Coarctation of Aorta in the Newborn Period (Report of Three Cases Admitted with Severe Metabolic Acidosis)

YENİDOĞAN DÖNEMİNDE AORT KOARKTASYONU (CİDDİ METABOLİK ASİDOZ İLE BAŞVURAN 3 OLGU SUNUMU)

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Abstract

Detection of the coarctation of aorta (CoA) prior to the onset of symptoms should result in lower morbidity and mortality. However, identification of severe CoA may escape in neonates as it may present with non-recognizable signs and symptoms in this period. In this article we report 3 cases, referred from other hospitals for severe metabolic acidosis and shock with the suspect of a metabolic disease or sepsis, but diagnosed as CoA in our NICU, in order to remind that CoA should always be remembered while searching the etiology of severe metabolic acidosis and shock in newborns.

Key Words: Coarctation of aorta, metabolic acidosis, shock

Özet

Aort koarktasyonu, semptomlar ortaya çıkmadan önce saptandığı taktirde morbidite ve mortalitesi düşük olan bir konjenital kalp hastalığıdır. Ancak yenidoğan döneminde, ciddi aort koarktasyonu olan hastaların her zaman spesifik semptom ve bulgularının olmaması tanıda güçlüklere neden olmaktadır. Bu yazıda, ciddi metabolik asidoz ve şok nedeni araştırılan yenidoğanlarda, ayırıcı tanıda erken dönemde aort koarktasyonu düşünülmesi gerekliliğine dikkat çekmek amacı ile metabolik hastalık veya sepsis düşünülüp kliniğimize sevk edilen, yenidoğan yoğun bakım ünitemizde aort koarktasyonu tanısı konulan 3 olgu sunulmaktadır.

Anahtar Kelimeler: Aort koarktasyonu, metabolik asidoz, şok

Turkiye Klinikleri J Pediatr 2005, 14:204-208

Coarctation of aorta (COA) is an anatomic narrowing of the descending aorta, most commonly at the site of insertion of the ductus arteriosus.[1] CoA, a relatively common defect present in 5-8% of all congenital heart defects and it is the predominant feature in 0.2-0.6/1000 live births.[2-4] However, coarctation can be difficult to diagnose clinically in the newborn as the pulses and upper/lower limb blood pressures may be normal while the ductus is open and if collapse occurs all pulses can be weak if left ventricular dysfunction is marked.[2] When CoA is encountered in a newborn it tends to be more severe than if it occurs later in life. In an infant with severe coarctation, especially associated with a large ventricular septal defect, acute heart failure, shock and severe acidosis often develop suddenly. Multiorgan system failure, necrotising enterocolitis and subsequent death occur rapidly unless definitive medical and surgical intervention are provided immediately. The mortality rate for these infants is 85% without surgery.[5]

We report three cases, referrred from other hospitals for signs of shock, severe metabolic acidosis and renal failure due to the possible diagnosis of a metabolic disease or sepsis but diagnosed as CoA in our Neonatal Intensive Care Unit (NICU).

Case Report

Case 1.

A four day old male infant, born at term, was transferred from another hospital with the diagnosis

Geliş Tarihi/*Received*: 14.04.2004 **Kabul Tarihi/*Accepted*:** 26.09.2004

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of a metabolic disease or sepsis. He had developed hypotonia, poor sucking ability, tachypnea at 48 hours of age and became seriously ill. On admission, he had clinical signs of shock (tachycardia, poor perfusion, cold extremities, lethargy, weak pulses, tachypnea, hypotension, decreased urine output), seizures, hepatomegaly and there was no cardiac murmur. He was immediately entubated and ventilated mechanically. Arterial blood gas levels, obtained from radial vessel, were pH 6.8, PaO_2 52.8 mmHg, $PaCO_2$ 20.6 mmHg, O_2 saturation 62%, BE: -18, HCO_3 5.3 mEq/L and laboratory investigations revealed elevated blood levels of ammonia (486 mcg/dL), lactate (17 mmol/L), blood urea nitrogen (41 mg/dL), creatinine (1.9 mg/dL), phosphate (16.4mg/dL), uric acid (15.5 mg/dL), potassium (7.9 mg/dL) and decreased levels of calcium (7.4 mEq/L). Laboratory tests done for sepsis were negative. Bicarbonate supplementation, inotropic agents (dopamine infusion of 15 μg/kg/minute IV), empiric antibiotic therapy, phosphate binders and volume expanders were initiated. Blood and urine samples were sent for the analysis of amino and organic acids. After correcting his acid-base imbalance, electrolyte abnormalities and dehydratation, a significant difference was detected between the measurement of systolic/diastolic blood pressures of the upper and lower extremities (75/32 mmHg and 44/28 mmHg respectively) and his femoral pulses were still weak in contrast to bounding radial pulses. There was congestion and cardiomegaly on his telecardiography. Echocardiography was performed and severe juxtaductal aort coarctation with atrial septal defect and patent ductus arteriosus were detected. Diagnosis was confirmed by angiography and the systolic pressure gradient between the ascending and the descending aorta beyond the stenosis was found as 30 mmHg. After the initial stabilization he was transferred to another hospital for surgery. After 48 hours of his transfer, amino and organic acid analysis were reported as normal.

Case 2

A 20 days old female infant was referred to our hospital with the diagnosis of sepsis and congestive heart failure because of respiratory distress, cyanosis and poor sucking ability. On admission, the baby had clinical signs of shock and congestive heart failure and a second degree systolic murmur on the left sternal border. She was immediately entubated and put on mechanical ventilator. Arterial blood gas levels, obtained from radial vessel, were pH 7.0, PaO_2 50.6 mmHg, PaCO2 30.6 mmHg, O_2 saturation 54%, BE= -16, HCO_3 4.2 mEq/L and laboratory investigations showed elevated blood ammonia (526 mcg/dL), lactate levels (14 mmol/L) and increased anion gap (22 mEq/L). Her clinical and laboratory findings were compatible with cardiac failure, respiratory insufficiency and prerenal failure. Fluid management, empirical antibiotic therapy (although sepsis work-up was negative), inotropic agents (dopamine infusion of 10 μg/kg/minute IV), and bicarbonate supplementation were started immediately. A significant difference was detected between the systolic/diastolic pressures of her upper and lower extremities after the initial treatment, and her femoral pulses were weak in contrast to the bounding radial pulses. Pulmonary edema and cardiac enlargement were detected on her chest radiography. Echocardiography was performed. Preductal coarctation of the aorta with constricted ductus arteriosus and associated lesions including secondum type atrial septal defect and ventricular septal defect were detected. Angiography revealed a preductal-type of CoA and constricted ductus could not be visualized (Figure 1). The systolic pressure gradient between the ascending aorta and the descending aorta beyond the stenosis was 40 mmHg. Transfer for operation was planned as soon as possible but her clinical status deteoriated and she died. Analysis of blood and urine aminoacids and urine organic acids were reported as normal after her death.

Case 3

A male baby was referred on his 7^{th} day of life because of oliguria and metabolic acidosis. He had been diagnosed as a metabolic disease. On physical examination he had clinical signs of shock and a second degree continuous murmur on the left sternal border with no radiation. Arterial blood gas levels, obtained from radial vessel, were pH 7.1,

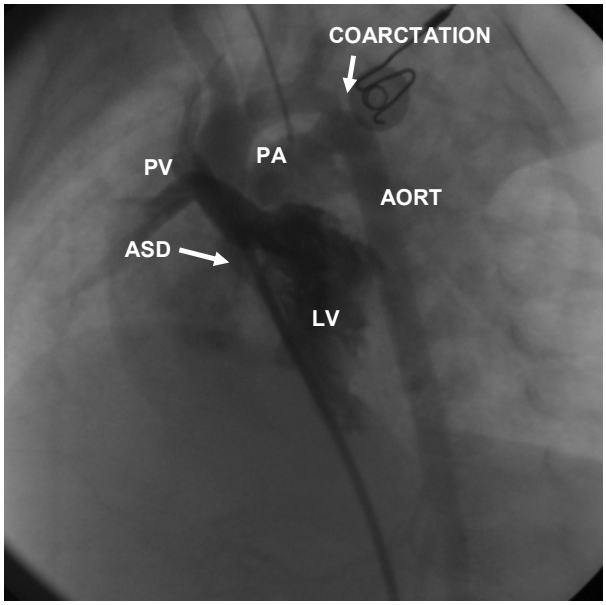

Figure 1. Upper right pulmonary vein injection of contrast material showed the secundum atrial septal defect and preductal coarctation of aorta (Case 2).

PaO_2 68.6 mmHg, $PaCO_2$ 38.4 mmHg, O_2 saturation 78%, BE: -15, HCO_3 6.2 mEq/L and laboratory investigations showed elevated levels of blood ammonia (298 mcg/dL), lactate (11 mmol/L), blood urea nitrogen (35mg/dl), creatinine (2.6 mg/dl). Renal failure indices supported intrinsic renal failure. Laboratory work-up for sepsis was normal and there was not any congenital renal malformation on renal ultrasonography. Volume expander and inotropic agent (dopamin infusion of 10 μg/kg/minute IV) were administered to correct hypotension and to improve renal perfusion. Metabolic acidosis persisted after the supplementation of bicarbonate. His urine output didn't increase neither with fluid challenge of normal saline nor with furosemide and dopamine administration so, he showed physical findings of volume overload. Cardiomegaly and pulmonary congestion findings were detected on his chest radiography. A significant difference was detected between the systolic/diastolic pressures of his upper and lower extremities after the initial treatment, and his femoral pulses were weak in contrast to the bounding radial pulses. So, echocardiography was performed. Juxtaductal CoA, and associated anomalies of secundum atrial septal defect, ventricular septal defect, patent ductus arteriosus were detected. Angiography revealed aort coarctation (the systolic pressure gradient between the ascending aorta and the descending aorta beyond the stenosis was 36 mmHg) (Figure 2). Volume overload could not be controlled with conservative management. So, dialysis and balon angioplasty were performed consequently before the operation. After his clinical stabilization he was referred for operation to repair his cardiac defects.

Discussion

Coarctation, congenital narrowing of upper descending thoracic aorta, is the 4th most common defect that occurs in 5-8% of patients with congenital heart disease.[1-3] Detection of CoA prior to the onset of symptoms should result in earlier referral, lower perioperative morbidity and mortality and better long term outcome. Therefore, early identification and repair of this lesion is highly desirable.[6,7]

Clinical features of the patients vary according to their age, severity of the coarctation and pres-

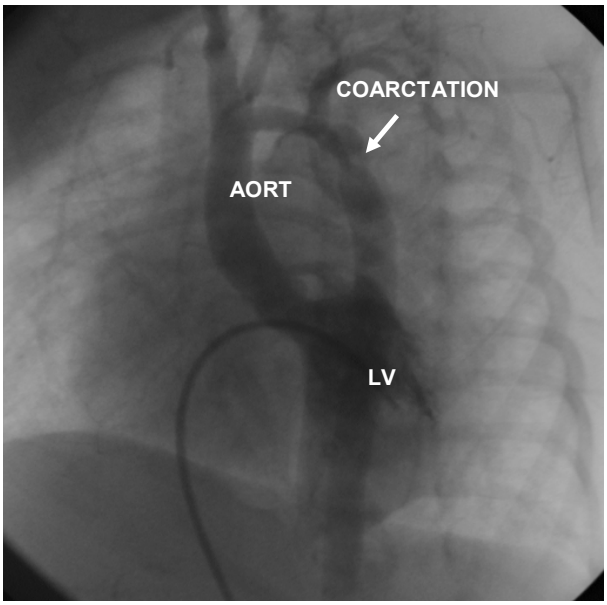

Figure 2. Left ventriculography of the patient showed the juxtaductal coarctation of aorta (Case 3).

ence or absence of associated major cardiovascular anomalies.[1,2,8] The classic sign of CoA is a disparity in pulsations and blood pressures of the arms and legs. The femoral pulses are weak in contrast to the bounding pulses of arms, so femoral pulses should always be palpated simultaneously with radial pulses. 82% of patients with CoA have isolated lesions and most of them are mild. These patients are usually asymptomatic. In contrast, patients with isolated but severe CoA or with associated cardiac defects as ventricular septal defect, atrial septal defect or aortic stenosis as well as CoA, may present with poor feeding, tachypnea, lethargy early in life and rapidly progress to overt congestive heart failure, metabolic acidosis and shock as the ductus arteriosus closes. Presentation of clinical findings are usually abrupt in these newborns. However developmental time of these symptoms may vary between 48 hours of age and early infancy period.[9] The diagnosis of CoA may be missed or delayed unless an index of suspicion is maintained as the babies with severe CoA usually have clinical signs of shock and metabolic acidosis on admission that may be misdiagnosed as inborn errors of metabolism, renal failure or sepsis.

We presented 3 infants who have been diagnosed as a metabolic disease or infection in other hospitals but later diagnosed as CoA in our NICU. Clinical and laboratory findings of these patients were similar but, the postnatal ages of presenting the signs (ranging between 48 hours and 20 days) were different. This difference might be related with the severity of obstruction, the degree of constriction in ductus arteriosus and with associated cardiac lesions as VSD and/or ASD in these babies.

Two-dimensional and Doppler echocardiography is generally accepted as a useful technic in assessment of CoA and intracardiac lesions associated with coarctation. High-quality echocardiographic studies may yield sufficient anatomic and physiologic data to enable the clinician to avoid diagnostic cardiac catheterization and angiography in many patients. We also obtained detailed information about the anatomical cardiac pathology with echocardiography. However, angiography was performed in our patients to confirm the diagnosis and to treat with Balloon angioplasty at the same time (for the 3rd patient) before transferring them to another hospital for surgical repair.

Coarctation, presenting with severe clinical findings demands immediate and aggressive treatment. Metabolic disturbances must be recognized and treated promptly.[10] After the initial stabilization, surgical intervention is recommended as early as possible. Our patients were in the clinical presentation of shock on admission to NICU. Occurence of shock can be expected in patients as the ductus becomes constrict. However, constriction could be detected in only one of our patients whose clinical presentation was also complicated with prerenal failure and she died after a few days. It is difficult to explain the clinical presentation of shock in others, especially in the first patient, as the systolic pressure gradient was not enough to be responsible from shock. Neither any pathologic laboratory finding to diagnose sepsis nor any pathologic blood or urine analysis to diagnose metabolic disease could be detected in these patients so, CoA seems to be the only reason for shock. However sepsis or infection generally presents with nonspesific signs and laboratory findings may not support the diagnosis of infection in the newborn period. Both of our patients have been referred from other hospitals after the initial treatment with antibiotics so these patients' clinical deteorization might stil be related with infection that could not have been detected with laboratory findings. Whatever the reason is, all of them were in the clinical presentation of shock so we could not detect the discrepancy in arteral pulses and systolic blood pressures between the upper and lower extremities on the initial physical examination. Second case was the worst and the oldest one. He had severe preductal type of CoA and we were late for his transfer as we could not be successful in correcting his metabolic problems rapidly. We planned to administer PGE_2 to him although the ductus was almost closed but, unfortunately we could not obtain it. Unfortunately it could not be possible to perfuse the lower parts of the body as the ductus was nearly closed and to overcome the

pulmonary congestion, so he died before performing an operation.

Balloon dilatation is recommended as palliation in patients with severe left ventricular dysfunction or in case of surgery is prohibitive for other reasons.[11] Balloon angioplasty was performed as a life saving procedure in our third patient because, volume overload could not be controlled despite conservative management and dialysis. He was transferred after the initial stabilization, for surgical repair.

In conclusion, CoA should be in the differential diagnosis of patients with suddenly occuring clinical signs of shock and metabolic acidosis due to unknown etiology and echocardiography should be performed as soon as possible in these patients. After the initial treatment of shock, extremity pulses should be palpated and blood pressure of four extremities should be measured in these patients again, in order to detect the discrepancy in arterial pulses and blood pressures between the upper and lower extremities.

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