Off-pump coronary bypass in a child with familial hypercholesterolemia: premature atherosclerosis of the ascending aorta

Familyal hiperkolesterolemili çocukta çıkan aortanın erken aterosklerozu nedeniyle çalışan kalpte baypas

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Introduction

Familial hypercholesterolemia is an autosomal-dominant disorder in which mutations in the low-density lipoprotein (LDL) receptor gene cause high levels of LDL and premature coronary artery disease in childhood (1). Extra coronary atherosclerotic lesions, including calcification of the aortic valve and root, are present in those patients. Atherosclerotic thickening and obstruction of the ascending aorta have also been documented in adult patients with familial hypercholesterolemia (2). When atherosclerosis involves the ascending aorta, coronary artery bypass grafting (CABG) becomes a high-risk procedure that can result in cerebrovascular complications. Coronary artery bypass grafting with the "no touch" aorta technique is preferred to avoid that devastating complication. However, it is unusual to apply this technique in pediatric patients who undergo CABG.

Case Report

A 12-year-old girl (weight, 27 kg) was admitted to our hospital with a history of chest pain of more than 2 years duration, during which time her angina symptom had ultimately become unstable. At the age of 5 years, she had been diagnosed with familial hypercholesterolemia, as had her twin sister. This patient had multiple xanthomas on her fingers, elbows, knees, and feet. Although she was being treated with cholestyramine and atorvastatin, her total cholesterol level was 675 mg/dL, her LDL level was 587 mg/dL, her high-density lipoprotein (HDL) level was 55 mg/dL and her triglyceride level was 164 mg/dL. The results of electrocardiography showed ST-T depression in the lateral leads, and echocardiographic Doppler scanning revealed mild aortic regurgitation. Left coronary ostial stenosis (95%) and irregularity of the ascending aorta were determined by coronary angiography (Fig. 1). Computed tomographic images of the thorax were obtained with a 4-detector scanner (Sensation 4, Siemens, Erlangen, Germany) without contrast enhancement, with contrast enhancement via a bolus of 40 mL of non iodinated contrast medium, and with electrocardiographic gating. The slice thickness and collimation were 2.5 mm. Diffuse; dense calcification was noted at the annulus level and diffuses but less dense calcification was detected in the entire ascending aorta and the aortic arch. The intimal side of the ascending aorta was very irregular and had a thickened wall (Fig. 2). Computed tomographic guidance enabled us to perform the surgical procedure without manipulating the aorta.

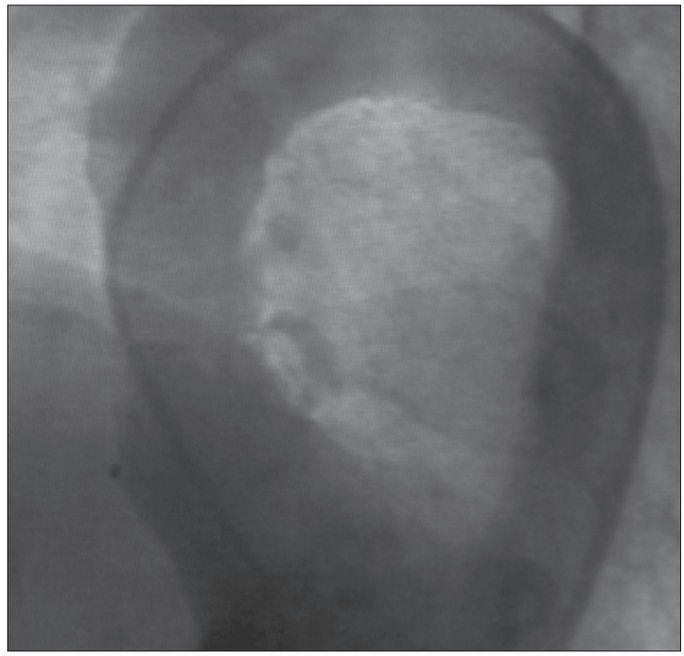

Figure 1. Coronary angiography reveals left main coronary artery ostial stenosis and irregularity of the ascending aorta

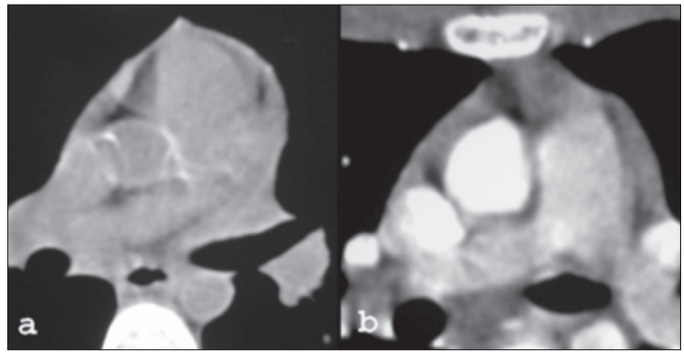

Figure 2. A) A nonenhanced computed tomographic scan shows that the entire ascending aorta is diffusely calcified
B) A contrast-enhanced computed tomographic scan in the upper portion of the ascending aorta shows the irregularity of the intimal surface of the ascending aorta

The patient underwent myocardial revascularization and anastomosis of the left internal thoracic artery to the left anterior descending artery was performed on the beating heart via median sternotomy. She was discharged from the hospital on the fourth postoperative day with antilipidemic therapy and instructions for a maintenance program of plasmapheresis every 2 weeks. Her control total cholesterol level was 278 mg/dL, her LDL cholesterol level was 234 mg/dL, her HDL cholesterol level was 28 mg/dL, and her triglyceride level was 81 mg/dL. At the time of her 38-month follow-up examination, the patient was free of angina.

Discussion

Coronary artery bypass grafting is performed in pediatric patients with familial hypercholesterolemia and the graft of choice is the internal mammary artery, which has the potential for growth and prolonged patency (3, 4). Bilateral internal mammary artery usage is also docu-

mented in childhood period (4). Conventional CABG is usually performed in children; off-pump CABG is rarely preferred for use in pediatric patients because of the small diameter of their vessels (5).

Extracardiac vascular involvement (including that involving the aortic root and carotid artery) has been documented in children with familial hypercholesterolemia and the incidence of such involvement increases with the age of the patient. Evaluation for atherosclerosis of the aorta is not routinely performed preoperatively in children, although atheroembolism from the ascending aorta is a major etiologic factor for stroke in adult patients undergoing cardiac surgery. Cohen et al. (6) showed that non calcified plaques are associated with a higher risk of vascular events and surgical manipulation has been reported to cause new mobile lesions in a diseased aorta (7). Soft atheromas are most likely to embolized as a result of manipulation. In our patient, the preoperatively detected atheromatous ascending aorta led us to prefer the use of off-pump CABG, because the non calcified, unstable, lipid-laden plaques were thought to have the potential to form emboli.

Conclusion

To prevent procedure-related neurologic complications, the possibility of diffuse and dense atherosclerosis of the ascending aorta must be kept in mind in children with familial hypercholesterolemia who undergo CABG.

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Available Online Date/Çevrimiçi Yayın Tarihi: 18.05.2011

doi:10.5152/akd.2011.090

Kawasaki disease presenting as meningitis in a two months old infant

İki aylık bir bebekte menenjit biçiminde ortaya çıkan Kawasaki hastalığı

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Introduction

Kawasaki disease (KD), is an acute febrile multisystem vasculitic syndrome characterized by fever, bilateral non-exudative conjunctivitis, erythema of lips and oral mucosa, cervical lymphadenopathy, changes in extremities and polymorphous exanthema (1). Although infants and young children have the highest incidence of KD, it is rarerly reported in infants ≤3 months of age (2). The diagnosis in this age group is difficult because the presentation is usually incomplete and similar to other diseases (3).

In this report, we describe an 8 week old infant with KD to remind that suspicion and proper evaluation are necessary for timely diagnosis and treatment.

Case Report

A two months old boy presented with fever diarrhea and vomiting. On admission body temperature was 38.5°C, skin turgor normal, lung and hearth were unremarkable at examination. Laboratory investigations revealed white blood cells (WBC) 10. 400/mm^3, erythrocyte sedimentation rate 85 mm hourly, hemoglobin 9.6 g/dl and platelet count 351.000/mm^3. Urinary analysis revealed 25 leukocytes per high power field and cerebrospinal fluid (CSF) examination revealed pleocytosis with normal glucose and protein values. Antibiotic therapy was initiated but fever persisted and a generalized macular rash on his trunk and edema of extremities appeared on sixth day of admission. Leukocytosis was detected and C-reactive protein increased to 68 mg/dL, which was normal at the beginning. His blood, CSF and urine cultures remained sterile. The next day hypoalbuminemia and generalized edema developed. On 12th day of his fever, physical examination revealed tachycardia with an S3 gallop rhythm. Red fissured lips, desquamation of fingers, thrombocytosis and perianal dermatitis accompanied other findings. An echocardiographic examination demonstrated dilation of both coronary arteries (right - 3.9 mm, z score -3.83, left main coronary artery - 3.7 mm, z score - 3.66) (Fig. 1, 2) and minimal mitral and aortic regurgitation. Cardiac contractions were in normal range.

High-dose intravenous gammaglobulin (IVIG) and aspirin were administered with a diagnosis of KD. Fever subsided only after a second dose of IVIG. His hemoglobin decreased progressively to 3.8 g/dl at 16th day of admission and he was transfused with erythrocyte suspensions three times during his stay in hospital. Repeated echocardiogram pointed out coronary artery aneurysm formation and on follow-up coronary artery dilations persisted necessitating continuation of aspirin at a dose 3-5 mg/kg/day.