



A CASE OF DOUBLE AORTIC ARCUS

ÇİFT ARKUS AORTA SAPTANAN BİR OLGU

Remziye TANAÇ¹ Esen DEMİR¹ Figen GÜLEN¹ Ayşe YENİGÜN¹ Demet CAN¹
Aytül PARLAR² Ruhi ÖZYÜREK² Ertürk LEVENT² Alp ALAYUNT³

¹Ege University Faculty of Medicine, Department of Pediatrics, Division of Allergy and Pulmonology, İzmir

²Ege University Faculty of Medicine, Department of Pediatrics, Division of Cardiology, İzmir

³Ege University Faculty of Medicine, Department of Cardiovascular Surgery, İzmir

Anahtar Sözcükler : Vasküler ring, çift aortic ark, trakeoözofageal obstrüksiyon

Key Words : Vascular ring, double aortic arch, tracheoesophageal obstruction

SUMMARY

Abnormalities of vascular ring which constitute 0.7 % of the congenital heart diseases are an important cause of tracheoesophageal obstruction in neonates and in early infancy. The symptoms may be life-threatening due to the degree of compression. We present here a 5-month-old boy presenting with the complaints of cough and noisy respiration. He experienced cyanosis and respiratory failure requiring ventilatory support and rapidly hospitalised. Thorax HRCT demonstrated that descending aorta was at the right paravertebral area. Barium scan of esophageus and Thorax Magnetic Resonance Imaging (MRI) led to the diagnosis of double aortic arcus. Right aortic arcus was larger than the left at cardiac catheterisation. After surgery the symptoms improved strikingly.

We conclude that vascular ring should be considered in the patients presenting with recurrent pulmonary infections, inspiratory stridor, and dysphagia. Early diagnosis and treatment may prevent chronic, irreversible complications.

ÖZET

Konjenital kalp hastalıklarının %0.7 sini oluşturan vasküler ring anomalileri, yenidoğanlarda ve erken bebeklik döneminde trakeo-özofageal obstrüksiyonun önemli bir nedenidir. Trakea ve özofagusa olan basının derecesine bağlı olarak, belirtiler yaşamı tehdit edecek kadar şiddetli olabilir. İlk kez 5.5 aylıkken öksürük, hırıltılı solunum yakınmalarıyla başvuran erkek olgu, siyanoz ve solunum yetmezliği gelişmesi üzerine entübe edilip, ventilatörde izlendi. Çekilen Toraks HRCT de, desenden aorta sağ paravertebral alanda izlenmesi üzerine, vasküler bir patoloji olabileceği düşünülerek, Baryumlu özofagus grafisi ve Toraks MR istendi. Çift aortic ark saptanan olgunun yapılan kardiyak kateterizasyonunda sağ tarafın daha gelişmiş olduğu izlendi. Cerrahi girişim uygulanan hastanın, sol arkus aortası divize edildikten sonra semptomları belirgin geriledi.

Sık akciğer enfeksiyon öyküsü olan, özellikle inspiratuar stridor, yutma güçlüğü olan olgularda vasküler ring mutlaka akla gelmelidir. Kronik ve irreversibl komplikasyonların önlenmesi, erken tanı ve tedavi edilmesi ile mümkün olacaktır.

INTRODUCTION

Abnormalities of vascular ring which constitute 0.7 % of the congenital heart diseases are an important cause of tracheoesophageal obstruction in neonates and in early infancy (1,2). The absolute incidence is not known because of asymptomatic cases and since incomplete types tend to occur more frequently than complete ones.

Clinical findings may be a barking cough, a noisy breathing, inspiratory stridor, dyspnea, dysphagia, aspiration pneumonia and frequent lung infections in early infancy. Feeding problems that become predominant with the introduction of the solid food are secondary to difficulties in the passage of food through esophageus. Symptoms may be severe and life threatening due to the degree of the compression to trachea and esophageus (3-5).

Yazışma adresi : Figen GÜLEN, Ege University Faculty of Medicine, Department of Pediatrics, Division of Allergy and Pulmonology, İzmir
Makalenin geliş tarihi : 21.12.2003 ; kabul tarihi : 02.02.2005

CASE REPORT

A 5-month-old boy hospitalized due to cough, noisy breathing and respiratory distress required ventilatory support following the development of cyanosis and respiratory failure. Auscultation of the lungs revealed rhonchi and prolonged expiration. Liver was palpable three centimetres below the costal margin though spleen was not. The examinations of the other systems including the cardiac system were found to be normal. He experienced an upper respiratory tract infection when he was 4 months old.

Laboratory analysis: Leukocyte count: 15700/mm³ (polymorphonuclear cells 60 %, lymphocytes 38 % and stabs 2 %). Erythrocyte sedimentation rate: 45 millimetres per hour.

C-reactive protein was negative and there was no bacterial growth in the blood cultures. Bilateral hyperaeration of lungs in the PA chest X ray was accompanied by the parallel ribs, flat diaphragms the image of a subatelectasis in right middle lobe. Immediate treatment of the patient was instuted as inhalation salbutamol, ipratropium bromide and parenteral dexamethasone. Prolonged course of respiratory distress required administration of TPN. Aminophylline infusion was also added to the treatment. High Resolution Computed Tomography (HRCT) of the thorax performed following improvement in clinical finding. HRCT showed that descending aorta was in the paravertebral area of the right lung and parenchyme of the lungs was completely normal. Barium scan of esophageus revealed compression of esophageus at the level of arcus aorta posteriorly (Figure 1).



Figure 1. Barium scan of esophageus

Thorax MRI displayed the double aortic arcus encircling the trachea and narrowing its lumen moderately just before the level of bifurcation (Figure 2).



Figure 2. Thorax MRI

Cardiac catheterization demonstrated that the left common carotid and the left subclavian arteries branched at the level of bifurcation just before left aortic arc joined the right one respectively. In other words, right and left aortic arcs were joining after the branching point of the subclavian arteries.

The right arch was larger than the other one. The patient could be operated at the age of 2 and a half due to the poor control and socio-economical causes. The symptoms strikingly lessened after the left arcus aorta was divided. He is still follow-up our outpatient clinic (Pediatric Allergy and Respiratory Diseases).

DISCUSSION

The simplest way to understand the anatomy and development of double aortic arch and other forms of vascular ring is to begin by considering the bilateral system of pharyngeal arch vessels in the early embryo. Early in the course of embryonic morphogenesis, 6 pairs of pharyngeal arch arteries develop in conjunction with the branchial pouches. The segments of the bilateral aortic arch system that normally regress include the distal portion of the sixth arch and the right sided dorsal aorta. Vascular rings are formed when this process of regression and persistence does not occur normally, and the resulting vascular anatomy completely encircles the trachea and

esophageus. A double aortic arch is formed when both fourth arches and both dorsal aortas remain present. In the presence of double aortic arcus, a complete abnormality of vascular ring, ascending aorta separates into two branches (right and left) around the trachea and esophageus which assemble again as a single descending aorta at the dorsal side (1,3).

Vascular rings have been well documented to cause respiratory and gastrointestinal symptoms in infants and children. Vascular rings are subclassified as double aortic arch, right arch/left ligamentum arteriosus, pulmonary artery sling and innominate compression (6). Double aortic arch is a rare vascular anomaly which causes tracheal and esophageal compression usually in the first months of life. Typical symptoms in the early childhood should lead to prompt diagnosis and surgical treatment of this malformation. In adults this anomaly is extremely rare. A case of a severely symptomatic 29 year-old woman was presented (7).

In cases presenting with inspiratory stridor and difficulties in swallowing, posteroanterior chest radiographs may demonstrate the presence of emphysematous changes, a widened mediastinum, and a compression of the trachea from right or left sides. Lateral graphs may reveal a sign of compression of the anterior part of the trachea. Roentgenographic examination of the barium scan of esophageus is the useful non-invasive technique leading to diagnosis (1,5,8). Compression of the esophageus from different sides can be seen. Echocardiography, magnetic resonance imaging and computed spiral tomography are the other helpful diagnostic methods. Angiography is needed to confirm the diagnosis and to prepare the patient

for surgery. Aortography leads to the accurate diagnosis (1,3,8).

Surgery is indicated if symptoms are severe or life-threatening of the patient. Infancy is the most appropriate period for operation. The fundamental principle of surgical management of double aortic arch is division of the ring to relieve compression of the trachea and esophageus. In general, this is achieved by dividing the minor arch through an ipsilateral thoracotomy. When the minor arch is atretic, the atretic segment is ligated or clipped and then divided. When the minor arch is patent, it usually is ligated and divided between the subclavian artery and descending aorta (1,3,4,5).

Magnetic resonance imaging has become the standard means of imaging paediatric airway obstruction due to vascular anomalies. The development of spiral or helical computed tomography provides an alternative imaging modality for evaluating paediatric airway obstruction (9).

A retrospective study of 45 cases operated due to tracheobronchial compromise secondary to vascular compression at a large children's hospital between July 1983 and February 1996. Revealed that 34 of these had innominate artery compression, ten had with a double aortic arch and one had with an anomalous right subclavian artery (10).

As a conclusion, we suggest that abnormalities of vascular ring should be considered in the differential diagnosis patients presenting with frequent pulmonary infections, inspiratory stridor and dysphagia. Early diagnosis and treatment may prevent chronic, irreversible complications.

REFERENCES

1. Park SC, Zuberbuhler JR. Vascular ring and pulmonary sling. In Anderson RH, Maccartney FJ, Shinebourne EA, Tynan M (ed). *Pediatric Cardiology* (1st ed). Edinburgh: Churchill Livingstone 1987; 1123-1137.
2. Goor DA, Lillehei CW. *Congenital Malformations of the heart*. Newyork: Grune and Stratton, (1st ed), 1975; 286-297.
3. Akçevin A. Arkus aorta anomalileri. Bozer Y (ed) *Kalp Hastalıkları ve Cerrahisi Cilt 1: Ankara*, 1985; 163-199.
4. Chun K, Colombani PM, Dudegon DL, Heller JA. Diagnosis and Management of Congenital Vascular Rings. A 22- Year Experience. *Ann Thorac. Surg.* 1992; 53: 597-603.
5. Han MT, Hall DF, Manche A, Rittenhouse EA. Double Aortic Arch Causing Tracheoesophageal Compression. *The American Journal of Surgery* 1993; 165: 628-631.
6. Backer CL, Mavroudis C. *Congenital Heart Surgery Nomenclature and Database Project: vascular rings, tracheal stenosis, pectus excavatum.* *Ann Thorac Surg* 2000 Apr; 69 (4 Suppl): 308-18.
7. Brockes C, Vogt PR, Roth TB, Turina J. Double aortic arch: diagnosis missed for 29 years. *Vasa* 2000 Feb; 29(1): 77-79.
8. Lillehei CW, Colan S. Echocardiography in the preoperative evaluation of vascular rings. *Journal of Pediatric Surgery* 1992; 27: 1118-1121.
9. Gustafson LM, Liu JH, Link DT, Strife JL, Cotton RT. Spiral CT versus MRI in neonatal airway evaluation. *Int J Pediatr Otorhinolaryngol* 2000 Apr 15; 52(2): 197-201.
10. Erwin EA, Gerber ME, Cotton RT. Vascular compression of the airway: indications for and results of surgical management. *Int J Pediatr Otorhinolaryngol* 1997 Jun 20; 40 (2-3):155-162.