Turkish Journal of Thoracic and Cardiovascular Surgery 2019;27(2):261-262
http://dx.doi.org/doi: 10.5606/tgkdc.dergisi.2019.16350

Letter to the Editor / Editöre Mektup

Cor triatriatum sinister mimicking mitral stenosis

Mitral darlığı taklit eden kor triatriatum sinister

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As a highly rare congenital defect, cor triatriatum sinister (CTS) in which the left atrial space is divided into two parts by a fibromuscular membrane represents only 0.1% of congenital cardiac anomalies.[1]

Although CTS can be an isolated lesion, it is frequently associated with other congenital cardiovascular anomalies, most frequently with secundum atrial septal defects (ASDs).[1,2] Cor triatriatum sinister is a cardiac anomaly associated with pulmonary venous stenosis, pulmonary arterial and venous hypertension, and congestive heart failure.[2] The degree of pulmonary hypertension and congestive heart failure is determined by the presence of an interatrial connection, the fenestration of fibromuscular membrane, and the type of additional cardiac anomalies (particularly ASDs and anomalous pulmonary venous return).

Cor triatriatum sinister is similar to mitral stenosis in symptomatology and has a new diagnosis in a wide range from newborn to geriatric age group.[3] It may be symptomatic in different decades, depending on the location of the ASD, transition between the proximal and distal chambers, and the presence or absence of additional cardiac defects. In the majority of patients with CTS, symptoms are seen with pulmonary venous obstruction findings during early childhood.[3,4] However, some cases may remain asymptomatic until adulthood due to the wide fenestration on the membrane or even be diagnosed as an incidental echocardiographic finding. The pathophysiology of the disease begins with pulmonary venous congestion secondary to pulmonary hypertension. Progressive pulmonary hypertension may result in tricuspid insufficiency and right ventricular failure. In certain cases, CTS can be complicated by systemic embolism or embolic stroke.[1,3]

To date, several techniques have been used to establish the diagnosis, such as transthoracic echocardiography, transesophageal echocardiography, catheter angiography, computed tomography, and magnetic resonance imaging. Diagnosis is usually based on echocardiography, including the measurement of the trans-membrane pressure gradient to identify the anatomic and physiological abnormalities and identifying other intra-atrial pathologies. Mitral stenosis, supravalvular mitral ring, pulmonary vein stenosis, and atrial septal aneurysms should be considered in the differential diagnosis.[2-4]

Surgery is a simple and effective method, when there is no complex heart disease associated with CTS. Surgical excision of the membrane provides effective long-term results with low morbidity and mortality. The type of incision varies due to the type of cardiac pathology associated with CTS. The surgical approach consists of left or right atriotomy, excision of the obstructing membrane, and repair of the associated intracardiac anomalies.

Received: April 28, 2018 *Accepted:* November 16, 2018 *Published online:* April 24, 2019

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Cite this article as:
Işık O, Akyüz M, Bakiler AR. Cor triatriatum sinister mimicking mitral stenosis. Turk Gogus Kalp Dama 2019;27(2):261-262

In conclusion, recognizing this abnormality to ensure early and accurate diagnosis during echocardiographic evaluation has an important role in determining cor triatriatum sinister mimicking signs and symptoms of mitral stenosis. Of note, the lack of symptoms does not completely rule out the possibility that there would be no complications without serious morbidity in these patients. If this rare cardiac anomaly is left undiagnosed, irreversible pulmonary disease and right heart dysfunction may present a poor prognosis. Thus, surgical treatment in eligible patients can be achieved with good long-term survival rates and low mortality and morbidity.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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