A very rare case: Cor polyatriatum, a variant of cor triatriatum

Kor triatriatumun alışılmadık bir varyantı: Kor poliatriatum

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Summary– Cor triatriatum sinister is a rare congenital heart anomaly. We present an elderly patient with cor polyatriatum, which is a variant of cor triatriatum. The patient was admitted to our hospital with symptoms of congestive heart failure. Echocardiographic evaluation revealed that the left atrium was divided into three spaces of the fibromuscular membrane, resembling a railway. Diagnosis was confirmed by cardiac magnetic resonance imaging. The patient did not accept surgical treatment and was followed medically.

Özet– Kor triatriatum sinister nadir bir doğuştan kalp anomalisidir. Bu yazıda ileri yaşta, kor triatriatumun bir varyantı olan kor poliatriatumlu olgu sunuldu. Hasta konjestif kalp yetersizliği semptomları ile başvurdu. Ekokardiyografik inceleme ile sol atriyumun tren yolu şeklinde, birbirine paralel fibromusküler zarlarla üç adet boşluğa bölündüğü saptandı. Tanı kardiyak manyetik rezonans görüntüleme ile doğrulandı. Cerrahi tedaviyi kabul etmeyen hasta tıbbi izleme alındı.

Cor triatriatum sinister is a very rare congenital heart defect with an estimated incidence of 0.1%.[1] In this malformation, the left atrium is divided by a fibromuscular membrane into posterosuperior and anteroinferior chambers. The pulmonary veins drain blood to the posterosuperior chamber and communicate with the anteroinferior chamber through a fenestrated membrane. Prognosis depends on the size of the fenestrations. If the fenestration is small, the majority of cases become symptomatic during early childhood and patients die at younger ages. Some cases may present in adulthood and the diagnosis is mostly made incidentally. Sometimes, the atrium is divided into more than two chambers, which is defined as cor polyatriatum.

In this paper, we present a case, in which cor polyatriatum was diagnosed after the development of heart failure.

CASE REPORT

A 60-year-old man was hospitalized for the evaluation of congestive heart failure. The history revealed that the patient was previously admitted to the emergency department with fatigue and dyspnea. Further interrogation of the patient about his symptoms revealed that the patient had undergone coronary angiography, which was reported as normal, two years prior. He was then transferred to the cardiology department for management of decompensated heart failure. He was on diuretics, digoxin and ACE inhibitor therapy. On physical examination, he had a regular pulse of 110 beats/min and blood pressure of 145/90 mmHg. Cardiovascular examination revealed a 2/6 holosystolic murmur best heard at the apex. There were bilateral crackles at the mid and base fields of the lung. Electrocardiography showed a normal sinus rhythm with normal QRS axis. Chest X-ray showed bilateral infiltrates with Kerley-B line consistent with congestive heart failure. Complete blood count, chemistry panel and cardiac enzymes were within normal range. The transthoracic echocardiography (TTE) revealed two thin membranes that were parallel to each other dividing the left atrium into three chambers (Fig. 1a). There was moderate mitral and tricuspid insufficiency

Abbreviations:

MR	*Magnetic resonance*
TEE	*Transesophageal echocardiography*
TTE	*Transthoracic echocardiography*

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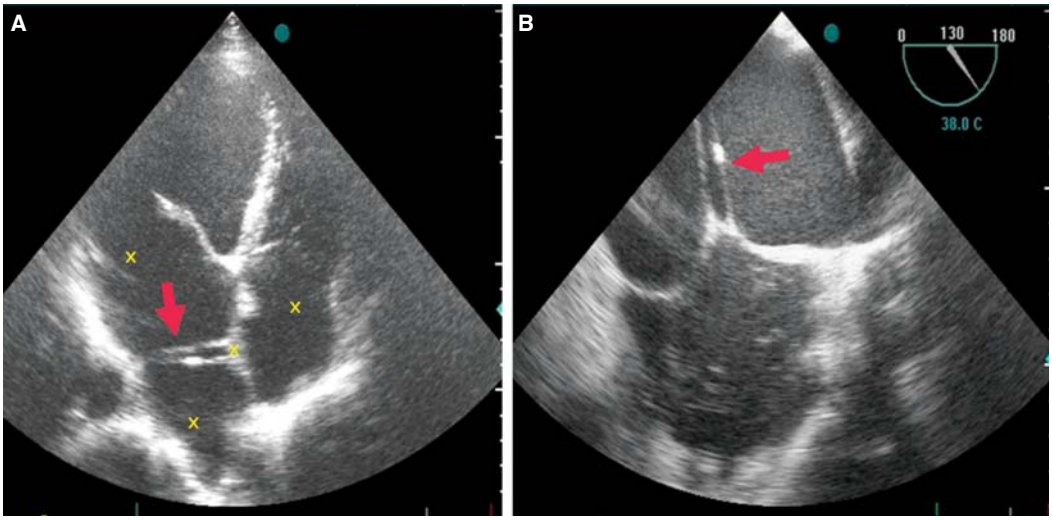

Figure 1. **(A)** Two fibromuscular membranes, which have a railway appearance and divide the left atrium into three chambers are seen in the apical four chamber view of transthoracic echocardiography. **(B)** Two fibromuscular membranes with a railway appearance dividing the left atrium into three chambers are seen in transesophageal echocardiographic view.

and the ejection fraction was 25%. Transesophageal echocardiography (TEE) confirmed the fibromuscular membranes (Fig. 1b). The patient responded well to diuretic treatment. Meanwhile, magnetic resonance (MR) imaging of the heart was performed and showed two thin atrial membranes at the mid left atrium extending from the left pulmonary veins to the fossa ovalis and consistent with cor polyatriatum (Fig. 2). Multiple fenestration areas were present at the membrane. MR imaging showed a railway image of the fibromuscular membranes. No other congenital abnormalities were noted. With optimal medical therapy, the patient's symptoms resolved. The patient is on regular follow up.

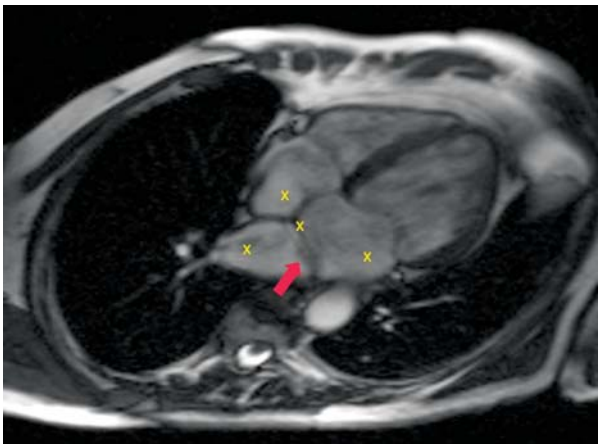

Figure 2. In cardiac magnetic resonance imaging, two fibromuscular membranes are clearly seen dividing the left atrium into three chambers.

DISCUSSION

Cor triatriatum is a very rare congenital cardiac malformation first described by Church in 1868.[2] The left atrium is divided into proximal and distal chambers by a fibromuscular membrane. The fenestrations in the fibromuscular membrane allow blood flow from the proximal chamber to the distal one. The embryological mechanism of cor triatriatum is a controversial subject. There have been some theories suggesting abnormal septum primum overgrowth, incomplete incorporation of the pulmonary veins and impingement of the left atrium by the persistence of a left-sided superior vena cava resulting in a membrane.[3,4] Cor triatriatum is sometimes associated with other cardiac anomalies, including atrial septal defect, persistence of left-sided superior vena cava, anomalous partial pulmonary venous return, and patent ductus arteriosus.[5] The clinical manifestation of cor triatriatum depends on the size of the fenestration. The fenestration may be single or multiple; small or large; central or eccentric. The clinical presentation of cor triatriatum can mimic mitral stenosis, supravalvular mitral ring, pulmonary venous stenosis or left atrial thrombus. Diagnosis can be established by TTE, TEE, 3D echocardiography, computed tomography or MR imaging. TEE is superior to TTE by providing bet-

ter images of the left atrium and fibromuscular band. An interesting finding in our case was that there were no signs of inflow obstruction despite pulmonary congestion. There are three possible explanations:[1] the patient was not tachycardic during echocardiography. The diastolic pressure is highly dependent on heart rate. A lower heart rate decreases the gradient so that it is not possible to detect with echocardiography;[2] the presence of accompanying left ventricular dysfunction may have caused an increase in pressure in the ventricular compartment of the left atrium. This increase may have lowered the pressure gradient between the left atrial compartments;[3] it is also possible that the fenestrations were so wide that the result was no gradient, thus causing no obstruction.

Better resolution, multiple image planes and the non-invasive nature of MR imaging make it the preferred imaging modality. In our case, the unique image of the membrane was examined with MR imaging. Asymptomatic patients, especially with incidental diagnoses, can be followed up regularly by TTE or MR imaging. For symptomatic patients, surgical resection of the fibromuscular membrane is indicated. Successful percutaneous balloon dilation has also been described.[6] Farber et al.[7] described a case of polyatriatum with MR images. Our case also demonstrates the use of TTE, TEE and MR imaging to evaluate cor polyatriatum and to our knowledge the best TTE and TEE images in the literature. Cor polyatriatum should also been considered in patients with a septated left atrium and findings of congestive heart failure.

Conflict-of-interest issues regarding the authorship or article: None declared

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Anahtar sözcükler: Kor poliatriatum/tanı; kor triatriatum/tanı; ekokardiyografi; kalp yetersizliği/komplikasyonları; manyetik rezonans görüntüleme.