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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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.

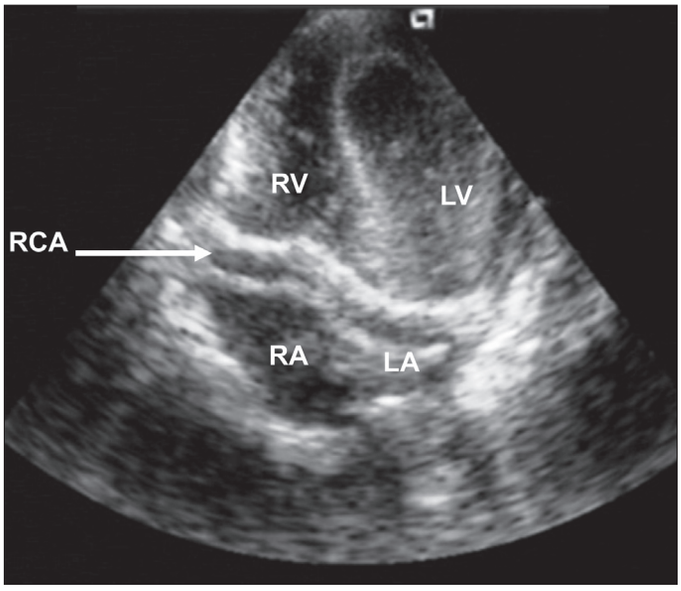

Figure 1. Apical four-chamber echocardiographic image: dilated right coronary artery (arrow)
LA - left atrium, LV - left ventricle, RA - right atrium, RCA - right coronary artery, RV - right ventricle

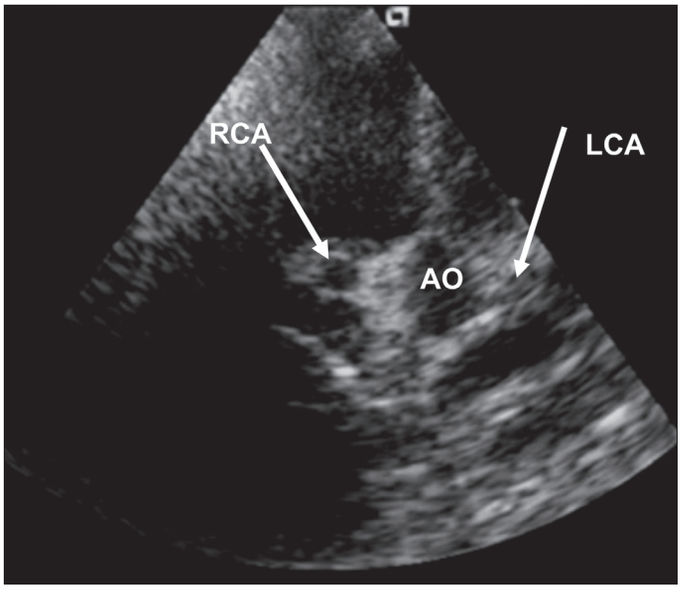

Figure 2. Modified apical five-chamber echocardiographic image: dilated right and left coronary arteries (arrows)
AO - aorta, LCA - left coronary artery, RCA - right coronary artery

Discussion

It is well known that there is a higher incidence of incomplete presentations in younger patients with KD (3, 4). It was reported that super antigen neutralization by transplacental antibody by mothers, cross reaction of antibody generated by frequently active immunization and weak vasculitis phenomenon caused by inadequate immune response in this age period may explain the incomplete presentation and late diagnosis in young patients (5).

Another reason for difficulty in diagnosis is similarity with other illnesses. In the reported case, urinary tract infection and meningitis were considered at the beginning. According to the algorithm presented by Newburger et al. (4) infants ≤6 months old on day ≥7 of fever without other explanation for the febrile illness should undergo laboratory testing and, if evidence of systemic inflammation is found, an echocardiogram should be considered even if they have no clinical criteria.

Newburger et al. (4) also suggested that patients who have unusual manifestations should be called atypical KD. Our patient was atypical in some aspects including age at presentation, generalized anasarca type edema and severe anemia requiring multiple transfusions. In a survey covering a period of 25 years, infants under 3 months constituted only 1.6% of patients with KD (2). The youngest reported case is that of a Japanese girl with onset at 8 days of age (6). Peripheral edema involving hands and feet is quite common but generalized edema has not been reported. Young patients have higher WBC counts, higher platelet counts and lower hemoglobin values which suggest a more severe inflammatory process (3). Nevertheless severe hemolytic anemia requiring transfusions is rare and may be related to IVIG infusion (7).

Conclusion

Young infants with KD are at increased risk for formation of coronary artery aneurysms due to delayed diagnosis and decreased responsiveness to IVIG treatment (8). In addition to his age our patient had other bad prognostic signs including male sex, prolonged fever in spite of IVIG treatment, anemia and hypoalbuminemia. Echocardiogram is an important implement in diagnosis which would help treatment if performed earlier.

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