Cor Triatriatum with Ankylosing Spondylitis

Ankilozan Spondilit ile Birlikte Cor Triatriatum

M. Onur Hanedan, A. İhsan Parlar, Seyhan Babaroğlu, Kerem Yay, Garip Altıntaş
Türkiye Yüksek İhtisas Hospital, Cardiovascular Surgery Clinic, Ankara, Turkey

Özet

Cor triatriatum özellikle sol atrium olmak üzere atrial boşluklardan birinin fibromusküler membran ile ikiye bölünmesiyle oluşan nadir bir konjenital defekttir. Ankilozan spondilit kardiyak tutulumunda görülebildiği kronik sistemik inflamatuvar romatizmal hastalıktır. Ankilozan spondilit ile takip edilen 27 yaşında kadın, kardiyak tutulum için taranırken, cor triatriatum ekokardiyografi ile teşhis edildi ve tanı MRI ve CT anjio ile doğrulandı. Membran kardiyoplejik arrest ile kardiyopulmoner baypas altında cerrahi olarak çıkarıldı.Hasta şifa ile hastaneden taburcu edildi. Sonuç olarak, Ankilozan spondilit'li hasta kardiyak tutulum için taranmalıdır.

Anahtar Kelimeler

Cor Triatriatum; Ankilozan Spondilit

Abstract

Cor triatriatum is a rare congenital defect that one of the atrial chambers especially left atrium, is divided into two by a fibromuscular membranee. Ankylosing spondylitis is a chronic systemic inflammatory rheumatic disorder that cardiovascular involvement can be seen. 27-year-old woman, who has been followed for ankylosing spondylitis searched for cardiac involvement, Cor triatriatum was diagnosed with echocardiography and diagnosis was confirmed with MRI and CT angiography. Membrane was resected surgically under cardiopulmonary bypass with cardioplegic arrest. Patient was discharged from the hospital with recovery. In conclusion, patient with ankylosing spondylitis must be searched for cardiac involvement.

Keywords

Cor Triatriatum; Ankylosing Spondylitis

DOI: 10.4328/JCAM.1670 Received: 08.02.2013 Accepted: 28.02.2013 Printed: 01.04.2016 J Clin Anal Med 2016;7(suppl 2): 85-6
Corresponding Author: Kerem Yay, Türkiye Yüksek İhtisas Hospital, Cardiovascular Surgery Clinic, 06100, Sıhhıye, Ankara, Turkey.
T.: +905054587576 F.: +90 3122290148 E-Mail: keremyay@gmail.com

Introduction

Cor triatriatum is a rare congenital defect that one of the atrial chambers is divided into two by a fibromuscular membrane. It is found 0.4% in a series of 474 patients' autopsy with congenital heart defect (1). More often patients present in infancy but some cases can remain asymptomatic until adulthood. Morphology of cor triatriatum left atrium division is more common. Cor triatriatum sinister is only 0.1% of the congenital heart diseases (2).

Ankylosing spondylitis is a chronic systemic inflammatory rheumatic disorder that mainly involves young males and primarily effects on axial skeleton. Peripheral joints, tendons and ligaments may be affected. Uveitis, cardiovascular and pulmonary involvement may also occurs (3). Cardiovascular involvement is found 42.5% in a retrospective study (4). Aortic insufficiency, atrio-ventricular block, bundle branch block, Wolff-Parkinson-White syndrome, short PR syndrome, ischemic heart disease, mitral regurgitation, diastolic dysfunction (left ventricular dysfunction), tachy-brady syndrome, atrial fibrillation are cardiac involvement (4, 5). However, coexisting with cor triatriatum did not reported previously.

Case Report

27-year-old woman, who has been followed for ankylosing spondylitis from March 2006 with pelvic articulation involvement, has no cardiac symptoms. On examination, the patient was found to be normotensive with a regular pulse and a normal electrocardiogram. A transthoracic echocardiography was performed to search for, if there is any cardiac involvement of ankylosing spondylitis. It is showed that there was a membrane below 1.3 cm of the mitral valve and had an 11/5 mmHg gradient, 2 degree mitral insufficiency with a 45 mmHg systolic pulmonary artery pressure. After that, a transoesophageal echocardiography was performed which showed a membrane in the left atrium between left atrial appendix and left upper pulmonary vein (Figure 1). Cardiac magnetic resonance imaging and cardiac computed tomography angiography were performed, both were reported as cor triatriatum sinister (Figure 2a and 2b).

Surgical Technique

In the general anesthesia, median sternotomy was performed. After the heparin was administered, total cardiopulmonary bypass (CPB) was established between the ascending aorta and both superior vena cava and inferior vena cava. Under moderate hypothermic conditions, CPB flow rate was maintained at 2.5 L/min/m², aorta was cross-clamped and cardiac arrest was maintained with antegrade cardioplegia. Left atriotomy was performed and a membrane, which had 0.5 cm2 opening, was noticed in the left atrium. Membrane was resected. Mitral valve was competent. There was no atrial septal defect. Left atriotomy was closed. Intraoperative transesophageal echocardiography was performed which shown no abnormalities after resection . Postoperative period was uneventful and patient was discharged from the hospital with healthy.

Discussion

Cor triatriatum is a rare congenital defect. Morphologically left atrium division is more common. More often patients present in infancy, but some cases can remain asymptomatic until adulthood (1,2). Ankylosing spondylitis is a chronic systemic inflammatory disorder that cardiovascular involvement can be seen (3). This involvement was reported about 42.5% in a retrospective study (4). This involvement generally makes valvular insufficiency and rhythm disorders due to involvement of conduction system in the heart. This is the first case of cor triatriatum sinister with ankylosing spondylitis in the literature.

It is important to search ankylosing spondylitis patients for cardiac involvement. Electrocardiography and echocardiography must be performed to search for any arrhythmias', possible valve abnormalities, and congenital disorders (5).

The treatment is surgery when cor triatriatum is found. Surgical treatment of cor triatriatum is a membrane resection and repair of concomitant abnormalities.

In conclusion, patient with ankylosing spondylitis must be searched for cardiac involvement.

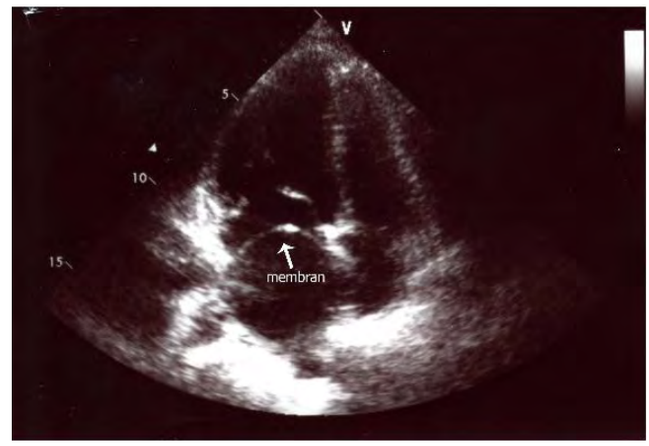


Figure 1. View of the preoperatif transthoracic echocardiography, arrow indicate membrane in the left atrium.

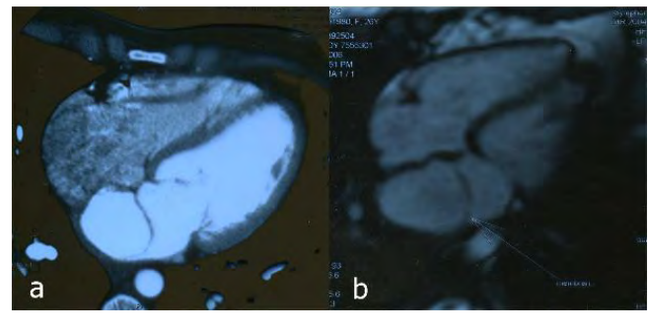


Figure 2. View of the membrane with Magnetic Resonance Imaging (A) and Computed Tomography Angiography (B).

Competing interests

The authors declare that they have no competing interests.

References

1. Kutsal A. Cor triatriatum. In: Duran E, editor. Kalp ve Damar Cerrahisi, 1st edition. Çapa Tıp Kitapevi: İstanbul; 2004. p. 251-61.
2. Von Son JA, Danielson GK, Schaff HV, Puga FJ, Seward JB, Hagler DJ, Mair DD. Cor triatriatum: diagnosis, operative approach and late results. Mayo Clin proc 1993; 68: 854-9.
3. Khan MA. Clinical features of ankylosing spondylitis. In: Hochberg MC, Silman AJ, Smolen JS, Weinblatt ME, Weisman MH, Editors. Rheumatology, 3rd edition. Mosby, London; 2003. p 1161-81.
4. Sukenik S, Pras A, Buskila D, Katz A, Snir Y, Horowitz J. Cardiovascular manifestation of ankylosing spondylitis. Clin Rheumatol. 1987; 6(4): 588-92.
5. Brunner f, Kunz A, Weber U, Kissling R. Ankylosing spondylitis and heart abnormalities: do cardiac conduction disorders, valve regurgitation and diastolic dysfunction occur more often in male patients with diagnosed ankylosing spondylitis for over 15 years than in the normal population? Clin Rheumatol 2006; 25(1): 24-9.

How to cite this article:
Hanedan MO, Parlar Aİ, Babaroğlu S, Yay K, Altıntaş G. Cor Triatriatum with Ankylosing Spondylitis. J Clin Anal Med 2016;7(suppl 2): 85-6.