Kawasaki Disease in an Infant – Missed Clues

Patients who do not fulfill the criteria of classic Kawasaki disease are diagnosed as ‘incomplete’ Kawasaki disease, a diagnosis that is often based on echocardiographic findings of coronary artery abnormalities [1]. Incomplete Kawasaki disease is more commonly diagnosed in young infants than older children [2]. We discuss the case of an infant who was diagnosed with Kawasaki disease in later part of his clinical presentation.

A 40-day-old boy was referred from a peripheral hospital in view of persistent fever for two weeks, despite intravenous antibiotics. At admission, he was febrile and tachypneic. Examination revealed hepatosplenomegaly without any skin changes. Blood investigations showed hemoglobin 9.6 g/dL, leucocyte count 44.6×10^9/L, platelet count 3.59×10^9/L, and raised C-reactive protein (20mg/dL). Cerebrospinal fluid examination revealed 90 lymphocytes with normal biochemistry. He was started on intravenous meropenem and vancomycin awaiting blood culture report; which was later sterile. The child was accepting breast feeds over next 2 days but fever persisted. On day 7 of admission, he developed features of congestive heart failure (CHF) and was detected to be anemic (hemoglobin 6.2 g/dL). He recovered partly with blood transfusion but remained febrile. Two days later, he developed features of cardiogenic shock and was detected to have a systolic murmur at apex. Repeat blood counts were similar and CRP was still raised. Electrocardiogram showed sinus tachycardia. He was started on vasopressor and ventilatory support. In view of persistent fever with acute cardiac event and elevated markers of inflammation, a possibility of Incomplete Kawasaki disease was suspected. A bedside echocardiography revealed dilated coronaries with mitral regurgitation suggestive of Kawasaki disease. The baby was started on intravenous immunoglobulins but died in next 24 hours due to refractory shock.

Incomplete Kawasaki disease should be considered in children with unexplained fever for more than 7 days duration with laboratory evidence of systemic inflammation even in the absence of principal features [1]. The diagnosis of incomplete KD in febrile infants with rash and cerebrospinal pleocytosis is challenging as their presentation closely mimics viral meningitis [3]. Acute congestive heart failure is an extremely uncommon early manifestation of Kawasaki disease [4]. The first manifestation of CHF which was attributed to anemia, along with persistent fever, were perhaps the early clues.

Currently echocardiography is recommended in all cases with unexplained persistent fever and supplemental laboratory criteria [1]. However, echocardiography is not often ordered in an infant with unexplained persistent fever in settings where infections are common. Possibly, an early echocardiography in our case could have detected coronary changes. The delay in starting treatment accounted for poor response of immunoglobulins, which has also been reported earlier in incomplete Kawasaki disease [1,4].

We conclude that high index of suspicion for Kawasaki disease should be kept in all infants and young children who present with persistent fever with/without supporting features such as rash or lymphadenopathy. Timely diagnosis and treatment may alter the course of disease, and may prove lifesaving.

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REFERENCES