A Case of Symptomatic Aberrant Right Subclavian Artery

Semptomatik Aberran Sağ Subklavian Arter Olgusu

Şit Uçar, MD.
Department of Pediatrics,
Doktor Sami Ulus Education and Research Hospital
situcar@gmail.com

Pelin Zorlu, MD.
Department of Pediatrics,
Doktor Sami Ulus Education and Research Hospital
pelin.zorlu@gmail.com

Atilla Çifci, MD.
Department of Pediatrics,
Doktor Sami Ulus Education and Research Hospital
dratillacifci@gmail.com

Abstract
Vascular rings are formed when there is a failure of normal regression of the embrionic arch segments resulting in a persistence of a vascular ring tissue surrounding the trachea and esophagus. Aberrant subclavian artery is congenital anomaly that usually does not produce symptoms. A patient with Aberrant right subclavian artery and wheezing, defined in early infancy, was presented.

Key words: **Congenital abnormalities; magnetic resonance angiography; subclavian artery.**

This study was presented in the Turkish Pediatrics Congress, 2006, Antalya, Turkey.

Özet
Arkus aortayı oluşturan embriyonik yapıların involüsyonundaki anormallikler nedeni ile oluşan vasküler ringler, çocukluk çağında trakea ve özofagusu sıkıştırarak solunum zorluğu ve yutma güçlüğüne neden olmaktadır. Aberran sağ subklavian arter, genellikle semptomlara yol açmayan konjenital bir anomalidir. Erken bebeklik döneminde wheezing ile birlikte olan aberran sağ subklavian arterli bir olgu sunuldu.

Anahtar kelimeler: **Konjenital anomali; manyetik rezonans anjiyografi; subklavian arter.**

Submitted : July 31, 2007
Revised : August 02, 2007
Accepted : September 05, 2007

Corresponding Author:
Şit Uçar, MD.
Department of Pediatrics,
Doktor Sami Ulus Education and Research Hospital
Ankara, Turkey

Telephone : +90 - 312 3056194
E-mail : situcar@gmail.com

Introduction

Vascular rings, congenital abnormality of arcus aorta, make various levels of pressure on trachea and esophagus, causing various symptoms to arise. They compose 1-2% of congenital cardiovascular abnormalities. Structural abnormalities of arcus aorta is more frequent compared to patients diagnosed. They can be asymptomatic when do not apply pressure on respiration routes. Vascular rings that cause pressure appear with life threatening clinical symptoms (1-2).

Vascular rings may present some symptoms in the early childhood period of the kids. The most important clinical symptoms are repeated stridor, wheezing, dysphasia, difficulty in nutrition and respiration based on pressure of trachea and esophagus (1-2). These are divided into two groups: complete and incomplete. Complete vascular rings are right arcus aorta with ligamentum arteriosum and double arcus aorta. These rings surround the circle of trachea and esophagus completly. Incomplete vascular rings are aberrant right subclavian artery, innominate artery and abnormal left pulmonary artery (vascular sling). These rings surround the circle of trachea and esophagus incompletly, but apply pressure on. Double arcus aorta is the most frequently seen type of vascular ring. Aberrant right subclavian artery composes 20% of all cases (2-4).

Case Report

A - 5- month - old male infant applied with the complaints of coughing, wheezing, rapid breathing that have been going on in the last two weeks and fever in the last three days. During the anemnesis, it was revealed that he had wheezing problem since birth and was hospitalized twice (two and four months old) with diagnose of bronchiolitis. His breathing complaints got not better during he was hospitalized and his complaints changed with position and were not related with nutrition. There was no another specific feature in his history.

In his physical examination, his general status was good and number of respirations was 64/min., pulse was 160/min., oxygen saturation was 90%, body temperature was 38^0C. Body weight-height-head perimeter percentiles was compatible with his age; intercostals and subcostal retractions were monitored. Wheezing, harder and longer exprium, bilateral crepitant rale and diffuse rhonchus were heard on auscultation of the chest. Other examination findings were normal.

On laboratory examinations, hemoglobin was 11.8 gr/dL, count of leukocytes was $18.9x10^3/mm^3$, count of thrombocytes was $386x10^3/mm^3$. There was left shift and in peripheral blood smear and CRP was detected to be 83 mg/L. In arterial blood gas light hypoxemia was detected and liver-kidney functions, blood sugar and serum electrolytes were found to be within normal limits. In double way lung graph, ventilation increase and perihilar-peribronchial infiltration was seen.

Antibiotic was administered to the patient with repeated wheezing, acute bronchiolitis and acute community acquired pneumonia. Oxygenization and hydration were ensured and bronchodilator treatment was applied. No response was obtained after bronchodilator, adrenalin and steroid treatments.

In examinations carried out in the previous hospitalization periods, ppd was determined to be negative. Sweat test was normal, tuberculosis PCR in hunger gastric juice was negative, family history for tuberculosis was normal, immunoglobulin M, G, A and E values were normal limits according to age. In reflux scintigraphy that was performed one month ago, reflux was determined and no specific response could be taken in gastroesophageal reflux treatment.

It was observed that congenital wheezing which could not be cured was less in lying position and became clear in sitting position. Upon this examination finding, congenital anatomic pathology was considered and esophagography with barium was taken. Indentation in esophagus and reflux in gastroesophagus were determined in posteroanterior and lateral esophagography with barium (Figure 1).

Right subclavian outlet was not observed in echocardiography. Pressure symptom in esophagus and absence of right subclavian outlet lead to consider vascular ring abnormality. Aberrant right subclavian artery was diagnosed with magnetic resonance angiography (Figure 2).

In the patient to whom catheter angiography was applied regarding right subclavian artery, after consultation to the committee, observation decision had been taken. During the 14 month observation period, acute bronchiolitis was experienced twice and the patient was monitored in the hospital once. Other than bronchiolitis attacks, it was seen that wheezing lessened with time and that his development was compatible with the age.

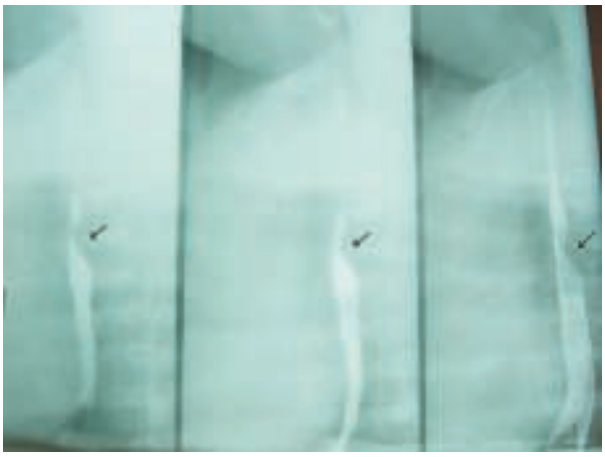

Figure 1. Oblique indentation is seen in esophagus at posterior-anterior and lateral esophagusgraphy with barium.

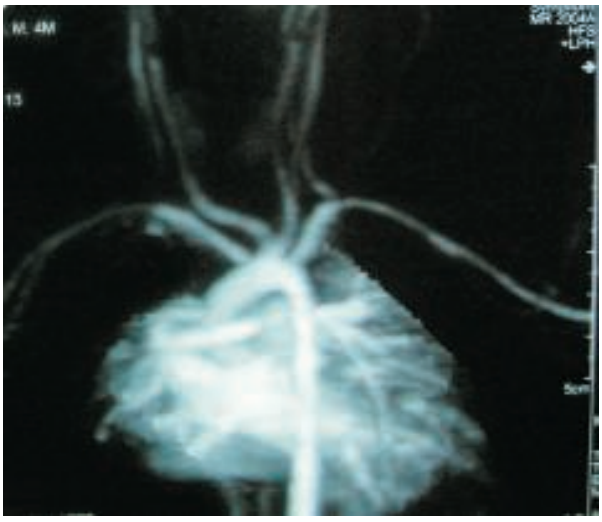

Figure 2. MR angiography image. Normal contours of ascending, descending, arcus aorta; right and left truncus arteries branched from the aorta as single truncus and divide to right and left branches after 0.5cm. It is seen that right subclavian artery aberrantly branched the arcus aorta and apply pressure on esophagus and trachea from the back.

Discussion

Repeated wheezing may be caused in breast feeding period due to many reasons. However early complaints, not having problem in perinatal period and not completely healing in intermediate periods lead to thinking of congenital respiration route abnormalities, gastro esophageal reflux disease, cystic fibrosis, tuberculosis, congenital heart and coronary disease (2- 4).

After therapy, no specific response was received for gastroesophageal reflux determined in reflux scintigraphy. Especially, complaints continued after birth and symptoms increased with position led to consideration of vascular ring abnormalities.

Vascular rings are resulted from abnormal involution of vascular structures forming arcus aorta or formation of involution in different spots than normal in embryonic life being. These vascular structures constrain trachea and/or esophagus and apply pressure, causing symptoms. Weight and onset of symptoms and findings depend on type of abnormality and pressure level on esophagus and trachea. Real frequency of the disease is not known due to asymptomatic cases however it is believed that this is higher than notified cases (2, 4, 5).

Vascular rings can be seen together with congenital intracardiac abnormalities or by themselves also. Most frequently observed cardiac abnormalities are tetralogy, transposition of larger arteries and aorta coarctation (6). Etiology of the disease is not well known. Teratogenic agents are accused however it is known that 22q11 deletion is frequently seen in isolated vascular ring abnormalities (7). In the case presented, no other cardiac abnormality accompanying right subcavian aberrant artery was detected.

Aberrant right subclavian artery occurs due to regression of right fourth arcus between carotid and subclavian artery Right subclavian artery lies towards the up and right side of the esophagus coming out of arcus aorta as fourth branch and forms a partial ring (5-6).

The most frequent complaints of patients are coughing, wheezing, difficulty in swallowing, stridor and especially manifest breathing difficulty in inspirium. Very early appearance of symptoms should make us think about the possibility of a congenital abnormality. When vascular ring does not apply any pressure on respiration routes, patients can be asymptomatic. However serious pressure creating vascular rings may appear along with life threatening clinical symptoms. Aberrant right subclavian artery cases are mainly asymptomatic in early period. Patients older than 1 years of age frequently admit to clinics with difficulty in swallowing problem. Some cases with obscure findings can be detected coincidently years after birth (2, 5, 8-10).

Since the presented case admitted with repeated wheezing in early infant period, it has been possible to diagnosis in early period with pressure symptoms.

In vascular ring cases, direction of arcus and pushing on trachea in anteroposterior lung graphy and narrowing of trachea lumen, flexure of trachea and retrotracheal opacity in lateral lung graphy can be monitored (11). Arcus direction can be determined in suprasternal long axis sections ecocardiographically. The fact that in presence of both right and left arcus, not being able to show bifurcation of brachiosephalic artery leads us to consider abberant right subclavian artery (2-6). In echocardiography of our case right subclavian outlet is not observed.

For the diagnosis, indentation in esophagus can be seen in posteroanterior and lateral esophagusgraphy with barium. Oblique indentation at the right subklavian artery is observed in lateral esophagusgraphy with barium (2-4). In the present case, these pressure finding were also seen in esophagusgraphy (Figure 1). MR angiography or computerized tomography (CT) angiography are advenced methods intended for definite diagnosis (12-13). In the diagnosis of vascular ring, the contribution of bronchoscope is restricted. Pre-operative catheter angiography may also be carried out in order to show the anatomic details (2, 6). The vascular ring are also diagnosed by using fetus echocardiograph (14).

Surgery is frequently suggested in symptomatic patients with vascular ring. Essential part of the surgical treatment is dividing the point where hypoplasic arcus intersects with aorta from proximal. Along with this, ligamentum arteriosum should be cut and all vascular structure shall be released and obstruction can be fully repaired. Videoscopic thoraxopic methods can also be applied In treatment of vascular rings (6, 9, 15, 16).

It was shown that when symptoms are not severe enough, surgical intervention does not increase the level of life quality. According to vascular ring type, vascular ring can be monitored clinically. The present case was taken under observation after catheterization and operation has been planned in case of symptoms continued. In some vascular ring cases, breathing problems after operation may continue. This condition results from tracheomalacia acquired because of long term pressure. Furthermore, it has been reported that cases with aberrant right subclavian artery aneurysm is reported (15, 16).

As a result, vascular rings should be considered and investigated in the diagnosis of patients that applied with breathing problems starting with early age, difficulty in breathing, wheezing, stridor, and repeated lower respiratory disease infection. Patients, whom indentation is determined in anteroposterior and lateral esophagusgraphy with barium, should also be evaluated with MR or CT angiography.

Kaynaklar

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