Kawasaki Disease – A Case Report

Case Report

An 11 month-old male child presented with moderate to high grade fever (for 14 days) and cough (for 7 days) was admitted in the month of April 2006. Before admission to the hospital the child was under the treatment of a private physician with no clinical improvement. On examination the child had - conjunctival congestion, strawberry tongue, erythematous rash over the palms and soles, generalized lymphadenopathy (largest cervical gland - 1.5 x 1.5 cm) and tachycardia. A clinical diagnosis of Kawasaki disease was considered.

On admission, his WBC count was moderately increased (12000/cumm), Hemoglobin was low (7.5 gm%), Platelet count was mildly increased (4,80, 000/cumm), which subsequently gradually increased to 6,75,000/cumm in a span of 5 days. His ESR at onset was 38 mm at the end of 1 hour and increased to 57 mm at the end of 1 hour by day 5. His CRP was positive. His 2D Echo was suggestive of dilatation of coronary arteries with right coronary artery of 2.9 mm, left coronary artery of 3 mm. His left anterior descending artery showed a giant aneurysm (internal diameter of 8 mm) with an area of severe narrowing.

He was immediately started on a high dose aspirin and was also given IVIg (2gm/Kg). On the second day of admission, he became afebrile. However, his platelet count increased to 9,88,000/cumm and ESR increased to 100mm. He was also given dipyridamole for the thrombocytosis to prevent thrombus formation.

On Day 5 of admission he again became febrile. He was again treated with IVIg (2gm/Kg). Aspirin in high doses was continued.

On the 7^{th} day of admission, the child became afibrile, however, his ESR remained high (100mm). There was a slight reduction in the platelet count (8,50,000/cumm).

On the 9th day of admission, in the night (at about 11pm), during sleep, the child suddenly started crying and developed vomiting, dyspnea, cardiovascular collapse and shock. The child could not recover from shock and expired at about 11.30 pm. The cause of death was severe myocardial infarction. Thus, we wish to highlight the treatment failure of aspirin and IVIg in a child with Kawasaki disease who had already developed complications.

Discussion

This disease was first described in 1967 by Kawasaki in Japanese children (Kawasaki, 1967). This disease is also known as mucocutaneous lymph node syndrome or infantile polyarteritis. It is a leading cause of acquired heart disease among children living in developed countries.

In India, Kawasaki disease has not been reported as frequently as it should be, only 25 cases have been reported so far (Apurba G, et al 2000). It is now known that coronary artery abnormalities (dilatation, aneurysms etc.) develop in approximately 20 to 25% of children with untreated Kawasaki disease (Takahashi, 1995; Kato et al , 1982; Newburger et al., 1986). This disease has been reported throughout the world (Taubert, 1997).

Kawasaki disease occurs more often in boys than in girls (ratio of about 1.5 to 1). Approximately 80% of affected children are less than five years old and fewer than 2 % of children have recurrences (Mason et al., 1993). The cause of this disease remains unknown, but bacterial toxins, viruses, and autoimmunity have been postulated in its etiogenesis (Shulman et al., 1995; Leung et al., 1993; Deresiewicz et al., 1996; Terai et al., 1995). This disease occurs year-round, but a greater number of cases are reported in the winter and spring (Shulman et al., 1987; Rauch, 1987; Khan et al., 1995).

This disease is characterized by fever (lasting for at least 5 days), bilateral nonpurulent conjunctival injection, strawberry tongue, erythema of hands or feet with desquamation usually beginning periungually, polymorphous

truncal rash, and cervical lymphadenopathy. Transient arthritis may also occur. Other manifestations may include diarrhea, vomiting, abdominal pain, hydrops of gall bladder, myositis, ulcerative stomatitis, aseptic meningitis, cranial or peripheral nerve palsies, and hepatosplenomegaly. Cardiac involvement is the most important manifestation of Kawasaki disease (Dajani et al., 1993; Taubert and Dajani, 1996). Between 10-40% of untreated children have evidence of coronary vasculitis within 2 weeks of illness as seen by coronary dilatation or aneurysm on 2D Echo that may lead to myocarditis and on long term early myocardial infarction.

Laboratory investigations show leucocytosis, thrombocytosis (which may be marked), elevated C-reactive protein, mild to moderate anemia, and high ESR. Hypoalbuminemia, and elevated serum immunoglobulin E levels may also be observed. Proteinuria and sterile pyuria of urethral origin are also common.

Current recommendations for initial therapy (preferably given within the first 10 days of the onset of illness) include the intravenous administration of immunoglobulin (IVIg) to help prevent coronary artery abnormalities and the oral administration of acetylsalicylic acid (aspirin) in high dosages to hasten resolution of the acute manifestations of Kawasaki disease, especially fever (Dajani et al, 1993). In the acute phase IVIg is given as a single dose (2 gm/Kg) over 10 - 12 hours along with high dose aspirin (80 - 100 mg/Kg/day) in four equally divided doses until the patient has been afebrile for several days. Low dose aspirin (3-5 mg/kg/day) orally is advocated for subacute and convalescent phases for 6-8 weeks after the active disease subsides for its anti-thrombotic effects and until coronary lesions resolve.

For patients unresponsive to initial treatment with IVIg, a re-treatment with second dose of IVIg is recommended. It is found that patients with low Hb (Hb<10gm/dl), leucocytosis, and low albumin are at risk of failure of initial IVIg and may require re-treatment with a second dose of IVIg (Dajani et al., 1993). The same thing happened with the present case.

It is observed that initial therapy should be started as early as possible (preferably within 10 days of the onset of illness) to prevent treatment failure and development of complications (Dajani et al., 1993). In the present case, the initial therapy was delayed because of the child was admitted to the hospital late (on about 14th day of illness) with complications already developed and this led to the failure of the treatment.

In some patients, coronary artery obstruction may be severe enough to warrant surgical revascularization (Kitamura et al., 1994). Worldwide, at least 13 patients with Kawasaki disease have undergone cardiac transplantation because of severe ischemic disease (Checchia et al., 1997).

Dr. Mansoor Quli Khan
MBBS, MD, PhD (USA)
Visiting Consultant Haemato-oncologist,
Hosmat Hospital (and other hospitals),
Bangalore, India.
Faculty Member, Akamai University, USA

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