Case Report

Incomplete Kawasaki Disease in a Three Month Old Infant

Ashraf M, Taleb A, Shamsah A, Al-Sharhan H

From, Department of Pediatrics, Pediatric Emergency Unit, Mubarak Al-Kabeer Hospital, Kuwait

Correspondence to: Dr Hind Al-Sharhan, Department of Pediatrics, Pediatric Emergency Unit, Mubarak Al-Kabeer Hospital, Kuwait. Email - hd.alsharhan@live.com

Received: 04 December 2014 Initial Review: 20 December 2014 Accepted: 31 December 2014 Published Online: 09 January 2015

ABSTRACT

Kawasaki disease (KD) is an acute systemic vasculitis of unknown aetiology that has largely replaced rheumatic heart disease as a cause of acquired heart disease in children of many developed countries. We report a case of incomplete KD in a three-month-old girl. The diagnosis of incomplete KD was made after exclusion of conditions with similar presentation. She was treated with intravenous immunoglobulin following which she made an uneventful recovery but demonstrated thrombocytosis in the second week of convalescence. It is important for the treating physicians to become aware of the incomplete KD as prompt diagnosis and early treatment of these patients with intravenous immunoglobulin is vital for the prevention of lethal coronary complications. Physicians need to have a "high index of suspicion" for KD and even, higher for IKD.

Keywords: *Coronary ectasia, Incomplete Kawasaki Disease (IKD), Thrombocytosis, Vasculitis*

Kawasaki disease (KD) is an acute vasculitis of unknown etiology that has been found in all pediatric age groups. However, most cases occur in children aged 6 months to 8 years old. After Henoch-Schonlein purpura, KD is the commonest vasculitic disorder of children [1-6]. It is a disease of variable incidence. In Japan, the incidence is 80-100 per 100,000 children under five years of age, 8 in the USA, 3-6 in Europe [1-3]. Despite such high incidence worldwide, the number of reported cases from Kuwait remains meager and is largely confined to typical KD. It is likely that many cases of KD and more of incomplete KD (IKD) go unnoticed, especially in the absence of a precise diagnostic test.

The diagnosis is confirmed by the presence of fever lasting for at least 5 days and four of the following criteria, without evidence of another known disease: a) bilateral non-purulent conjuctival injection; b) changes of mucous membranes in the upper respiratory tract such as injected pharynx and lips, strawberry tongue; c) polymorphous rash; d) changes in the extremities such as peripheral edema, erythema or periungual desquamation; and e) acute, usually unilateral, non-purulent cervical adenopathy. Since there is no single diagnostic laboratory test, physicians use laboratory markers of inflammation (high white blood cell count, C-reactive protein, and erythrocyte sedimentation rate) to establish the diagnosis of Kawasaki disease. In some cases not all the criteria are met, and the coronary artery abnormalities develop later as a complication. The term incomplete (or atypical) Kawasaki is used for such patients [7-9]. Coronary artery aneurysms develop in 20–25% of untreated patients but administration of intravenous immunoglobulin within the first 10 days of fever significantly reduces this incidence [9]. We report a case of a three month old infant with IKD who presented with fever followed by red lips, strawberry tongue and non purulent conjunctivitis.

CASE REPORT

A three months old girl presented with six days history of fever and lethargy. There was no history of vomiting or diarrhea. Her past medical history was uneventful.

Systemic review was noncontributory apart from mild eczema. She was seen three times by different pediatricians who diagnosed her as having acute otitis media; she was given amoxicillin with clavulinic acid and cefpodoxime respectively, with no changes in her overall wellbeing despite treatment for five days.

On physical examination, she looked unwell; her temperature: 39.6^{o}c; oxygen saturation: 98% in room air; heart rate: 150 per minute; respiratory rate: 35 per minute; there was obvious non purulent conjunctivitis and mucus membrane changes in the form of red lips and strawberry tongue.

The mother agreed that the above two features appeared within the preceding 48 hours. There were no skin rashes, no lymph node enlargement and no extremity changes.

Although, she was three month old with only two features of Kawasaki disease, IKD was suspected and the following investigations were ordered. Her complete blood count (CBC) showed white blood cell count (WBC) of 27,000/mm^3; hemoglobin (Hb): 9.7 gm/dl, platelets: 915,000/mm^3; erythrocyte sedimentation rate (ESR): 97 mm/hour; C reactive protein (CRP): 75 mg/dl; There was markedly elevated liver transaminases. Urine analysis showed mild pyuria (30/ml). Blood and urine samples were also collected for culture and sensitivity which were sterile.

The clinical features, the duration of fever, the unresponsiveness to antibiotics and the laboratory results, all indicated that the diagnosis of IKD was highly probable. Accordingly, she was immediately started on IVIG and high dose aspirin. Pediatric cardiologist was consulted and echocardiogram was done and it was unremarkable. She showed dramatic response to IVIG within 24 hours. Her fever subsided, her conjuctival and mucus membrane changes improved. She was kept under observation for another 24 hours during which she remained asymptomatic then she was discharged home on aspirin.

A follow up CBC and Echocardiogram were arranged. Platelet count normalized within 4 weeks, her second Echocardiogram (after 6 weeks) and third Echocardiogram (after 3 months) showed coronary ectasia, consequently low dose aspirin was continued.

DISCUSSION

Kawasaki disease is a generalized vasculitis that affects medium-size arteries. Ninety percent of cases involve children younger than five years. The disease is relatively uncommon in children younger than six months. Boys are affected about 50% more often than the girls. The disease occurs year round, but cases tend to cluster in the winter and spring [10]. Patients may be found to have incomplete Kawasaki disease if they exhibit fever and some of the classic symptoms but not enough to meet the diagnostic criteria. Infants, especially those younger than six months, are especially likely to have an incomplete presentation. The finding most consistently absent in incomplete Kawasaki disease is cervical lymphadenopathy (present in only 10%). Rash is absent in 50%, and extremity changes are absent in 40%. Mucous membrane changes are the most consistent finding.

Laboratory evaluation is recommended for the patients a) younger than six months with unexplained fever for ≥7 days, even if they exhibit none of the classic symptoms of Kawasaki disease and b) of any age with unexplained fever for ≥5 days who exhibit three or fewer of the classic symptoms [11]. Laboratory evaluation should include complete blood cell count, ESR, CRP level, urinalysis, alanine aminotransferase level, and serum albumin level.

The disease tends to be self-limiting and usually resolves without treatment within about 12 days [12]. However, serious cardiac complications can occur, such as coronary artery aneurysms, decreased myocardial contractility, congestive heart failure (CHF), arrhythmias, and myocardial ischemia. Early recognition and treatment significantly reduces the incidence of these complications. Without treatment, 20% to 25% of patients develop cardiac complications while with treatment, the incidence decreases to 4% [10-14]. Treatment should be initiated as soon as the diagnosis is made and should involve the administration of intravenous immunoglobulin (IVIG) and high-dose aspirin.

The overall prognosis for patients with Kawasaki disease is dependent on the severity of coronary artery involvement as a risk factor for myocardial ischemia [15]. Patients without any cardiovascular abnormalities tend to do well and are generally asymptomatic at their long-term follow-up examination.

CONCLUSION

It is important for the treating physicians to become aware of the incomplete KD as prompt diagnosis and early treatment of these patients with intravenous immunoglobulin is vital for the prevention of lethal coronary complications. Physicians need to have a "high index of suspicion" for KD and even, higher for IKD.

REFERENCES

1. Melish ME, Hicks RV. Kawasaki syndrome: clinical features, pathophysiology, etiology and therapy. J Rheumatol 1990;17:2-10.
2. Dhillon R, Newton L, Rudd PT, Hall SM. Management of Kawasaki disease in the British Isles. Arch Dis Child 1993;69:631-38.
3. Schiller B, Fasth A, Bjorkhem G, Elinder G. Kawasaki disease in Sweden: incidence and clinical features. Acta Paediatr 1995;84:769-74.
4. Paul DK, Gupta A, Lahiri M. Kawasaki Disease in Calcutta. Indian Pediatr 2000;37:1264-5.
5. Singh S, Kumar L. Kawasaki disease-treatment with intravenous immunoglobulin during acute stage. Indian Pediatr 1996;33:689-92.
6. Singh S, Kumar L, Trehan A, Marwaha RK. Kawasaki Disease at Chandigarh. Indian Pediatr 1997;34:822-5.
7. McMaster P, Cooper S, Isaacs D. Is it Kawasaki disease? J Pediatr Child Health 2000;36:506-8.
8. Rowley AH, Goonzalez-Crussi F, Gidding SS, Duffy CE, Shulman ST. Incomplete Kawasaki disease with coronary artery involvement. J Pediatr 1987;110:409-13.
9. Burns JC, Glode MP. Kawasaki syndrome. Lancet 2004;364:533–44.
10. Sundel R. Epidemiology and etiology of Kawasaki disease [monograph on the Internet]. Waltham (MA): UpToDate, inc; 2008 Jun 3 [cited 2008 Nov 4]. Available from:www.utdol.com/patients/content/topic.do?topicKey=~45zSjiWwxeTWX4.
11. Bourusly M, Jawad N. Case report: Kawasaki disease in a Kuwaiti family. Kuwait Medical Journal 2005;37(2):110–2.
12. Sundel R. Clinical manifestations and diagnosis of Kawasaki disease [monograph on the Internet]. Waltham (MA): UpToDate, Inc; 2008 Jun 3 [cited 2008 Nov 4]. Available from: www.utdol.com/patients/content/topic.do.
13. Yanagihara R, Todd JK. Acute febrile mucocutaneous lymph node syndrome. Am J Dis Child 1980 Jun;134(6):603–14.
14. Husson RN, Mark EJ. Case records of the Massachusetts General Hospital. Weekly clinicopathology exercises. Case 23-1999. A 10-month-old girl with fever, upper lobe pneumonia, and a pleural effusion. N Engl J Med 1999 Jul 29;341(5):353–60.
15. Newburger JW, Takahashi M, Gerber MA, et al. Diagnosis, treatment, and long-term management of Kawasaki disease: a statement for health professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Circulation 2004;110(17):2747-71.

How to cite this article: Ashraf M, Taleb A, Shamsah A, Al-Sharhan H. Incomplete Kawasaki Disease in a Three Month Old Infant. Indian J Case Reports. 2015;1(1):11-13.

Conflict of interest: None stated, Funding: Nil