LETTER TO EDITOR (VIEWERS' CHOICE)

OBSTRUCTIVE COR TRIATRIATUM: A RARE PRESENTATION

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An 8 years old girl presented with left-sided chest pain for 15 days. Examination was essentially normal except for systolic murmur in the tricuspid area. Her weight for age was 76%, height for age was 92.4% and body mass index was 14.6. Chest X ray and electrocardiogram were normal. Echocardiography was suggestive of cor triatriatum (obstructive type) with normal bi-ventricular dimensions and normal valves and left ventricular ejection fraction of 63%.

Cor triatriatum is a rare congenital cardiac anomaly, incidence being about 0.1%. (1) In this, a fibromuscular membrane divides the left atrium into two chambers. The proximal chamber communicates with the distal chamber (the true left atrium with left atrial appendage) through one or more openings. It may be obstructive or non-obstructive depending on the presence and size of fenestrations in the dividing membrane and obstruction to the pulmonary blood flow. It has a varied age of presentation from infancy to old age. (2) Clinical presentation depends on the size of the opening(s) in the membrane, as symptoms are directly related to the degree of obstruction to pulmonary venous drainage and pressure loading on the right side of the heart. Presentation in infancy mimics that of Ebstein's anomaly and in adults mimics that of mitral stenosis. (3) Presentation in infancy is usually fatal with signs of pulmonary edema and hypertension. (4) In our case, initial presentation was at 8 years of age despite it being obstructive in nature. Also, the only complaint was chest pain. Association with other cardiac anomalies is also seen in 70 -80% cases, most common being patent foramen ovale and ostium-secundum atrial septal defect. (5) Electrocardiographic alterations are non-specific. (5) Echocardiography is required for definitive diagnosis. Medical treatment in cor triatriatum depends on symptoms. An incidental echocardiographic finding of a left atrial extra-membrane with no pressure gradient in an asymptomatic patient does not require medical management. Definitive treatment involves surgical excision of the abnormal fibromuscular membrane followed by closure of the inter-atrial septum with a pericardial baffle or patch. Long term prognosis depends on the underlying congenital heart disease (6) and is generally good after surgical correction in isolated cases.

Thus, obstructive varieties of this cardiac anomaly may not be limited only to infancy and an echocardiogram should definitely be performed when other investigations for chest pain are inconclusive, even when it is the only presenting complaint.

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