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CASE REPORT

Coarctation of the Aorta Presenting with Fever and Abdominal Pain: A Case Report

Ateş ve Karın Ağrısı ile Başvuran Aort Koarktasyonu: Olgu Sunumu

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ABSTRACT

Aortic coarctation is among the congenital heart disease which can easily be diagnosed in infancy and childhood by means of a deliberate physical examination and echocardiography. Children without a prompt diagnosis are represented with several complications that depend on the severity of coarctation. It is of vital importance to perform stent implantation, balloon dilatation or surgical repair as soon as possible after the diagnosis, because severe complications may emerge in aortic coarctation. The present report delineates the case of a six-year old patient who was presented with fever and abdominal pain, and diagnosed with echocardiography to have aortic coarctation.

Keywords: Aortic coarctation; child; hypertension

ÖZ

Aort koarktasyonu dikkatli bir fizik muayene ve ekokardiyografi ile bebeklik ve çocukluk çağında kolayca tanı konulabilen doğumsal kalp hastalıklarından biridir. Erken tanı alamayan hastalar koarktasyonunun şiddetine bağlı olarak bebeklik, çocukluk ve erişkin dönemde çeşitli komplikasyonlar ile başvururlar. Aort koarktasyonu olan vakalarda ağır komplikasyonlar gelişebileceği için aort koarktasyonu teşhis edilir edilmez vakaların yaşına ve koarktasyonun durumuna göre stent implantasyonu, balon dilatasyonu veya cerrahi onarım yapılması hayati önem taşımaktadır. Biz burada ateş ve karın ağrısı ile müracaat eden ve ekokardiyografide aort koarktasyonu saptanan altı yaşında bir hastayı sunduk.

Anahtar Kelimeler: Aort koarktasyonu; çocuk; hipertansiyon

Introduction

Aortic coarctation (AC) is prevalent at a rate of 5-8% in all age groups among congenital heart diseases (CHD) while it is 1.7 times more common in male (1). It occurs mostly at the level of the ductus arteriosus, and the length and degree of stenosis can vary. Although it is usually detected as isolated aortic coarctation, it is sometimes can be accompanied by other CHDs (2). The most common CHDs are bicuspid aortic valve, ventricular septal defect, transverse arch hypoplasia and aortic stenosis (3). AC pathophysiologically emerges in the fetal life even though it clinically can manifest at various post-natal ages. In the newborn, AC may present with heart failure and metabolic acidosis whereas findings such as hypertension, cerebrovascular disorders, and intermittent claudication can persist later on (1,4). Endovascular balloon dilatation, stent implantation, and open surgical interventions are contemporarily available treatment approaches (5,6). Prompt diagnosis and timely treatment are important as for prognosis. Here, we presented a case of a six-year-old boy who was hospitalized with the preliminary diagnosis of urinary tract infection and then diagnosed with aortic coarctation during etiological investigation of hypertension.

Case

A six-year-old male patient was admitted to the pediatric emergency department with complaints of fever (up to 40 oC) for four days and abdominal pain. On physical examination, there was tenderness, abdominal guarding and rebound in the right lower quadrant. Heart rate was 98/min, and body temperature was 38.6 oC. Laboratory findings were neutrophilic leukocytosis (WBC: 45430/mm3, neutrophil: 90%) and elevated C reactive protein (CRP)(195 mg/L). The patient was transferred to the pediatrics ward to be examined for the etiology of abdominal pain and fever. Samples for the complete urinalysis, routine biochemistry, and urine and blood culture were taken. Complete urine and biochemistry tests were normal. Imaging was planned to rule out acute abdomen for the patient who had abdominal guarding and rebound on physical examination. Abdominal and pelvic ultrasonography were found normal. Abdominal computed tomography imaging was planned for the patient. In abdominal tomography, right kidney dimensions were relatively lower compared to the left, and heterogeneous densities were observed in the upper pole of the right kidney which may be

considered consistent with infectious pathologies in case of the correlation with laboratory results. Empirical ceftriaxone treatment was started with a preliminary diagnosis of acute urinary tract infection. In the routine blood pressure measurement, the upper extremity blood pressure was 121/79 mmHg which is high for his age (95-99 p). Renal doppler ultrasonography was performed to investigate the etiology of hypertension other than renal parenchymal disease, and it was evaluated as normal. In the repeated examination of the patient whose upper and lower extremity systolic pressures varied more than 20 mmHg, femoral pulses could not be detected. Echocardiography was planned for the patient who was thought to have aortic coarctation. In echocardiography, aortic coarctation (gradient 51 mmHg, flow velocity 3.5 m/sec) and 5.5 mm secundum atrial septal defect (ASD) were detected (Fig 1,2). Enalapril maleate (1*2.5 mg) was started against hypertension. Due to the detection of Klebsiella spp. (100,000 colonies) producing extended-spectrum beta-lactamase (ESBL) in urine culture, ceftriaxone treatment was discontinued on the 2nd day, and ertapenem was started according to the culture-antibiogram results. In the follow-ups, their fever and acute phase reactants regressed (WBC: 7.240 /mm3, NEU: 27% CRP: 6.81 mg/L). Dimercapto succinic acid (DMSA) was applied to the patient following the completion of antibiotic treatment and detection of no growth in the control urine culture. In nuclear imaging, a hypoactive area secondary to pelvicalyceal dilatation was observed in the medial part of the left kidney. The right kidney was small in size and there was contour irregularity in its upper and lower poles which can be associated with renal parenchyma damage. In the voiding cystourethrography taken afterwards, stage 1 vesicourethral reflux was detected in the passive phase on the right. After the eradication of the urinary infection, balloon dilatation for aortic coarctation was performed in an external center. One month after the procedure, residual aortic coarctation (gradient: 27 mmHg) was observed in the control echocardiography. Blood pressure was measured at the upper limit of normal (110-120/70-80 mmHg [>95p]). The patient who received propranolol treatment instead of enalapril maleate which was discontinued at the operated center, was followed up clinically.

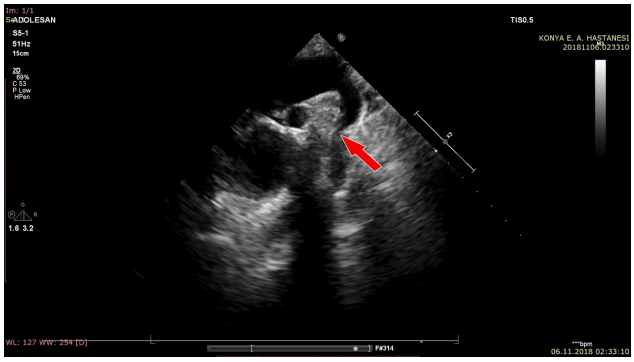

Fig 1: Coarctation of the aorta at the duct level in the aortic arch(arrow)

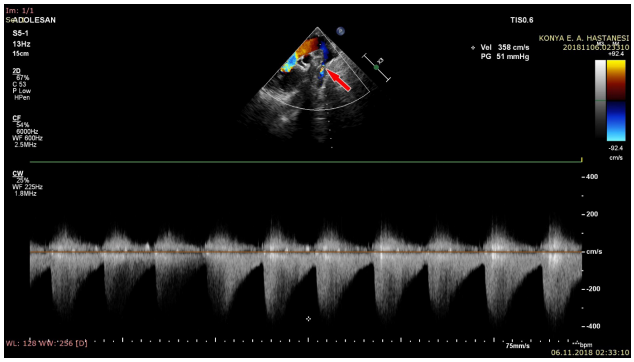

Fig 2. Mosaic image on color doppler in the region with aortic coarctation (arrow).

Discussion

Aortic coarctation (AC), described in the middle of the eighteenth century, may show diverse clinical presentations in different ages and patients (7). The pathophysiological process related to the disease starts from the fetal period. Therefore, early diagnosis and treatment of aortic coarctation is of great importance for survival and life quality.

Although AC is usually observed isolated or along with bicuspid aortic valve, ventricular septal defect, transverse arch hypoplasia, aortic aneurysm, double aortic arch, right descending aorta, persistent left superior vena cava, and abnormal pulmonary venous return anomalies may also accompany (2,3). In our patient, secundum atrial septal defect was observed accompanying aortic coarctation.

AC can manifest with various clinical symptoms and signs in different periods of life. While it usually presents with heart failure, acidosis and shock in infancy, in advanced ages; they present with symptoms and signs such as hypertension, cerebrovascular incidents, claudication, exercise intolerance, aortic dissection and rupture, weak or absent femoral pulses, systolic-diastolic murmur, hypertensive retinopathy, and cold feet (1,4). Unlike these, Alpsoy et al. (8) have presented a 41-year-old patient who was observed with atrial fibrillation and found that he had aortic coarctation. This once again shows how wide the range of symptoms and signs can be in adults. Our patient was incidentally diagnosed with aortic coarctation by the mean of transthoracic echocardiography which was utilized because hypertension was observed in the routine blood pressure measurement and a difference more than 20 mmHg between lower and upper extremities were noticed along with undetectable femoral pulses. Hypertension accompanies almost all acquired and congenital types of renal parenchymal disease and frequently causes decreased glomerular filtration rate (GFR) (9). In our patient, the aforementioned hypertension may have stemmed from existing renal parenchymal injury and aortic coarctation. However, renal parenchymal hypertension was ruled out primarily due to the pressure difference in the lower and upper extremities.

In patients with aortic coarctation, rib notching can be observed in the anteroposterior thorax radiogram because of collaterals that develop with advancing age. At the same time, pre- and post-stenotic dilation can be seen (9). Both findings were not found in our patient.

Since electrocardiography does not show any typical findings except for left ventricular hypertrophy, transthoracic echocardiography is the gold standard for diagnosis in newborns and infants. In adults, magnetic resonance/computerized tomography and hemodynamic tests may be more useful for planning the treatment (10,11). In our patient, transthoracic echocardiography was used for diagnosis. The coarctation gradient was 55 mmHg and there was severe stenosis. Even so, the patient who was six years old, did not have any complaints related to aortic coarctation, according to repeated detailed anamnesis following the diagnosis of aortic coarctation which was detected incidentally.

Today, endovascular balloon plasty-stent and open surgical repairment can successfully be performed in patients in need. Depending on the type of aortic coarctation, end-to-end anastomosis, extended end-to-end anastomosis, patch angioplasty or graft interposition techniques is used in open surgery (12). Patients undergone surgery should be followed for re-coarctation, aortic aneurysm, and persistent hypertension (5,6). The study by Koç et al. (12) in which 106 cases repaired with 4 different techniques were compiled, reported that re-coarctation was rarest in end-to-end (12.5%) and extended end-to-end (12.5%) anastomoses, and was most frequent in patch angioplasty (14.8%). In addition, all patients who developed re-coarctation were operated before the age of sixth months. In our patient, balloon angioplasty was performed and residual coarctation (gradient 25 mmHg) was observed. While there was reportedly no hypertension in the early follow-ups of the external center, the patient was noticed to be hypertensive in our examinations. Therefore, he is still being followed by pediatric cardiology and nephrology.

Pulse oximetry measurement in the newborn, and routine femoral pulse check and blood pressure measurement in later ages are of critical importance in the diagnosis of aortic coarctation, considering complications and symptoms that can be prevented with early diagnosis and treatment in line with current knowledge. As this case report shows, physical examination should be performed very carefully in newborns and children in order not to overlook the cases. Aortic coarctation is a congenital heart disease that can be treated in any age group. When the diagnosis is achieved, stent implantation should be applied immediately to suitable cases, and balloon angioplasty or surgical treatment should be considered for unsuitable patients.

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