

A Rare Complication of Left Atrial Dilatation in Childhood: Vocal Cord Paralysis and Ortner Syndrome

Çocukluk Çağı Sol Atriyal Dilatasyonun Nadir Bir Komplikasyonu: Vokal Kord Paralizi ve Ortner Sendromu

Dilek GİRAY¹, Kemal GÖRÜR², Murat ÖZEREN³, Derya KARPUZ⁴, Olgu HALLIOĞLU

¹ Van Education and Research Hospital, Turkish Republic Ministry of Health, Department of Pediatric Cardiology, Van.

²Mersin University, Department of Otolaryngology, Mersin

³Mersin University, Department of Cardiovascular Surgery, Mersin

⁴Mersin University, Department of Pediatric Cardiology, Mersin

Geliş Tarihi: 04.11.2019, Kabul Tarihi: 17.12.2019

ÖZET

Bu yazıda, çocukluk çağında literatürde sıklıkla bildirilmeyen, rekürren romatizmal kardit ve buna ikincil sol atriyal dilatasyona bağlı Ortner sendromu gelişen hasta sunuldu. Sydenham koresi tanısı olan ve penisilin profilaksisini kullanmayan 15 yaşındaki kız çocuğu tekrarlayan romatizmal kardit atağı ile başvurdu. Hasta yatırıldı ve oral steroid tedavisi ile intramusküler benzatin profilaksisi başladı. Ayrıca hastanın yatışı sonrası başlayan ses kısıklığı, sıvı gıda alımı sonrası öksürük ve hematez yakınmaları vardı. Yapılan laringoskopik muayenede sol vokal kord paralizi saptandı. Hastaya vokal kord enjeksiyonu yapıldı ve sıvı alımı sonrası öksürüğü geriledi, ancak ses kısıklığı devam etti. Sol atriyal basıncı azaltmak ve kalıcı laringeal sinir hasarı gelişimini önlemek için mitral ve aort kapak replasmanı yapıldı. Ortner sendromu nadir görülebilmesine rağmen, ses kısıklığı nedeniyle başvuran çocuklarda da akılda tutulmalıdır.

Anahtar Kelimeler: Sol atriyal genişleme, Ortner sendromu, romatizmal kardit, Sydenham koresi.

ABSTRACT

In this report, a patient was presented with the diagnosis of Ortner syndrome secondary to recurrent rheumatic carditis and left atrial dilatation, which was not commonly reported in the literature in childhood. A 15-year-old girl with Sydenham's chorea who did not use penicillin prophylaxis was presented with recurrent rheumatic carditis attack. She was hospitalized and oral steroid therapy and intramuscular benzathine prophylaxis was initiated immediately. She also had the complaints of hoarseness, cough after liquid intake, and hematemesis after her hospitalization. In the laryngoscopic examination left vocal cord paralysis was detected. Vocal cord injection was performed and her cough after liquid intake regressed, but hoarseness did not. Mitral and aortic valve replacement was performed in order to decrease left atrial pressure and to prevent the development of permanent laryngeal nerve damage. Ortner syndrome although seen rarely should be kept in mind also in children who have been admitted because of hoarseness.

Key words: Left atrial enlargement, Ortner syndrome, rheumatic carditis, Sydenham's chorea.

INTRODUCTION

Left-sided vocal cord paralysis and hoarseness developing as a result of compression of the recurrent laryngeal nerve secondary to cardiac pathologies, such as left atrial dilatation, aortic aneurysm or pulmonary hypertension, between the pulmonary artery and the aorta is called Ortner syndrome. In this report, a 15-year-old girl who developed Ortner syndrome secondary to recurrent rheumatic carditis and left atrial dilatation was presented since it was rarely reported in the literature in childhood.

CASE

The patient who was diagnosed as Sydenham's chorea because of weakness and involuntary movements in the extremities three years before the last admission, was presented with recurrent rheumatic carditis attack and she did not use the prophylaxis. The patient had aortic (3rd degree) and mitral (4th degree) valvular regurgitation, the left atrium and ventricle were markedly dilated in echocardiography (figure 1 and figure 2). Cardiac contraction and valvular movements of her heart were decreased, ejection fraction was calculated as 45% in M-mode echocardiography. Steroid and congestive heart failure treatment was started to the patient, but she had the complaints of hoarseness, cough after liquid, and hematemesis. The endoscopic examination was performed by the gastroenterology department and the upper gastrointestinal system was evaluated as normal. Laryngoscopic examination was performed due to hoarseness and all laryngeal structures were found normal other than left vocal cord paralysis (figure 3). Vocal cord injection was performed by using calcium hydroxyapatite (radiess) material, the only long-acting injectable laryngeal material approved by the FDA, as the temporary treatment and aspiration attacks were improved despite hoarseness. Because of the severe regurgitations of mitral and aortic valves, two valve replacement was performed to reduce left atrial pressure and to prevent the development of permanent laryngeal nerve damage. In the surgery the recurrent laryngeal nerve on the left side was seen close to the left atrium and ventricle there was evidence of stretch injury on pericardium on the left side. Hoarseness and the other complaints of the patient regressed consecutively within a month after surgery.

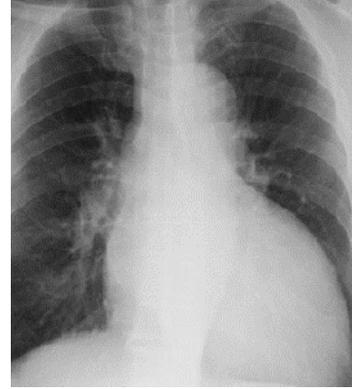


Figure 1: Chest x ray of the patient showing marked left heart dilatation

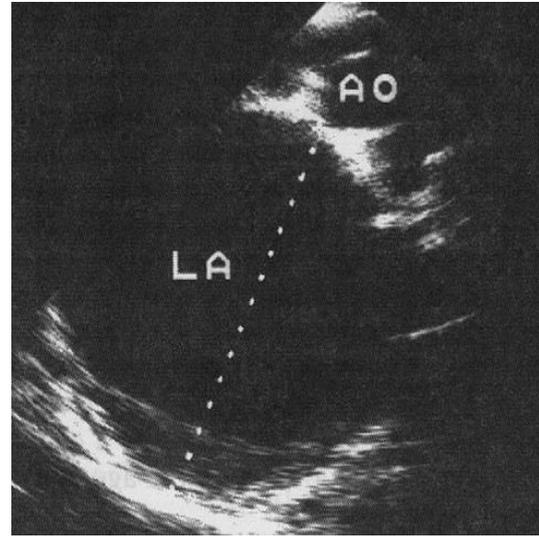


Figure 2: Echocardiographic imaging of the patient showing marked left heart dilatation



Figure 3: Laryngoscopic examination imaging of the patient showing left vocal cord paralysis.

DISCUSSION

Vocal cord paralysis may occur due to intralaryngeal or more frequently extralaryngeal pathologies. The most common two causes of unilateral vocal cord paralysis are intra-thoracic malignancies causing mediastinal lymphadenopathies, and surgical interventions (Karataş et al., 2006). Ortner syndrome was first described by Ortner in 1987 as a compression of the recurrent laryngeal nerve between the enlarged left atrium and the aortic arch in a patient with mitral stenosis (Ortner, 1987). However, it was accepted as a hoarseness due to compression of the recurrent laryngeal nerve that innervates the vocal cords between the pulmonary artery and the aorta or aortic ligaments (Thirlwall, 1997). Ortner syndrome, also known as cardio-vocal syndrome, is not as common in childhood as in adults. Congenital heart diseases such as atrial septal defect, ventricular septal defect, patent ductus arteriosus, total abnormal pulmonary venous return, idiopathic pulmonary hypertension have been reported as the most common causes of Ortner syndrome in the pediatric population.

Rheumatic mitral stenosis has been shown to be the most common cause in older children (Karataş et al., 2006). In this case, the etiology of the disease was recurrent rheumatic carditis leading to left atrial dilatation.

Although it is mainly hoarseness but dyspnea, hemoptysis, chest pain, cough are the other common symptoms of the disease. In our case, hoarseness, cough and hematemesis were the symptoms developed during the course of the treatment of rheumatic carditis.

Imaging methods have an important role in the diagnostic algorithms of causes of vocal cord paralysis (Yuan, 2014). Contrasted thorax tomography showed no pathology other than left atrial dilatation in accordance with the echocardiographic examination in our patient.

The main aim of the treatment is the elimination of the pressure on the nerve. In our patient, contractility of the heart returned to the normal with the treatment of heart failure, but patient's symptoms did not regress. In the laryngoscopic examination left vocal cord paralysis was detected and prevention of permanent nerve injury was planned the patient had aortic and mitral valve replacement. There was a marked reduction in the

left atrium diameter after the surgery and the hoarseness of the patient regressed.

The hoarseness caused by isolated left atrial dilatation as a result of recurrent rheumatic carditis in children has not been previously described in the literature. However, few cases with similar complaints in different cardiac diseases have been reported in the adult age group (Özyurtlu et al., 2013). Ortner syndrome should be kept in mind also in children who have been admitted because of hoarseness but who have left atrial dilatation on echocardiography and who do not have any pathological findings on laryngoscopic examination other than vocal cord paralysis.

REFERENCES

- Karataş A, Altıntaş O, Bilgili M, Veyseller B, İpek A. Aort anevrizmasına bağlı sol vokal kord paralizi (Ortner Sendromu- Kardiyovokal Sendrom). *Türk Klin J Cardiovasc Sci* 2006;18:258-60.
- Ortner N. Recurrent laryngeal nerve paralysis due to mitral valve stenosis. *Wien Klin Wochenschr* 1897; 10: 753-5.
- Thirlwall AS. Ortner's syndrome: A centenary review of unilateral recurrent laryngeal nerve palsy secondary to cardiothoracic disease. *J Laryngol Otol* 1997; 111: 869-71.
- Yuan S. Ortner (Cardio-vocal) Syndrome: A collective review. *Kuwait Med J*. 2014;46(1):3-13.
- Özyurtlu F, Acet H, Bilik MZ, Tasal A. Dev disekan aort anevrizmasına bağlı Ortner sendromu. *Türk Kardiyol Dern Arş* 2013;41(3):225-7.