International Journal of Contemporary Pediatrics
Thumjaa A et al. Int J Contemp Pediatr. 2014 Aug;1(2):122-124
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291
DOI: 10.5455/2349-3291.ijcp20140813

Case Report

Atypical Kawasaki disease

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Received: 28 July 2014
Accepted: 5 August 2014

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ABSTRACT

A five year old girl presented with fever for about three weeks, rashes for about 1 week with mild congestion of eyes, redness of mucosa of oropharynx, strawberry tongue, and desquamation of skin in the perineal region. Investigation showed Pleomorphic leukocytosis and thrombocytosis, all bacterial cultures were negative, echocardiography was normal. We had a high suspicion of Kawasaki disease with atypical presentation, therefore it should be quickly suspected in unexplained prolonged febrile children with associated features for early diagnosis and treatment to prevent coronary artery abnormality.

Keywords: Atypical Kawasaki disease, Diagnostic criteria, Treatment of Kawasaki disease

INTRODUCTION

Kawasaki disease was first described by Dr. Tomisaku kawasaki in 1967.[1] Since then hundred thousand cases has been reported worldwide.[2] Kawasaki disease predominantly affects young children. In developed countries Kawasaki disease is now considered to be the commonest cause of acquired heart disease in children.[3] We report a case of atypical presentation of Kawasaki disease.

CASE REPORT

A 5 year old girl presented with fever for about 3 weeks, rashes over the trunk and back for 1 week. She has been treated in two separate hospitals. She was febrile, physical examination revealed mild congestion of eyes, dry and cracked lips, redness of mucosa of oropharynx, she had strawberry tongue (Figure 1), maculopapular rash over the trunk and back, desquamation of skin over the perineal region (Figure 2), no significant palpable lymph nodes, palms and soles was normal. Systemic examination was normal including cardiovascular system.

Initial laboratory findings showed white blood cell count - 22000/mm^3, with a differential count of 77% neutrophils, 20% of lymphocytes, 3 % of monocytes. The platelet count was 532000/mm^3, ESR was normal, CRP was negative, urine routine was normal with negative culture, blood culture was normal, Widal - negative, renal function test and liver function test was normal, chest X-ray was normal.

Day 1 we had a high clinical suspicion that the illness might be atypical presentation of Kawasaki disease, patient was sent to cardiology unit for echocardiography which was normal. Even though with high clinical suspicion of Kawasaki disease, the patient was started on high dose aspirin 100 mg/kg/day and IVIG infusion was given over 24 hours. The patient improved and she became afebrile in 48 hours after the treatment, and she was discharged after one week with low dose aspirin 5 mg/kg/day for 6 weeks. After 6 weeks, all the clinical symptoms and signs were disappeared, complete blood count was normal, repeat echocardiography was done which was also normal.

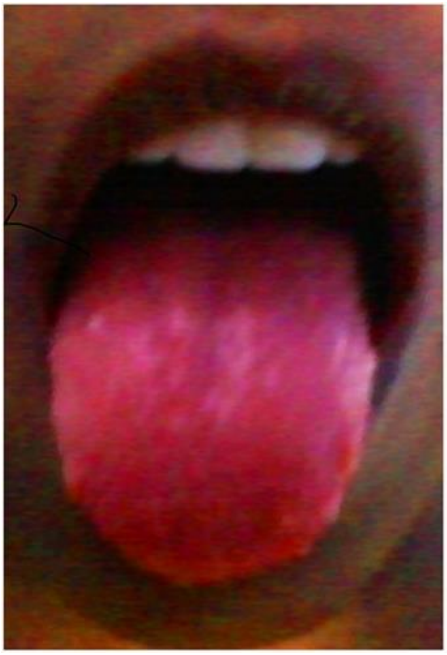

Table 1: Dry and cracked lips with strawberry tongue.

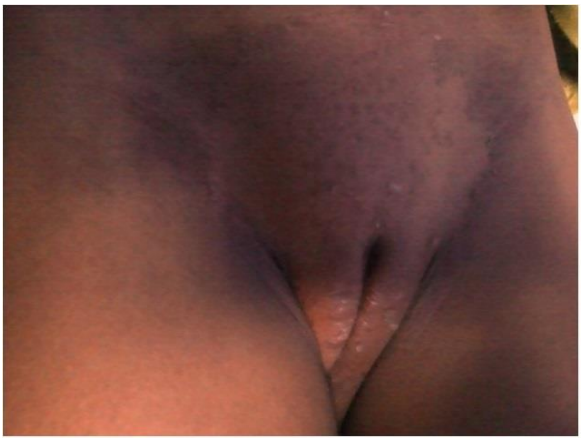

Table 2: Desquamation of skin in the perineal region.

DISCUSSION

As the exact etiology of Kawasaki disease and its specific laboratory tests remains unknown, the diagnosis of Kawasaki disease mainly depends upon the clinical presentation. The diagnostic criteria of Kawasaki disease is extremely useful, lack of such criteria may result in atypical features of the illness has in our patient. The essential diagnostic criteria of Kawasaki disease is

1) Fever lasting 5days or more.

2) Presence of atleast 4 of the following 5 principal criteria

   a. Bilateral non purulent conjunctivitis

   b. Redness of mucosa of oropharynx, strawberry tongue, fissured lips

   c. Changes in extremities such as edema and erythema of Hands and feet and later periungual desquamation which may involve palms and soles.

   d. Polymorphous vesicular rash

   e. Cervical lymphadenopathy of atleast 1.5 cm in size usually unilateral

3) Illness not explained by another known disease.

In our patient there was fever for 3 weeks, mild congestion of eyes, redness of mucosa of oropharynx, strawberry tongue, dry and cracked lips, desquamation of skin in the perineal region, maculopapular rash over trunk and abdomen. With the essential diagnostic criteria our patient did not fulfill the criteria, where the patient had prolonged unexplained fever with less than 4 of the following 5 criteria, which is said to be atypical Kawasaki disease. Even though our patient did not have classical form of Kawasaki disease, with the high suspicion of atypical presentation of Kawasaki disease our patient was treated and improved.

Atypical Kawasaki disease is not a mild form of Kawasaki disease, children remain at similar risk of cardiovascular sequelae as that of Kawasaki disease,[4] since the disease has similar risk of coronary artery abnormality as Kawasaki disease,[5-8] it is necessary to make an accurate diagnosis in order to prevent the development of coronary artery abnormality.[4-7] Kawasaki disease should be considered in every infant presenting with long lasting unexplained fever with associated features, because children less than five years are at an extremely high risk of developing coronary arterial abnormalities, early diagnosis and appropriate therapy are important. Pediatrician who evaluates the children with unexplained prolonged fever with or without classical forms of Kawasaki disease should have high suspicion for the possibility of Kawasaki disease.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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DOI: 10.5455/2349-3291.ijcp20140813
Cite this article as: Thumjaa A, Selvakumar C. Atypical Kawasaki disease. Int J Contemp Pediatr 2014;1:122-4.