Shone’s complex and Cortriatriatum Sinister: A Rare Combination.

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ABSTRACT

Shone’s complex is a Congenital Heart Disease (CHD), consisting of multiple levels of left sided obstructive lesions including supravalvular mitral membrane (SVMM), parachute mitral valve, subaortic stenosis and coarctation of aorta. Cortriatriatum sinister is a rare congenital defect in which a membrane divides LA into two distinct chambers. Shone’s complex a rare congenital anomalies, described initially by Shone et al., consists of four obstructive lesions of the left side of the heart.

Key words: Shone’s complex, Cortriatriatum sinister, Congenital Heart Disease, 2DEchocardiography, Cortriatriatum dexter

INTRODUCTION

Shone’s complex is a Congenital Heart Disease (CHD), consisting of multiple levels of left sided obstructive lesions including supravalvular mitral membrane (SVMM), parachute mitral valve, subaortic stenosis and coarctation of aorta. Cortriatriatum sinister is a rare congenital defect in which a membrane divides LA into two distinct chambers. Shone’s complex a rare congenital anomalies, described initially by Shone et al., consists of four obstructive lesions of the left side of the heart. The LVOT obstruction features may include subaortic stenosis, valvar aortic stenosis, bicuspid aortic valve, and coarctation of aorta. Fewer than 100 patients with Shone’s complex have been reported in the literature. It is mostly detected in childhood as the patient becomes symptomatic by the age of 2 years. The usual symptoms are of heart failure and reduced cardiac output. The child usually has recurrent episodes of respiratory tract infections due to pulmonary congestion.

Cortriatriatum represents 0.1% of all congenital heart defects and 80% have associated cardiac defects in pediatric population (ostium secundum ASD, TOF, Bicuspid aortic valve, Anomalous pulmonary venous return, Coarctation of aorta, DORV, VSD, Common AV canal). It was first described by Church in 1868 and was named by Borst in 1905. It involves usually the left atrium (Cortriatriatum sinister) and rarely the right atrium (Cortriatriatum dexter). The atrium is divided into two chambers by a thick fibro-muscular or membranous septum into (i) a proximal or superior chamber which drains the pulmonary venous blood and (ii) a distal or inferior chamber (true atrium) which is in contact with the AV valve and contains the atrial appendage and the true atrial septum. Clinical features mimic those of mitral stenosis (but absence of loud S1 and opening snap). Pathophysiology the obstructive membrane leads to a pressure gradient with an associated rise in pulmonary arterial and venous pressure. Age and symptoms at the time of diagnosis are highly variable and depend on the degree of obstruction to the pulmonary venous return and presence of associated cardiac lesion. The definitive treatment of Cortriatriatum sinister is surgical excision of the membrane. Balloon dilatation can be an alternative in selected cases as a bridge to surgical repair. However our patient had undergone balloon coarctoplasty for severe coarctation of aorta in neonatal period. Later patient during follow up had been diagnosed with Shone’s complex with Cortriatriatum Sinister and was referred for surgical correction during follow up.

CONCLUSION

Shone’s complex and Cortriatriatum Sinister are rare congenital anomalies and the combination is extremely rare. cardiac imaging modalities, mainly two dimensional transthoracic and colour doppler studies, tran-
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soesophageal echocardiography, these valuable diagnostic tools help to optimise the management of this congenital anomalies whose rarity may frequently lead to misdiagnosis. Surgical correction is the definitive treatment for these two rare congenital anomalies.

CONFLICTS OF INTEREST

Nil

ETHICAL COMMITTEE CLEARANCE:

Obtained

REFERENCES:


