

Kawasaki's disease: An unusual presentation

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ABSTRACT

Atypical presentations of Kawasaki's disease have been described in the form of intestinal pseudo-obstruction, tonsillitis, hemorrhagic serous effusions, thrombocytopenia, and non-fulfillment of all criteria for diagnosis of Kawasaki's disease. However, presentation of Kawasaki's disease with shock and need for inotropic support have been rarely described. We present a 4-year-old girl with Kawasaki's disease who presented with anasarca, oliguria, shock, and presence of dilated coronary arteries within 5 days of fever and responded to intravenous immunoglobulin (IVIG) and inotropic support.

Key words: Atypical presentation, children, Kawasaki's disease

INTRODUCTION

Kawasaki's disease is an acute febrile multisystem vasculitis of early childhood with a striking predilection for the coronary arteries.^[1] Patients usually present with fleeting macular skin eruptions, fever, leukocytosis, conjunctivitis, cervical adenitis, strawberry tongue, and oral cheilosis.^[2] If the condition is unrecognized or treated sub-optimally, there is a 25% risk of developing serious cardiovascular complications.^[3] Early diagnosis and prompt treatment with aspirin and intravenous immunoglobulin (IVIG) within 10 days of illness can reduce coronary artery lesions to 5%.^[4] Thus, high suspicion of Kawasaki's disease and a watch for atypical presentations can prevent coronary complications. We present a 4-year-old girl who presented with fever, shock, and anasarca and developed coronary dilation within 5 days of admission.

CASE REPORT

A 4-year-old female child presented with intermittent fever since 4 days, generalized edema with oliguria since 2 days, erythematous rash that started on trunk and limbs simultaneously since 1 day, and vomiting - 2 episodes. There was no history of jaundice or similar episode in the past. On examination, she was febrile with tachycardia (heart rate = 134/min) with signs of shock (hypotension: blood pressure = 88/50 mm Hg, poor peripheral perfusion). She had anasarca with macular erythematous rash over trunk, hands, and legs. She had multiple, nontender, nonmatted cervical lymphadenopathy, with largest being 1.5 × 1.5 cm, and oral cheilosis. There was no strawberry tongue or conjunctival congestion. On systemic examination, she had tender hepatomegaly with splenomegaly. A differential diagnosis of dengue, leptospirosis, sepsis, and Kawasaki's disease was considered. Her investigations initially showed hemoglobin of 8.7 gm%, white cell count of 10,000/cu.mm (72% polymorphs, 26% lymphocytes, 1% monocytes), platelets = 430,000 cells/cu.mm, and ESR of 4 mm at end of 1 h. Her blood culture was negative for any organism and peripheral smear showed no malarial parasites. Her liver function tests were deranged with SGPT of 155 IU/L, total proteins of 4.8 g/dL, and albumin of 2 g/dL. Her dengue IgM and

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leptospira IgM were negative. Cardioscope monitoring did not show any arrhythmia or any abnormal wave. Serum creatinine and CPK were normal. She was treated with IV Ceftriaxone and normal saline bolus, and because blood pressure did not improve, Dopamine was added. However, fever persisted and she developed red lips and strawberry tongue although rash disappeared in the next 2 days. Dopamine was stopped after 4 days of admission. Her repeat hemogram after 5 days showed platelet count of 477,000/cu.mm and ESR of 135 mm at end of 1 h. A 2D echocardiography was done, which showed mild pericardial effusion and dilated coronary arteries (Right main coronary artery = 2.8 mm, left anterior descending = 2.2 mm). She was treated with intravenous immunoglobulin (IVIG) on the suspicion of Kawasaki's disease (2 g/kg) to which the patient responded. The edema subsided, inotropic support was omitted, and ESR decreased to 40 mm on Day 9 of admission and 20 mm on Day 15. She developed desquamation of the soles and palms in the 3rd week of illness. She was started on Aspirin and advised regular follow-up.

DISCUSSION

In a literature review of atypical Kawasaki's disease, less than 150 case reports of atypical Kawasaki's disease have been reported.^[1-8] Most atypical presentations of this mucocutaneous lymph node syndrome have been reported in infants.^[5,8,9] Also, incidences of coronary artery complications are more common in these infants.^[5] In older children, atypical features reported have included co-infection with cytomegalovirus (CMV),^[1] intestinal pseudo-obstruction,^[7] acute tonsillitis,^[3] hemorrhagic serous effusions with hepatic dysfunction, and thrombocytopenia.^[6] Reports of Kawasaki's disease presenting with shock is rare.^[10-12] Compared with patients without shock, patients with Kawasaki's disease shock syndrome are more often female and have higher C-reactive protein concentrations, and lower hemoglobin concentrations and platelet counts^[10] as was seen in our patient. They may require anti-inflammatory in addition to IVIG.^[10] Our patient, however, responded to IVIG. Shock in Kawasaki's disease with edema may be due to an important inflammatory state and vascular leakage, as our patient showed no signs of cardiac dysfunction on echocardiography. This is similar to that described by Natterer *et al.* in 3 children in 2011.^[11] Terai *et al.* have described that the increased vascular permeability in the acute phase of Kawasaki's disease is in relation with the elevation of vascular endothelial growth factor expression. They found a direct relation between the severity of edema and the

development of coronary aneurysms. They concluded that vascular leakage may be a key feature of the physiopathology of Kawasaki's disease.^[12]

Although our patient had rash, cervical lymphadenopathy, fever, and oral cheilosis, other features such as shock, hepatosplenomegaly, and edema were atypical. Owing to the fact that the diagnosis of this disease is infrequent in the pediatric intensive care unit and that the way of presentation with shock may mimic toxic shock syndrome or sepsis, there is a risk of delayed diagnosis and, therefore, delayed treatment. A high suspicion made us do echocardiography, which showed dilated coronary arteries. Thus, a high suspicion of Kawasaki's disease should be considered in patients who do not meet all the criteria of diagnosis, and early echocardiography with early treatment may help to prevent complications and treatment failure.

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