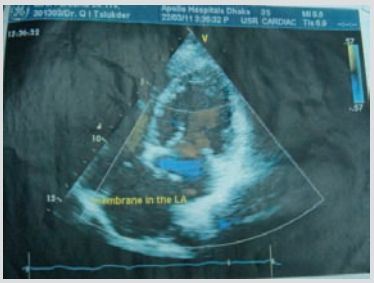

Fig 1: Echo shows membrane in left atrium

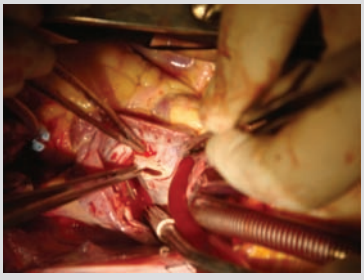

Fig 2: Small opening in the membrane dividing the left atrium into two chambers

1. Senior Registrar, Department of Cardiothoracic Surgery, Apollo Hospitals Dhaka 2. Specialist, Department of Cardiothoracic Surgery, Apollo Hospitals Dhaka 3. Registrar, Department of Cardiothoracic Su 4. Senior Registrar, Department of Cardiothoracic Anesthesia, Apollo Hospitals Dhaka 5. Registrar, Department of Cardiothoracic Anesthesia, Apollo Hospitals Dhaka 6. Resident Medical Officer, Department of Cardiothoracic Surgery, Apollo Hospitals Dhaka 7.Consultant & Coordinator, Department of Cardiothoracic Surgery Apollo Hospitals Dhaka

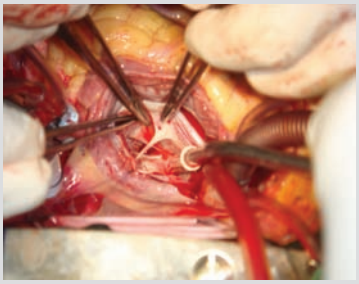

Fig 3: Left upper and lower pulmonary veins are seen in hugely dilated third atrium

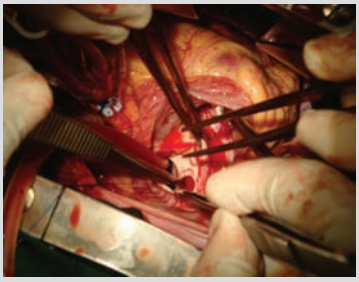

Fig 4: Right upper and lower pulmonary veins are seen in hugely dilated third atrium

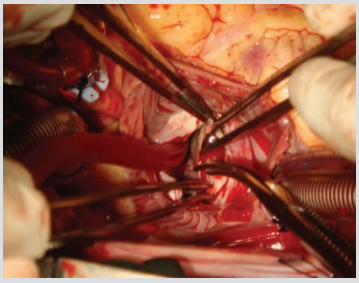

Fig 5: Fibro muscular membrane is being excised

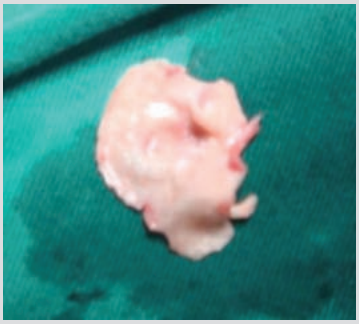

Fig 6: Excised membrane

Discussion

Cor-triatriatum is a rare congenital heart disease with incidence of about 0.1 – 0.4%.[2] Classically, patients are diagnosed in infancy, although in some cases they remain asymptomatic until adulthood. Pathophysiologically the obstructive nature of the membrane leads to creation of a pressure gradient, with an associated rise in pulmonary arterial and venous pressures.[3] Total excision of the accessory septum utilizing cardiopulmonary bypass is presently the appropriate surgical treatment of this entity.[4] From May 1960 to January 1992, 13 patients with Cor-triatriatum underwent surgical correction at the Mayo Clinic. Their ages ranged from 7 months to 57 years. Echocardiography was the procedure of choice for diagnosing Cor-triatriatum. The membrane was excised through a left atrial approach in seven patients and through a right atriotomy in six. One critically ill patient who underwent an emergency operation died early postoperatively, and one patient with chromosomal abnormalities and multiple cardiac defects died 2 months after an uneventful postoperative course. Postoperative angiography or echocardiography in other patients showed no residual inter atrial shunt or recurrent left atrial membrane.[5]

Conclusion

Cor-triatriatum sinister is a rare congenital heart disease and rarely found in adults. Surgical treatment is the gold standard for treating cor-triatriatum even if it presents in adult age. An expert surgical team with proper ICU support is essential.

Reference

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