Journal of Surgery and Medicine

An idiopathic aorta-right atrial fistula: A rare case

İdiopatik aorta-sağ atrial fistül: Nadir bir olgu

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Informed Consent: The authors stated that the written consent was obtained from the patient presented with images in the study. Hasta Onami: Yazar calismada görüntüleri sunulan hastadan yazılı onam alındığını ifade etmiştir.

Conflict of Interest: No conflict of interest was declared by the authors Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemislerdir.

Financial Disclosure: The authors declared that this study has received no financial support. Finansal Destek: Yazarlar bu calışma için finansal destek almadıklarını beyan etmişlerdir.

> Published: 2/29/2020 Yayın Tarihi: 29.02.2020

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Abstract

Atrial fistulas are the non-natural and rarely seen connections between the left atrium and ascending aorta. This pathological connection can be classified as anterior or posterior according to its location. It can be distinguished angiographically by the absence of myocardial branches. With the effect of systemic pressure, a tunnel occurs from the congenitally weak area in the aortic structure to the low-pressure atrium. There may be no symptoms in the patients, or serious clinical findings, including those related to congestive heart failure may be seen. Even if aorto-atrial fistulas are asymptomatic or hemodynamically insignificant, they should be treated to prevent possible complications. Closure can be performed with the catheterization method or open-heart surgery when appropriate. In this case report, we would like to share the treatment approach of a 49-year-old male patient who was admitted to our hospital with chest pain and whose angiographic and echocardiographic examinations revealed a tunnel from the aorta to the atrium. An informed consent was obtained from the patient.

Keywords: Aorta-atrial fistula

Öz

Aorta atriyal fistüller nadir olarak görülen sol atriyum ile asendan aorta arasındaki doğal olmayan bağlantılardır. Bu patolojik bağlantı yerleşim yerine göre anterior yeya posterior olarak sınıflandırılabilir. Miyokardiyal dalların olmaması ile anjiografide ayırt edilebilir. Sistemik basıncın etkisi ile aort yapısındaki konjenital zayıf olan bir bölgeden düşük basınçlı atriyuma tüne açlması durumudur. Hastalarda semptom olmayacağı gibi konjestif kalp yetmezliğine bağlı semtomlar dahil olmak üzere ciddi klinik yansımaları olabilir. Asemptomatik veya hemodinamik açıdan önemsiz olsa bile olası komplikasyonları nedeniyle tedavi edilmeleri gerekmektedir. Kateterizasyon yöntemi ile yapılabileceği gibi uygun hastalarda açık kalp cerrahisi ile kapama işlemi yapılabilmektedir. Burada 49 yaşında olan, göğüs ağrısı ile hastanemize başvuran, yapılan anjiografik inceleme ve ekokardiyografik değerlendirmede aortadan atriyuma geçiş tespit edilen hasta için yaptığımız tedavi yaklaşımını kendisinin onayı alındıktan sonra paylaşmak istedik. Anahtar kelimeler: Aorta-atrival fistül

Introduction

Tunneling from the aorta to the right atrium is a very rare vascular pathology. In this pathology, there is a shunt arising from the aortic root and terminating in the right atrium. It may occur as a complication of infective endocarditis, mitral valve insufficiency, aortic valve insufficiency, transseptal catheterization, connective tissue diseases, and aortic dissection [1,2]. It is usually detected incidentally during an imaging process and no spontaneous closure was reported [2]. In the period from diagnosis to treatment, it requires a multidisciplinary approach and cardiovascular team co-operation.

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Case presentation

A 49-year-old male patient was admitted to our hospital with long-standing chest pain. The patient had no diagnosis of chronic illness in his medical history. There was no risk factor other than smoking. Physical examination of the cardiovascular system revealed no other pathological finding except a precordial murmur. Other system examinations were normal. In twodimensional echocardiographic examination of the patient with normal sinus rhythm, the presence of an anteriorly located tunnel from the aortic root to the right atrium was detected (Figure 1a-1b).



Figure 1a: Two-dimensional echocardiographic image

Figure 1b: Two-dimensional echocardiographic image

Transesophageal echocardiography was performed for a clearer evaluation of the tunnel in the patient whose ejection fraction was 60%, and the shunt from the right sinus of Valsalva to the right atrium was observed. His biochemical parameters were within normal range. Coronary angiography via the right radial artery was performed to evaluate coronary vascularization. No pathology was detected in the main vascular structures. The tunnel structure did not contain myocardial branches. After the joint evaluation with the cardiology clinic, the decision to surgically intervene was made, and routine cardiac surgery preparations were started. After pre-operative preparations, the surgery was performed. The mediastinum was reached by median sternotomy. Cardiopulmonary bypass (CPB) was established after aorto-bicaval cannulation. After retrograde aortotomy, cardiac arrest was achieved with cold blood selective and antegrade cardioplegia from the coronary ostium, and it was continued intermittently. The tunnel structure extending from the right coronary sinus to the atrium was observed (Figure 2a, 2b).

The fistula mouth was closed with Teflon supported suture passing from the external aorta and the coronary sinus. Atriotomy was performed and the other end of the fistula was seen. The fistula mouth adjacent to the tricuspid valve was closed with teflon-pledget supported sutures. After venting maneuvers, the patient was separated from pump with no problem, and the cross clamp was removed. After the rhythm was back to normal, echocardiography was performed intraoperatively, and the absence of aorta-atrial transition was confirmed. Then, decannulation was performed with routine surgical procedures and surgery was completed. The patient, who was observed in the intensive care unit for 24 hours, had an uneventful postoperative period and he was discharged on the 5^{th} postoperative day. No aorta-atrial transition was observed with echocardiographic examination performed at 3^{rd} month.



Figure 2a: Intraoperative views of aorta-atrial tunneling



Figure 2b: Intraoperative views of aorta-atrial tunneling

Discussion

This pathology, which was first described by Coto et al. [3] and evaluated as a tunnel extending from the aortic root to the right atrium, is quite rarely seen. It frequently originates from the left sinus, and rarely from the right sinus [4]. In this case, the pathological structure extended from the right sinus to the right atrium. It was argued that a tunnel from congenitally weak area in the tunica media of aorta to low pressure right atrium may occur due to high pressure in the aorta [5]. It may occur due to the presence of an aneurysm in the sinus Valsalva, surgical interventions in the region, infections, aortic dissection, trauma, or idiopathically. The pathology in our case was evaluated as idiopathic since there were no other factors that might play a role in its etiology.

Since some patients can be asymptomatic, this pathology can be detected incidentally by imaging procedures. It can also be detected by the acute form with chest pain, or by the chronic form with congestive heart failure symptoms. Although transthoracic echocardiography is an important guide for the detection of this pathology in patients, the diagnosis becomes clearer with a rate of 97% by transesophageal echocardiography [6]. Coronary angiography and aortography are especially important for the definitive diagnosis and the evaluation of coronary vascular structures [7].

In the treatment options, catheter-supported closure methods and surgical closure are available according to the location and size of the lesion. In our case, since the tunnel in the right atrium was very close to the tricuspid valve, the surgical closure was considered appropriate after evaluation with the cardiology team. Even if the closure procedure is controversial in asymptomatic patients, it was reported that closure procedure is recommended to prevent the development of heart failure in these patients [8].

Conclusion

Aorto-atrial fistulas are rarely seen and important pathologies that can lead to heart failure if it is detected late or not treated. All facilities of the cardiology team should be used in diagnosis. In addition to transthoracic and transesophageal echocardiography, the coronary angiography and aortography should be performed to detect accompanying vascular pathologies. We hereby presented our case of fistula, which is quite rarely seen and extends from right sinus to the right atrium, and our treatment approach.

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Suggested citation: Patrias K. Citing medicine: the NLM style guide for authors, editors, and publishers [Internet]. 2nd ed. Wendling DL, technical editor. Bethesda (MD): National Library of Medicine (US); 2007-[updated 2015 Oct 2; cited Year Month Day]. Available from: http://www.nlm.nih.gov/citingmedicine