CASE REPORT

Case report of a rarely seen long-segment middle aortic syndrome

Nadir görülen uzun segment middle aortik sendrom: Olgu sunumu

Kahraman Yakut, M.D., İlkay Erdoğan, M.D.

Department of Pediatric Cardiology, Başkent University Faculty of Medicine, Ankara, Turkey

Summary– Middle aortic syndrome (MAS) follows a course with distal thoracic and abdominal aorta stenosis. It is a rare disease that is usually diagnosed after the first decade of life. Clinical reflection of MAS is often in the form of hypertension and claudication in the lower extremities. Its etiology is unclear, but is known to be associated with congenital or acquired diseases. This pathology, which is accompanied by malignant hypertension, often does not respond to medical treatment. In patients with MAS, surgical treatment is first line recommendation to prevent complications such as hypertension, heart failure, intracranial bleeding, or aortic rupture. In order to draw attention to this disease, presently described is case of high blood pressure detected during routine examination of a child who had no complaint, and discovery of long-segment stenosis in the abdominal aorta identified with echocardiography and conventional angiography.

Özet– Middle aortik sendrom (MAS) distal torasik ve abdominal aortanın darlığı ile seyreder. Genellikle ilk dekattan sonra tanı konan nadir bir hastalıktır. Middle aortik sendromun klinik yansıması sıklıkla hipertansiyon ve alt ekstremitelerde klaudikasyon şeklindedir. Etiyolojisi tam bilinmemekle birlikte doğumsal veya akkiz hastalıklara eşlik ettiği bilinmektedir. Malign hipertansiyonla seyreden bu patoloji, çoğunlukla tıbbi tedaviye yanıt vermemektedir. Middle aortik sendromlu hastalarda hipertansiyon, kalp yetersizliği, intrakraniyal kanama, aort rüptürü gibi komplikasyonları önlemek için cerrahi tedavi birinci sırada önerilmektedir. Bu yazıda, herhangi bir şikayeti olmaksızın rutin muayene sırasında tansiyon yüksekliği saptanan, ekokardiyografi ve geleneksel anjiyografi ile abdominal aortada uzun segment darlığı belirlenen çocuk olgu, bu hastalığa dikkat edilmesi için sunuldu.

Middle aortic syndrome (MAS) is a rare pathology characterized by varying degrees of stenosis of the distal thoracic aorta, abdominal aorta, and its branches. The disease commonly involves the renal (91–80%) and splanchnic (50–70%) branches of the aorta, and is the most common clinical syndrome associated with stenotic aorto-arteriopathy in children.[1–5] This lesion, known as abdominal aortic coarctation, constitutes 0.5% to 2.0% of all coarctations.[6] MAS may be associated with congenital abnormalities, as well as Takayasu syndrome, Williams syndrome, neurofibromatosis, fibromuscular dysplasia, retroperitoneal fibrosis, and mucopolysaccharidosis.[3,7,8] Presently described is case with hypertension identified during routine examination and diagnosis of MAS confirmed with echocardiography and catheter angiography in the context of the current literature.

Abbreviation:

MAS Middle aortic syndrome

CASE REPORT

Hypertension was identified in a 12-year-old male patient who had no complaints during routine examination at another medical facility. Aortic coarctation was detected in echocardiographic examination, and the patient was referred to our clinic. On physical examination, no femoral pulses could be palpated on either side. Following 15 minutes of resting in a quiet environment, systolic and diastolic blood pressures were measured 3 times using mercury sphygmomanometer with appropriate cuff size, and average value was recorded. Both upper extremity blood pressures were measured as 200/100 mmHg. Kidney function tests were normal. On electrocardiographic examination, there were findings of left ventricular hypertrophy. Classic coarctation area was patent with no significant gradient could be gained. There were hypertro-

Received: July 02, 2015 *Accepted:* August 24, 2016

Correspondence: Dr. Kahraman Yakut. Başkent Üniversitesi Tıp Fakültesi, Çocuk Kardiyoloji Bilim Dalı, Ankara, Turkey.
Tel: +90 312 - 203 68 68 / 1382 e-mail: kahramanyakut@gmail.com

phic changes in the left ventricle. Subcostal study revealed stenosis in long axis image of the abdominal aorta. On catheter angiographic examination, diffuse hypoplasia was detected in the abdominal aorta, beginning at level of the diaphragm and extending to 2 cm above the iliac artery bifurcation. There was no observation of the right-left renal arteries, the superior mesenteric artery, or the celiac artery, and the kidney and the visceral organs were observed to be supplied through collateral vessels (Figure 1, Video 1*). There were no signs of Takayasu syndrome, neurofibromatosis, or retroperitoneal fibrosis. Due to lack of dysmorphic features, Williams syndrome was excluded. There was no history of catheter placement in the umbilical artery in neonatal period. The patient was therefore accepted as congenital MAS. Due to long-segment diffuse hypoplasia in the abdominal aorta and visceral arteries, surgical treatment was planned, since the patient was not suited to balloon angioplasty and/or stent application.

DISCUSSION

Congenital MAS is caused by developmental disorder of embryonic dorsal aorta fusion and maturation.[9] Stenosis in MAS may occur at various levels, such as the thoracic aorta, abdominal aorta, or visceral and renal branches below the diaphragm.[1,4,5,10] Clinical presentation has included hypertension, claudication, renal failure, headache, and intestinal ischemia.[2,4,5,7] Additional symptoms include fatigue upon effort, ischemic pain in the lower extremities, and intestinal angina.[1,2,6] Hypertension is frequently diagnosed as a coincidence during the investigation for disease.

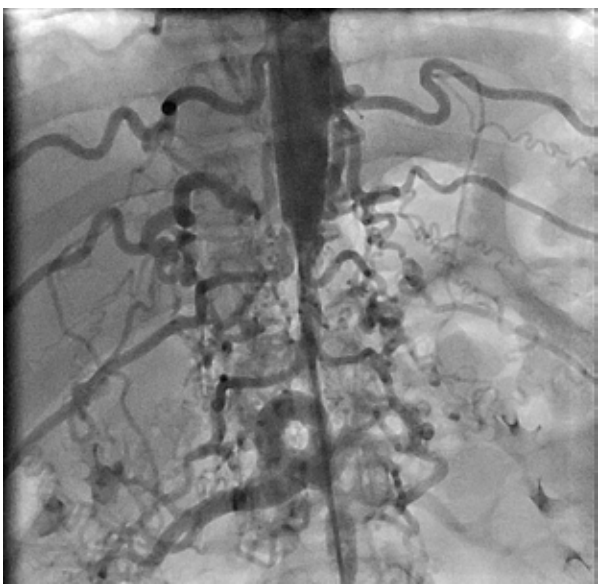

Figure 1. Catheter angiographic examination revealed diffuse hypoplasia in the abdominal aorta, starting from level of the diaphragm and extending to 2 cm above the iliac artery bifurcation. There was no observation of the right-left renal arteries, the superior mesenteric artery, or the celiac artery, and the kidney and the visceral organs were observed to be supplied through collateral vessels.

Severe renovascular hypertension may accompany absence or weakness of femoral pulses in patients. MAS may be diagnosed with echocardiography, aortography, magnetic resonance angiography, and Doppler ultrasound.[1,5,7] In cases of MAS, left ventricular hypertrophy with diastolic dysfunction is observed on echocardiography. In the present case, echocardiography indicated left ventricular hypertrophy with stenosis in the abdominal aorta. Diagnosis was confirmed with catheter angiography.

In cases of MAS, medical treatments, interventional procedures, and surgery are recommended, depending on nature of lesion and symptoms.[6,11] In the majority of patients, surgical intervention is required in addition to medical treatment to control hypertension.[1,12–14] Response to medical therapy alone has failed over long period, and balloon angioplasty treatment has shown only partial success. On the other hand, despite need for surgery and complex anatomy, long-term curative treatment has been provided for most patients.[6,12–14] In our patient, due to long-segment coarctation, balloon angioplasty was not considered beneficial; therefore, surgical treatment was planned. Most patients who were not treated have been reported to have died at around the age 40 with complications such as myocardial infarction, heart failure, intracranial hemorrhage, aortic rupture, or renal failure.[1,6]

Conclusion

As a result, measurement of blood pressure should be part of routine examination in all children and high values must be investigated further. In addition, during physical examination, all pulses should be checked and compared. Hypertension detected during routine examination led us to diagnosis of MAS. With no consensus on treatment of MAS, it should be known that surgical intervention has high rate of providing complete success.

Conflict-of-interest issues regarding the authorship or article: None declared.

****Supplementary video file associated with this article can be found in the online version of the journal.***

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Keywords: Coarctation; congenital heart disease; hypertension; middle aortic syndrome.

Anahtar sözcükler: Koarktasyon; konjenital kalp hastalığı; hipertansiyon; middle aortik sendrom.