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OLGU SUNUMU / CASE REPORT

An infant with congenital midaortic syndrome

Konjenital midaortik sendromlu bir bebek

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Abstract
Midaortic syndrome is an uncommon disease characterized by segmental or diffuse narrowing of abdominal or distal descending thorasic aorta. Majority of the cases are idiopathic but some are secondary to fibromuscular dysplasia, giant cell arteritis, Moyamoya disease or genetic diseases such as neurofibromatosis, Williams' syndrome and Alagille syndrome. It is very rare in newborn period. Here we present a 40 days old infant who admitted to hospital for respiratory distress, hypertension, heart failure and was diagnosed as midaortic syndrome. She had medical treatment but unfortunately died due to heart failure when she was two months old age.
Key words: Midaortic syndrome, aort coarctation, infant

Öz
Midaortik sendrom nadir bir hastalık olup abdominal veya distal inen aortanın segmental veya difüz darlığı ile karakterizedir. Olguların çoğu idiyopatik olmakla birlikte bir kısmı da fibromüsküler displazi, dev hücreli aterit, Moyamoya hastalığı veya nörofibramatozis, Williams' sendromu ve Alagille sendromu gibi genetik hastalıklara ikincildir. Burada 40 günlük iken hastaneye solunum sıkıntısı, kalp yetmezliği tablosunda başvuran ve midaortik sendrom tanısı alan bir olgu sunulmuştur. Olguda medikal tedavi tercih edilmiş ancak bebek iki aylık iken eksitus olmuştur.

Anahtar kelimeler: Midaortik sendrom, aort koartasyonu, bebek

INTRODUCTION

Midaortic syndrome (MAS), also called as midaortic dysplastic syndrome is characterized by segmental or diffuse narrowing of the distal descending or abdominal aorta and is a very rare syndrome in childhood. 0.5-2% of aortic stenosis are due to MAS.[1,2] There is segmental or diffuse narrowing in the abdominal aorta; and renal, superior mesenteric arteries may be involved. MAS may be congenital or acquired.[3]

Most of the patients are admitted with severe hypertension. Current management aims to control hypertension with medical treatment, however oral antihypertensive drugs may not be effective in most of the cases and balloon dilatation or transcatheter placed stents can be used to dilate the coarctation; or surgical procedures such as aortaaortic bypass, graft vascular replacement and patch angioplasty are needed[3]. Here we report an infant with MAS who admitted to hospital for respiratory distress, hypertension and heart failure.

CASE

Fourty days old girl was admitted to hospital for respiratory failure and poor feeding. She was the second child of nonconsanguinous parents and was born at 39 gestational weeks after an uneventful pregnancy. She had cutis marmoratus, respiratory distress, tachycardia (187/ min), hypertension and metabolic acidosis (capillary pH: 7.1, pCO2: 25 mm Hg, BE:-14, HCO3: 12 mmol/L) on admission. Capillary refill time was 4 seconds. Femoral pulses were weak. Arterial tension was 120/90 mm Hg. Oxygen saturation was 80% without oxygen. Nasal synchronised intermittent mandatory ventilation was

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applied, 10 ml/kg saline was infused in ten minutes and 120 ml/kg %0.34 NaCl was started. She had cardiomegaly on her chest X-ray. Her birth weight was 2500 g (<3p), and she was 2600 g (<3p) on admission. Her laboratory values were normal except for BUN: 28 mg/dl and creatinine: 0.98 mg/dl. Metabolic acidosis improved after hydration.

Echocardiography showed left ventricular hypertrophy. Although her clinic findings and left ventricular hypertrophy caused clinicians to suspect aort coarctation, no coarctation was seen in the classical location. Abdominal Doppler ultrasonography revealed high speed monophasic flow in abdominal aorta and external iliac artery. Angiography was performed in the 49th day of life. Femoral arterial catheterization could not be performed.

Percutaneous femoral venous catheterization was done, and via vena cava inferior, right atrium and foramen ovale radio opac material was injected to left atrium. Angiography showed no filling of the aorta after suprarenal level consistent with midaortic syndrome (Figure 1). As the patient had a stenosis in a long segment, stent placement or balloon dilatation could not be performed, also surgery had high-risk and she was managed with antihypertensive agents, propranol, amlodipine, methyl dopa and with a diuretic, furosemid. Unfortunately she died due to heart failure in 59th day of life. Informed consent was gained from the parents.



Figure. 1. Angiography shows narrowing of the abdominal aorta and no filling with radio-opac material beyond suprarenal level.

DISCUSSION

Midaortic syndrome is an uncommon disease characterized by segmental or diffuse narrowing of abdominal or distal descending thorasic aorta. Majority of the cases are idiopathic but some are secondary to fibromuscular dysplasia, giant cell arteritis, neurofibromatosis, Moyamoya disease, Williams' syndrome and Alagille syndrome[1-3]. The present case seems to be idiopathic.

Although the first patient was described about 6 decades ago[4], pathogenesis is not exactly known. Congenital coarctation has been thought to be due to incomplete fusion or overfusion of embryonic dorsal aortas during 4th week of gestation[5], rubella infection and abnormal migration of the kidneys[7], however they have not been proved. The most common histological finding in the abdominal aorta and visseral vessels in idiopathic MAS is fibroplasia of the intima and variable distortions of the internal elastic lamina[8,9]. Autopsy showed fibrointimal plaque in the preterm infant reported by Harmon et al[10].

Recently Rumman et al[3] reviewed childhood MAS in English literature and found 630 reported cases of children. 64% of the cases were idiopathic, 15% were due to genetic diseases and 17% were related to inflammatory diseases. The mean age of the patients were 9.1 years and 55% were girls. In 97% of the children, narrowing was present in abdominal aorta, while distal aorta was involved in 3% of the cases. Involvement of the anatomic site within abdominal aorta was not reported in 43% of the cases. In the remaining patients, 51% coarctation were in suprarenal, 20% were in supra to infrarenal, 15% were in interrenal and 14% were in infrarenal region. In our patient coarctation was in suprarenal region. In idiopathic cases high incidence of renal involvement were reported (91%, of which 61% were bilaterally affected)[11].

Angiography is the gold standart for diagnosis, however MR angiography, CT angiography, abdominal ultrasonography, echocardiography and 3-D CT may also be useful. Midaortic syndrome can present with hypertension, headache, leg claudication, fatigue or abdominal angina and hydrops fetalis. [3, 12] In Rumman et al's study [3], the most common finding at presentation was hypertension (87%), followed by headache (13.2%), claudication (10.3%), dyspnea (7.5%) and failure to thrive (3.8%). In our patient, dyspnea and heart failure were the leading findings. She also had failure to thrive and hypertension.

If not treated, most of the patients with MAS die

due to severe hypertension before 35-40 years old. Treatment can be pharmacological, endovascular (balloon expandable stents, stent grafts) or complex endovascular/ surgical. Each child needs individualized theraphy. Although antihypertensive agents may be useful in mild and moderate cases, endovascular and surgical interventions are required in severe cases. The present patient was treated with furosemid, propranol, amlodipine and methyl dopa. She had an improvement in blood pressure, however hypertension persisted. In uncontrolled hypertension, endovascular percutaneous transluminal angioplasty with or without stents or balloon dilation have been performed in children and adolescents, however symptomatic MAS in newborn period is almost fatal[3,11-14]. Balloon dilation and stenting are effective for short-segment lesions but do not yield favorable results in patients with long-segment stenosis[15]. As the present patient was only 2600 g, we wanted to do medical treatment allowing her to grow until surgery or stent placement. Unfortunately she died due to heart failure. To our knowledge there are only 9 newborn cases in the literature[10-14,16,17]. Five of 9 babies died secondary to intraventricular hemorrhage or cardiac failure [10,12-14]. Our patient was 40 days on admission.

In conclusion midaortic syndrome should be in mind in patients with clinic findings of aort coarctation even coarctation can not be detected in the classical location in echocardiography. Although MAS is rare in childhood it must be considered in unexplained cases of hypertension.

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