Middle Aortic Syndrome As A Cause of Dilated Cardiomyopathy

Dursun Alehan, MD, Gülden Kafalı, MD, Metin Demircin, MD*

From the Department of Pediatric Cardiology and Department of Thorax and Cardiovascular Surgery*, Hacettepe University, Faculty of Medicine, *Ankara, Turkey*

Introduction

Coarctation of the aorta (CoA) is one of the most common causes of dilated cardiomyopathy secondary to congenital cardiac defects. When CoA is in the distal region of left subclavian artery or juxtaductal region, it can be diagnosed easily by the physical examination and echocardiographic study performed in the suprasternal notch. Early diagnosis and therapy of CoA is important for preventing the development of secondary dilated cardiomyopathy. When the diagnosis is doubtful according to these diagnostic methods, subcostal Doppler echocardiographic study and catheter angiography may be required to rule out abdominal CoA (1-4). We present a three-year-old boy who has been followed-up with the diagnosis of idiopathic dilated cardiomyopathy for 2,5 years and in whom long-segment coarctation of the abdominal aorta and middle aortic syndrome were diagnosed during follow-up.

Case Report

At the age of 8 months, the patient had been admitted to hospital with the diagnosis of respiratory system infection. On his physical examination, cardiac murmur had been determined and he was referred to our hospital' s pediatric cardiology unit.

On admission to the hospital, his weight was 8 kg, pulse was 120 beats/min, respiratory rate was 22/min, femoral pulses in the lower extremities were weakly palpable and his blood pressure was measured as 110 / 70 mmHg. Respiratory and gastrointestinal systems were found to be normal. On his cardiac system examination, a grade II/VI holosystolic murmur was heard best at cardiac apex. Initial laboratory investigation revealed the following: hemoglobin-11.5 g/dl; hematocrit-34.3 %; white blood cell count-11.000/mm^3; platelet count-259.000/mm^3. The chest X-ray showed cardiomegaly, prominent main pulmonary artery segment and pulmonary venous congestion. Electrocardiography showed normal QRS axis, normal sinus rhythm and left ventricular hypertrophy. Echocardiographic assesment of left ventricular dimensions and function were performed: the left ventricular end-diastolic dimension (LVEDD) was 40 mm (normal value according to the body weight of the case: 24-31 mm), the left ventricular end-systolic dimension (LVESD) was 31 mm (normal value: 14-21 mm). Left ventricular ejection fraction (EF) and fractional shortening (FS) were measured as 53% and 23 %, respectively. Additionally mitral regurgitation (regurgitant jet velocity 4.7 m/sec) was detected. Because of decreased femoral pulses, coarctation of the aorta was searched carefully by echocardiography from suprasternal views, but no abnormalities were detected in the aortic arch. Decreased femoral pulse amplitude was attributed to his cardiomyopathy and low cardiac output. The patient has been followed with the diagnosis of dilated cardiomyopathy and mitral regurgitation and, has been treated by anticongestive medications. During follow-up, his left ventricle dimensions increased and functions decreased gradually.

During his last visit, when he was 3 years old, weak femoral pulses but strong radial pulses were noted, and his blood pressure in the both arms were measured as 170 / 110 mmHg. His weight was 14 kg (25-50 percentage). On cardiac examination; apical grade II to III / VI holosystolic murmur, and at the upper left sternal border, grade II to III / VI systolic ejection murmur were heard. Hepatomegaly was determined. Echocardiography revealed larger left ventricular dimensions and lower left venricular function (LVEDD: 49 mm, LVESD: 39 mm, EF: 41 %, FS: 20 %). In addition, mitral-E septum distance was measured as 21 mm. Echocardiography showed grade 2 to 3 mitral regurgitation (regurgitant jet velocity 4 m/sec), grade 1 aortic regurgitation (regurgitant jet velocity 2.5 m/sec), mild pulmonary regur-

Address for correspondence: Dr. Dursun Alehan, Hacettepe Üniversitesi Tıp Fakültesi, Çocuk Sağlığı ve Hastalıkları Anabilim Dalı, Kardiyoloji Ünitesi, Ankara / Türkiye
Tel: 90 - 312 - 3051157 (Work), 0-532-5503041 (Private), Fax: 90 - 312 – 3090220, E-Mail: dalehan@hacettepe.edu.tr

gitation (regurgitant jet velocity: 2.5 m/sec) and mild pulmonary stenosis. No coarctation was detected at its classical localization. Because of the stronger radial and weak femoral pulses, subcostal echocardiography was repeated in an attempt to see thoraco-abdominal aorta for possible coarctation. Detailed echocardiographic study revealed long-segment thoraco-abdominal coarctation of the aorta with a systolic pressure gradient of 90 mmHg extending into diastole. Thoracic and abdominal aortography confirmed the diagnosis and showed 10 cm long severe segmental narrowing of the thoracoabdominal aorta (Fig. 1). Furthermore, all other arteries originating from abdominal aorta (renal, visceral and iliac arteries) had a hypoplastic appearance. Systolic pressure gradient between the proximal and distal parts of the coarctation was measured as 103 mmHg during catheterization (200 mmHg vs. 97 mmHg).

During operation via a thoracoabdominal approach, CoA was found exactly in that segment described by aortography. Revascularisation was achieved by placing on aorto-aortic bypass from the proximal to the distal end of the coarcted segment using a Cardial collagen coated Dacron graft (no: 10). Unfortunately, histologic examination of coarcted segment was not performed. Preoperatively, the difference of blood pressure between the upper and lower extremities was 85 mmHg (120 mmHg, 35 mmHg, respectively), whereas postoperatively the difference of blood pressure was 55 mmHg (135 mmHg, 80 mmHg, respectively). Thirty hours postoperatively, suddenly hypotension and then cardiopulmonary arrest developed and the patient died unexpectedly.

Discussion

Coarctation of the aorta is one of the most frequently observed causes of secondary dilated cardiomyopathies originating from congenital heart diseases. When early diagnosis is made, progress to cardiomyopathy may be prevented. During physical examination simultaneous palpation of upper and lower extremity pulses is very helpful for the diagnosis. The combination of strong brachial and only weakly palpable femoral pulses should lead to the suspicion of an aortic coarctation in any case. Therefore, routine examination of blood pressures and pulses in both lower and upper extremities must certainly be made in all patients (5, 6).

Abdominal coarctation is an unusual form of coarctation characterized by segmental stenosis of the aorta and its branches. Due to the disagreement regarding the etiology of abdominal coarctation and the fact that the name ''coarctation'' implies a congenital origin, some authors prefer a more neutral name, such as ''middle aortic syndrome'' (1-4). The spectrum of middle aortic syndrome encompasses narrowing of the abdominal aorta and progressive involvement of the renal and visceral branches. It is an uncommon lesion, accounting for only 2 % of aortic hypoplastic lesions, but is increasingly recognized as a cause of hypertension in children and young adults. Coarcted segments are frequently longer and renal artery stenosis is found in 80 % of patients, and 25% have involvement of the superior mesenteric artery, inferior mesenteric artery or celiac axis. Middle aortic syndrome has been described in Takayasu arteritis, fibromuscular dysplasia, a number of genetic diseases, such as neurofibromatosis, muco-

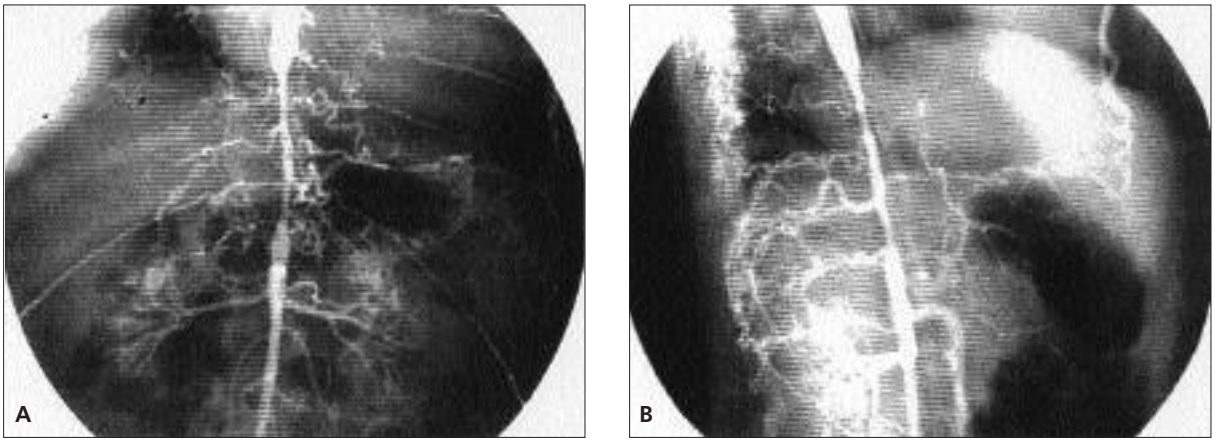

Figure 1. Aortography of the patient shows a severe long-segmental narrowing of thoraco-abdominal aorta in antero-posterior (A) and lateral (B) projections.

polysaccharidosis, Williams syndrome, Turner syndrome, Alagille syndrome, and congenital rubella syndrome (2, 7, 8). In addition, irradiation of abdominal or retroperitoneal neoplasms early in life, particularly in case of Wilms tumour, have resulted in a similar angiographic appearance (7).

Takayasu arteritis has the progressive nature and it is characterized histologically by aortic segments that are narrowed by marked intimal hyperplasia and that alternate with areas of poststenotic aneurysm. But, the diffuse nature of the aortic involvement and the frequent association with a systemic illness similar to collagen vascular disease further typify this disease entity. Whether isolated abdominal coarctation is ever a congenital anomaly or only exists as a postinflammatory narrowing of the aortic lumen is not clear (2, 4).

The above discussed syndromes and genetic diseases have the other specific symptoms and clinical findings of the disorder to make differential dignosis. Because of the absence of the stigmata relating to these disorders in our patient, the differential considerations are limited to Takayasu arteritis and fibromuscular dysplasia. Fibromuscular dysplasia is a congenital disorder of the connective tissue in the blood vessels and is a pathological diagnosis. Fibromuscular dysplasia and Takayasu arteritis may be distinguished only by histological examination. Histological findings of Takayasu arteritis are severely narrowed aorta and renal arteries by marked intimal fibrosis and adventitial inflammation and scarring. Whereas, histopathological appearance of fibromuscular dysplasia is lack of inflammation in all layers of vessel walls and severely narrowing of aorta and renal arteries by intimal and medial fibroplasia, medial hyperplasia and perimedial dysplasia (2, 9, 10). We could not obtain histological examination of our case. However, because of lack of the other stigmata observed in Takayasu arteritis, we think that abdominal coarctation of our case may result from fibromuscular dysplasia.

Balloon angioplasty or autotransplantation for renal artery stenosis and stent implantation or bypass graft for stenosed aortic segments are the suggested therapeutic options in the treatment of middle aortic syndrome. Also, Lillehei et al. (11) have suggested staged vascular repair to minimize renal ischemia. In our patient revascularization has been obtained in only thoracoabdominal coarcted segment via bypass graft. The patient died after thirty hours postoperatively despite successfull surgical procedure. Unfortunately, it is difficult to know the exact cause of death in our patient since we could not get permission for autopsy. However, we think that death may be secondary to dilated cardiomyopathy or complications due to renal or visceral ischemia although there was no measurable rise of blood urea nitrogen or serum creatinine levels postoperatively.

Consequently, all patients presenting with clinical findings of both dilated cardiomyopathy and coarctation (weak femoral pulses, hypertension) must certainly be evaluated for coarctation and thoracoabdominal coarctation should be excluded by detailed echocardiographic and/or angiographic study.

References

1. Wozniak G, Bauer J, Bohle RM, Dapper F. Coarctation of the thoraco-abdominal aorta: operative treatment with a cryopreserved arterial homograft in a seven-year-old boy. J Cardiovasc Sug 1998; 39: 483-8.
2. D' Sauza SJA, Tsai WS, Silver MM, et al. Diagnosis and management of stenotic aorto-arteriopathy in childhood. J Pediatr 1998; 132: 1016-22.
3. Shefler AG, Ostman-Smith CI. Middle aortic syndrome in a boy with arteriohepatic dysplasia. Pediatr Cardiol 1997; 18: 232-4.
4. Morris MJH, McNamara DG. Coarctation of the aorta and interrupted aortic arch. In: Garson A, Bricker JT, Fisher DJ, Neish SR, editors. The Science and Practice of Pediatric Cardiology (2nd ed). Baltimore, Philadelphia: Williams & Wilkins; 1997:p.1317-46.
5. Venugopalan P, Agarwal AK, Worthing EA. Chronic cardiac failure in children due to dilated cardiomyopathy: diagnostic approach, pathophysiology and management. Eur J Pediatr 2000; 159: 803-10.
6. Prabhu SS, Dalvi BV. Treatable cardiomyopathies. Indian J Pediatr 2000; 67: 279-82.
7. Roun ME, Skolkin MD. Peripheral vascular angiography. In: Garson A, Bricker JT, Fisher DJ, Neish SR, editors. The Science and Practice of Pediatric Cardiyology (2nd ed). Baltimore, Philadelphia: Williams & Wilkins; 1997:p.1041-63.
8. Sumboonnanonda A, Robinson BL, Gedroyc WM, Saxton HM, Reidy JF, Haycock GB. Middle aortic syndrome: clinical and radiological findings. Arch Dis Child 1992; 67: 501-5.
9. Jansen J, Vuong PN, Rothenberger-Janzen K. Takayasu' s arteritis and fibromuscular dysplasia as causes of acquired atypical coarctation of the aorta: retrospective analysis of seven cases. Heart Vessels 1999; 14: 277-82.
10. Matsushita M, Yano T, Ikezawa T, et al. Fibromuscular dysplasia as a cause of abdominal aortic aneurysm. Cardiovascular Surg 1994; 2: 615-8.
11. Lillehei CW, Shamberger RC. Staged reconstruction for middle aortic syndrome. J Pediatr Surg 2001; 36: 1252-4.