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

**Familyal hiperkolesterolemili çocukta çıkan aortanın erken aterosklerozu nedeniyle çalışılan kalpte bypass**

Öner Gülcan, Selman Vefa Yıldırım*, Rıza Türköz
Department of Cardiovascular Surgery and *Pediatric Cardiology, Adana Teaching and Medical Research Center, Başkent University, Adana-Turkey

**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 noniodinated 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.

**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-
Kawasaki disease presenting as meningitis in a two months old infant

İki aylık bir beşekte menenjit biçiminde ortaya çıkan Kawasaki hastalığı

Özden Türel, Alper Güzeltas1, Çiğdem Aydoğmuş, Nevin Hatipoğlu, Hüsem Hatipoğlu, Rengin Siraneci
Clinic of Pediatric, Bakırköy Maternity and Children’s Hospital, İstanbul
1Clinic of Pediatric Cardiology, Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul-Turkey

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 exantheme (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/mm3, erythrocyte sedimentation rate 85 mm hourly, hemoglobin 9.6 g/dl and platelet count 351,000/mm3. 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 hypalbuminemia 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, throratomy and perianal dermatitis accompanied other findings. An echocardiographic examination demonstrated dilatation 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.