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Cor triatriatum sinister with significant pressure gradient in an adult patient

Erişkin bir hastada ciddi basınç gradiyenti oluşturan sol kor triatriatum

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Cor triatriatum sinistrum is rare congenital cardiac malformation. It accounts for approximately 0.1-0.4% of all patients with congenital heart disease. Most cases are diagnosed in childhood, while adult cases are very rare. The condition is characterized by the presence of a fibromuscular membrane dividing the left atrium into two chambers a superior posterior chamber and an inferior anterior chamber. A 30-year-old male presenting with progressive exertional dyspnea for the past two years was admitted to our clinic. Color Doppler ultrasonography revealed an eccentric, mosaic pattern of continuous turbulent flow near the interatrial septum across the membrane. The peak velocity of the flow across the membrane was 2.70 m/s, indicating that the pressure gradient between two chamber was 11.5 mmHg. Surgical correction of the membrane was recommended. The membrane was excised and all symptoms resolved in the scheduled visit at one month following surgery. In this article, we report an adult case of incomplete cor triatriatum sinister with a significant pressure drop.

Key words: Cor triatriatum sinister; surgical correction; transesophageal echocardiography.

Kor triatriatum sinistra, nadir görülen doğuştan bir kardiyak malformasyondur. Doğuştan kalp hastalığı olan tüm hastaların yaklaşık olarak %0.1-0.4 kadarını oluşturur. Olguların birçoğuna çocukluk çağında tanı konulurken, yetişkin hasta sayısı çok azdır. Hastalık sol atriyumu superior-posterior ve anterior-inferior boşluklar olarak iki ayrı boşluğa bölen fibromusküler bir membran ile karakterizedir. Otuz yaşında erkek hasta kliniğimize son iki yıldır progresif artan egzersiz dispnesi ile başvurdu. Renkli Doppler ultrasonografide internal septumdan membranın karşı tarafına doğru uzanan egzantrik, mozaik patern oluşturan türbülan akım saptandı. Akımın membran çapındaki pik velositesi 2.70 m/sn olup, iki boşluk arasındaki basınç gradyentinin 11.5 mmHg olduğunu gösteriyordu. Membranın cerrahi olarak düzeltilmesi önerildi. Membran eksize edildi ve hastanın ameliyattan bir ay sonraki takip vizitinde tüm semptomlar iyileşmişti. Bu yazıda membran üzerinde ciddi basınç gradiyenti saptanan erişkin bir tamamlanmamış kor triatriatum sinistra olgusu sunuldu.

Anahtar sözcükler: Kor triatriatum sinistra; cerrahi düzeltme; transözofageal ekokardiyografi.

Cor triatriatum is rarely found in adults and accounts for approximately 0.1-0.4% of congenital heart disease.[1,2] In this report, we present a case of cor triatriatum sinister with significant pressure gradient that was treated successfully with surgical excision in an adult patient.

CASE REPORT

A 30-year-old male presented with progressive exertional dyspnea that had been occurring for the past two years. A transthoracic echocardiogram revealed a thin, linear echo-dense structure traversing the left atrium (LA). The patient's global left ventricular function was within normal limits. Color Doppler echocardiography revealed mild degrees of mitral and tricuspid regurgitation, and the systolic pressure gradient over the tricuspid valve was 35 mmHg. A transesophageal echocardiogram demonstrated a thin, immobile membrane in the LA attached medially to the intact interatrial septum that

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was separating a posterior chamber from an anterior chamber in connection with the left atrial appendage and a normal mitral valve (Figure 1). Color Doppler ultrasound (CDUS) revealed an eccentric, mosaic pattern of continuous turbulent flow near the interatrial septum across the membrane (Figure 2). The peak velocity of the flow was 2.70 m/s, indicating that the mean pressure gradient between two chambers was 11.5 mmHg, and surgical correction of the membrane was recommended. The membrane was excised, and at the one-month postoperative examination, all of the patient's symptoms had disappeared.

DISCUSSION

Cor triatriatum sinistrum is a rare, congenital cardiac malformation which occurs in about 0.1-0.4% of all patients with congenital heart disease. Most cases are diagnosed in childhood, with adult cases being extremely rare.[1,2] The condition is characterized by the presence of a fibromuscular membrane dividing the LA into a superior posterior chamber and an inferior anterior chamber. The superior chamber receives the pulmonary venous inflow, and the inferior chamber contains the left atrial appendage and the mitral valve orifice.[3] Communication between the two chambers usually occurs through a perforation in the membrane. Loeffler[4] classified hearts with cor triatriatum into three subgroups. Type 1 contains no opening in the membrane, type 2 has a small opening, and type 3 features a large perforation through the membrane. Classically, patients present during the neonatal period or early infancy, although patients with type 3 cor triatriatum may remain undetected until late adulthood depending on the diameter of the orifice and the degree of obstruction. Generally, patients are symptom-free when the diameter is >1 cm. Late presentation of cor triatriatum in advanced adulthood may be due to either fibrosis or calcification of the orifice. This can occur with long-standing turbulent flow through the membrane which can cause stenosis or with the development of mitral regurgitation or atrial fibrillation.[1] The hemodynamic consequences and symptoms of cor triatriatum resemble those of mitral stenosis, and various techniques are available to identify the intraatrial membrane in patients with this anomaly. Routine cardiac catheterization and angiography have been proven to be unreliable in differentiating cor triatriatum from other causes of left atrial inflow obstruction. Standard transthoracic echocardiography in adults also has its limitations in the imaging of posterior structures; hence, it may not allow for the complete characterization of the membrane or the flow across the orifice in the membrane. However, color flow Doppler US can depict abnormal flow across the membrane opening. A transesophageal echocardiogram is useful in determining the difference between intracardiac causes, such as supravalvular mitral ring, dissected LA, atrial myxoma, or thrombus, and extra-cardiac causes, such as mediastinal, pericardial, or a pleural cyst causing left atrial compression. Additional techniques for visualizing the intraatrial membrane include computed tomography (CT), magnetic resonance imaging (MRI), and three-dimensional echocardiography.[1,5] The only definitive treatment for cor triatriatum is complete surgical excision of the membrane, and the long-term results with this procedure are excellent, with 85% survival rates without recurrence.[3,6]

Declaration of conflicting interests

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

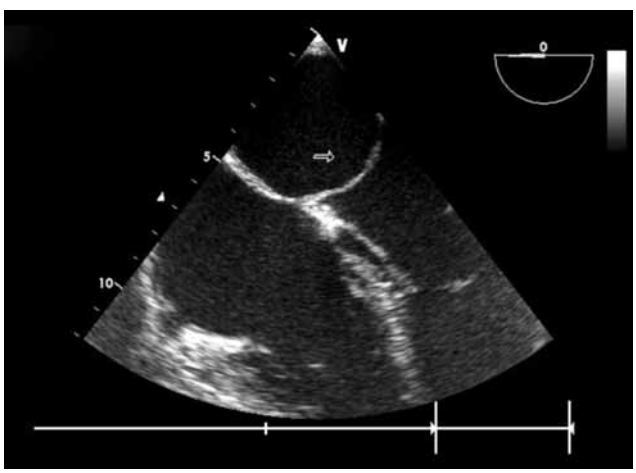

Figure 1. A transesophageal echocardiogram view reveals a linear echogenic density traversing the left atrium.

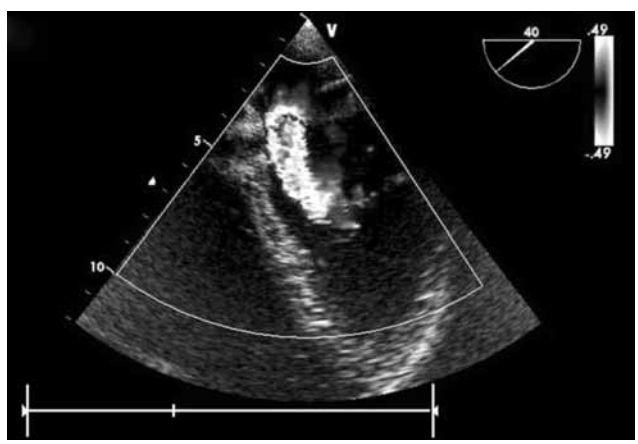

Figure 2. A transesophageal echocardiogram with color flow Doppler ultrasound shows a turbulent flow through an opening in the membrane near the intraatrial septum.

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