Letter to Editor

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Congestive Heart Failure – An Atypical Presentation of Kawasaki Disease

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Kawasaki disease (KD) is an acute febrile mucocutaneous lymph node syndrome with multisystem vasculitis mainly affecting infants and children less than 5 years of age, first described by Tomisaku Kawasaki from Japan in 1967[1]. In the absence of a diagnostic test for KD, the diagnosis is established when fever and four of the remaining five principal symptoms are present. In this report we describe 3 children who presented atypically with prolonged fever and congestive heart failure (CHF) that does not belong to principal symptomatology of KD[2,3].

Case 1: A 2 year 7 month old male child presented with history of fever for 10 days along with occasional vomiting and abdominal pain. There was history of sudden onset of shortness of breath, puffy eyelids and swollen feet on 11th day of illness for which he was referred to our hospital. On admission the child was clinically diagnosed to have congestive heart failure. Investigations revealed Hemoglobin (Hb) 10 gm%, white blood cell (WBC) 20000/cmm, (N 76,, L 22), platelet 605000/cmm, erythrocyte sedimentation rate (ESR) 80 mm/hr and C-reactive protein (CRP) 48 mg/L. Serum levels of urea creatinine, sodium and potassium were normal. Chest x-ray revealed cardiomegaly with perihilar infiltrates. He was treated conservatively for CHF, with fluid and sodium restriction, furosemide and captopril. He was started with IV antibiotics after urine and blood culture samples were taken. Fever did not subside and there was no improvement of CHF even after 72 hrs of therapy. In the meantime, urine and blood culture reports came to be negative. A two-dimensional echocardiography revealed aneurysmal dilatation of coronary arteries. High dose intravenous immunoglobulin (IVIG) (2 mg/kg over 12-24 hour infusion) and oral aspirin (80-100 mg/kg/day). The child responded very quickly, fever subsided and features of CHF improved within 24 hours of therapy.

Case 2: A 4 year 8 month old female presented with breathlessness, easy fatigability, and poor feeding along with history of continuous fever for 12 days. There was history of maculopapular eruption mainly over trunk and upper extremities on 4th day of fever and it disappeared completely by 7th day of illness. On admission, the child had features of CHF in the form of tachycardia, tachypnea, gallop rhythm and hepatomegaly. Investigations revealed Hb 11.5 gm%, WBC 17600/cmm (N 82, L 18), platelet 569000/cmm, ESR 100 mm/hr, CRP 24 mg/L, urine and blood culture no growth. Chest x-ray showed patchy infiltrates in perihilar region. She was initially put on anticongestive measures along with IV antibiotics. Echocardiography revealed aneurysmal dilatation of coronary arteries and diagnosis of KD was made. She responded dramatically with IVIG therapy.

Case 3: A 5 year old male was admitted with history of long-continued fever for 25 days. On admission, clinical examination was non-contributory and investigations were started in the line of pyrexia of unknown origin. 2 days after admission, he developed sudden onset of respiratory distress and was found to have tachycardia, tachypnea, and basal rales on chest auscultation. His feet were swollen and edematous. There was peeling of skin around toe-nails. Investigations revealed Hb 10.8 gm%, WBC 31700/cmm (N84, L14) platelet 378000/ cmm, ESR 114 mm/hr, CRP 48 mg/L. Other labrotary tests (serum urea, creatinine levels, liver function test, widal test, and cerebrospinal study) were normal. Blood and urine cultures

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showed no growth. Chest x-ray showed mild cardiomegaly. He was also put on IV antibiotics and anticongestive measures for CHF. But there was no clinical improvement. When echocardiography revealed aneurysmal dilatation of left main and anterior descending coronary arteries, diagnosis of atypical KD was made. IVIG therapy quickly improved the clinical condition.

Kawasaki disease is now known to be the commonest vasculitis in childhood[4]. Diagnosis of KD is based on principal symptoms as proposed in diagnostic criteria for KD.

At least five principal symptoms must be present for the diagnosis of KD. However, patients with four of the principal symptoms can be diagnosed to have KD when coronary artery aneurysm is recognized by echocardiography or coronary angiography. It is also said that if fever of unknown etiology continues for 10 days or more, and if one or two of the principal symptoms are present, then atypical KD can be diagnosed and IVIG treatment should be instituted[5,6].

In this report, we did not think of KD in the differential diagnosis because all cases presented with prolonged fever and features of CHF. Swollen feet in case 1 and case 3 was considered as manifestation of CHF, history of maculopapular eruption in case 2 was ascribed by parents as drug rash, and periungual desquamation in case 3 was misinterpreted as a non-specific skin lesion. Even 2D-echocardiography was delayed for 5–7 days after admission. Actually, in the absence of cardiac defect, we thought that CHF was due to fever and infection. When urine and blood culture reports came to be negative and there was no clinical improvement with conventional antibiotics and anticongestive measures, echocardiographic evaluation was undertaken. All cases showed aneurysmal dilatation of coronary arteries on echocardiogram. Literature supports the view that at least 10% of children with characteristic coronary artery aneurysms of KD never actually fulfill criteria for KD[7]. So our cases were diagnosed as atypical KD.

Hematological investigations revealed neutrophilic leucocytosis, elevation of ESR and CRP, normal to increased platelet count in all cases. Though the patients presented with CHF, their ESR values were very high. It is a contradictory finding because ESR is generally low in CHF[8].

Follow-up echocardiography was done in all cases at 6 weeks, 3 months, 6 months and 12 months after IVIG therapy. All follow-up echos were normal. All 3 children remain well on regular follow-up. Cases 1, 2 and 3 have completed their follow-up for 17 months, 15 months and 12 months respectively.

Key Words: Kawasaki Disease; Congestive Heart Failure; Coronary Artery; Aneurysm

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