January 2009

Incomplete Kawaski disease: are we missing it

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Case Report

Incomplete Kawaski Disease: Are we missing it?

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Abstract

Kawasaki disease, also known as mucocutaneous lymph node syndrome or infantile polyarteritis nodosa is an acute febrile vasculitis of unknown etiology with a predilection for coronary arteries and potential for aneurysm formation. In Incomplete Kawasaki disease, children with fever lack the sufficient number of criteria to fulfill the epidemiologic case definition and are diagnosed when coronary artery disease is detected. We present a case report of a one and a half years old girl who came with features of incomplete Kawasaki disease, high grade fever, irritability, history of conjunctivitis and cracking of lips. She was investigated and had a platelet count of 902 x 10^9/L, ESR was 71mm/hr and CRP was also raised to 12.8mg/l. Cardiac evaluation and echocardiography was done which showed dilated coronary arteries >3mm on the left side and 4mm on the right side with early aneurysmal changes . She was treated with immunoglobulin and aspirin and improved.

Introduction

Kawasaki disease is more common in Asian children and in children younger than five years.[1] In developing countries it has replaced rheumatic heart disease as the most common acquired heart disease in children.[2] The diagnosis of Kawasaki requires the presence of fever for at least five days and at least four of the following five signs: Bilateral bulbar conjuctival injection generally non purulent, changes in the mucosa of the oropharynx including injected pharynx, dry fissured lips and strawberry tongue, rash which is polymorphous and primarily truncal, changes of the peripheral extremities such as oedema or erythema of hands and feet in the acute phase and periungal desquamation in the sub acute phase and cervical lymphadenopathy >1.5 cm usually unilateral.[3] Incomplete cases in which a child has fever with fewer than four other features of the illness and then develops coronary artery disease have been described.[3] These cases are more common in infants. We present a case of Incomplete Kawasaki disease in a child.

Case Report

A one and a half year old immunized girl weighing 10 kg came with the complaint of fever for fifteen days. The fever was high grade and relieved only temporarily with anti pyretics. Along with the fever the child also developed bilateral non purulent conjunctivitis which subsided in three to four days. For these complaints she was admitted to a private hospital where she was treated as a case of enteric fever. The mother gave a history of cracking and redness of lips which improved in 3 days. She received injection ceftriaxone and then syrup cefixime for a total of ten days. But the fever was still persistent. The laboratory tests done at that time showed a raised C.R.P of 15 mg/dl but the rest of the laboratory examination was normal. When we examined the child she was febrile and very irritable and her temperature was 40°C. She was anaemic and her throat was found to be hyperaemic. There were no signs of conjunctivitis or mucosal involvement at that time and rest of the general physical examination was normal. On systemic examination the only finding was a grade 2/5 systolic murmur audible at the apex. The laboratory examination done revealed an Hb: 9.2 gm/dl, TLC: 23,000/mm^3 with a neutrophil count of 55% and lymphocytes of 45%.The platelet count was also raised to 902 x 10^9/L. The ESR was 71mm/hr and CRP was also raised to 12.8mg/l. Cardiac evaluation and echocardiography was done which showed dilated coronary arteries >3mm on the left side and 4mm on the right side with early aneurysmal changes. On the basis of the above findings a diagnosis of Incomplete Kawasaki disease was made and the child was given Intravenous immunoglobulin two doses and along with it aspirin was also started. The child improved and was discharged on aspirin for six weeks with the advice of a regular follow up.

Discussion

The etiology of Kawasaki disease is not known but evidence supports some infectious agent. The frequency is more in Asian children with a sex ratio of 1.5:1 males to females and the mortality rate is 0.1-2%.[1,2] The figures reported in Japan are almost 8-10%.[3] If untreated 25% of patients develop coronary artery aneurysm. Incomplete Kawasaki disease should be suspected in all children with fever for more than five days associated with two or three of the features of Kawasaki disease.[4] Kawasaki disease typically has a triphasic course. The acute phase has fever, conjunctivitis, oral changes, lymphadenopathy and rash and lasts for almost one to two weeks. The sub acute phase is characterized by desquamation of the hands and feet and conjunctivitis may also persist during this phase. The

convalescent phase starts when all the clinical signs have resolved and ends when the laboratory abnormalities have returned to normal usually in four to six weeks after the onset of disease.[5]

The two most commonly missing symptoms in atypical cases are cervical lymphadenopathy and polymorphous rash while mucosal changes are nearly always found.[6] These symptoms were also missing in our patient, although our patient had a history of conjunctivitis in the beginning of the illness. Some of the other clinical features which may be present are aseptic meningitis, facial palsy, pleural effusion, acute renal failure, hepatitis, pancreatitis and jaundice. Mucous membrane changes are most common, occurring in 90% of typical and incomplete Kawasaki disease. The laboratory findings in incomplete cases are the same as the typical ones.[7] Early recognition of these cases is important as these children may suffer from myocardial infarction or unexplained death years later and some cases have actually been diagnosed at autopsy. The principal cause of death in Kawasaki disease is due to myocardial infarction.[8]

Recovery is usually complete in children who do not have detectable coronary vasculitis and second attacks occur only rarely. Intravenous immune globulin is known to be safe and its early use in patients with suspected incomplete Kawasaki disease is appropriate.[9] The frequency of coronary artery abnormalities can be reduced from 25% to less than 5% by early treatment with intravenous immune globulin.

Conclusion

A diagnosis of incomplete Kawasaki disease should be considered in all febrile children in whom some but not all features of Kawasaki disease are present. It has been recommended that in children less than six months of age with fever greater than 7 days and children older than 6 months with fever of more than 5 days should be investigated for Kawasaki disease. Early recognition and treatment of Kawasaki disease can reduce the development of potentially life threatening coronary abnormalities, and institute prompt treatment.

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