Incomplete Kawasaki disease with coronary aneurysms presenting as neonatal sepsis in a 45 day old infant

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Introduction

Kawasaki disease was initially thought to be a disease confined to the Japanese population and was first reported by Tomisaku Kawasaki in 1967 [1]. We described the first case of Kawasaki disease in Sri Lanka in 1985 and subsequently the same authors described the first report of Kawasaki disease with coronary involvement in a Sri Lankan child in 1993 [2, 3].

Kawasaki disease is an acute febrile disease and a common form of vasculitis in childhood, seen particularly in East Asia. It is commonly seen among children in the age group three months to five years. It is extremely rare below the age of three months. The reported cases were usually incomplete and atypical variants and were associated with poor outcome probably because of delayed diagnosis [4-6]. Previous reports indicate that younger the child, the more atypical the features [7]. Kawasaki described specific criteria for the diagnosis, however, it is now known that these criteria may differ and may not fulfill the initially described criteria [1,8]. The youngest reported case in Japan did not have major features except a maculopapular rash at onset [6]. There is another report of a child with Kawasaki disease diagnosed at the age of twelve weeks [9]. Our patient is the third youngest to be reported in world literature.

Case report

A 45 day old previously well baby girl presented to a local hospital with documented high grade intermittent fever of 102° F for three days and raised inflammatory markers. Mother recalled a generalized transient erythematous rash on day three of illness which lasted for two days. She did not have any respiratory, gastro intestinal or urinary symptoms. Examination at the local hospital showed a lethargic baby with generalized erythematous rash, with otherwise unremarkable findings. She was the second child of healthy non-consanguineous parents. There were no perinatal complications and there were no risk factors for sepsis. She had a tiny ostium secundum ASD as shown in the initial echocardiograph done at the local hospital. She did not have any other cardiac abnormalities. Having considered the clinical presentation, the baby had been treated for bacterial meningitis with intravenous cefotaxime 50 mg/kg/six hourly at the local hospital on day 48 of life. Since there had been no response, the baby was transferred on day 7 of illness to Lady Ridgeway Hospital for Children in Colombo (LRH). On admission to LRH, the child remained febrile, but was otherwise well with unremarkable clinical examination findings. The rash which had been there at the onset was not seen. There were no skin changes or oedema in the extremities, no mucosal changes, no evidence of painful movement of limbs or lymphadenopathy. The fever spikes continued without resolution in spite of parenteral antibiotics. The septic screen was negative. Due to poor response to intravenous cefotaxime and persistently high CRP intravenous antibiotics were changed to vancomycin and ciprofloxacin on day 55 of life.

Investigations showed sterile pyuria and persistently high inflammatory markers. Urine, blood and cerebrospinal fluid cultures were sterile. Ultrasound scan of the abdomen was normal. Presence of pyuria, clinical features of sepsis and high inflammatory markers suggested urinary tract infection. Urine cultures were negative probably because the child was treated with parenteral antibiotics at the local hospital. An alternative diagnosis was considered with the appearance of thrombocytosis $(603 \times 10^9 / \mu l)$ on day 14 of illness. Repeated blood counts showed a platelet count of $908 \times 10^9/\mu l$ on day 15 of illness. Atypical Kawasaki disease was clinically suspected and echocardiogram was done. Table 1 shows the pattern of C-reactive protein (CRP) and platelet count with progress of acute presentation.

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Case report

2D Echo on day 17 of illness showed an aneurysm of the left main coronary artery measuring 3.4 mm, thin rim of pericardial effusion and grade II mitral regurgitation. The baby was administered intravenous immunoglobulin 2 g/kg and high dose aspirin (80 mg/kg/day) orally in four divided doses following the diagnosis of atypical Kawasaki disease. The spiking fever settled by crisis within 24 hours of initiating treatment. The antiplatelet dose of aspirin (5 mg/kg/day) was commenced after 14 days of high dose therapy and was continued for five months until echocardiographic improvement.

Repeated echocardiogram one week after the initial investigation showed a persisting aneurysm in the same location measuring 3.2 mm. Follow up investigation after three months showed a resolving aneurysm measuring 2.8 mm. 2D Echocardiogram five months after the initial presentation showed a resolved aneurysm in the same location.

Discussion

Young infants, especially neonates are more likely to present with incomplete Kawasaki disease due to atypical presentations and other differential diagnoses [10]. Final outcome is poor due to delayed diagnosis and treatment with immunoglobulin [10]. The diagnosis of incomplete Kawasaki disease might be made in cases with fever lasting five or more days and classical diagnostic criteria with several compatible clinical, echocardiographic, or laboratory findings and exclusion of other febrile illness [11, 12]. Diagnosis of atypical Kawasaki disease is usually reserved for those patients who have one or more atypical clinical manifestations such as renal impairment, testicular swelling, unilateral peripheral facial nerve palsy, diarrhea, vomiting, abdominal pain, pulmonary nodules and/or infiltrates, pleural effusions, acute surgical abdomen and hemophagocytic syndrome which are not seen in Kawasaki disease [8].

This child was only 45 days old and initially had clinical and laboratory features supportive of severe sepsis. Apart from the transient erythematous rash and fever the child did not have any other features of classical Kawasaki disease. Ultrasound scan of gall bladder was normal. These factors lead to the initial diagnosis of sepsis. The rising platelet count was the main indicator to prompt an echocardiogram which then confirmed the diagnosis. The diagnosis of incomplete Kawasaki disease in early infancy has been associated with higher mortality most likely due to coronary artery involvement and delayed treatment [13]. Mitral regurgitation detected in this child is also a known acute complication of Kawasaki disease seen in approximately 15% of children with cardiac manifestations [14].

The American Heart Association (AHA) recommends that echocardiography should be considered in any infant aged <6 months with fever of ≥7 days’ duration, laboratory evidence of systemic inflammation, and no other explanation for the febrile illness since young infants may present with fever and have few, if any, classical diagnostic criteria [8]. Thus we would like to emphasize the possibility of Kawasaki disease to be considered in any ill neonate or young infant with persistent fever not responding to treatment. These children should be reassessed for incomplete Kawasaki disease according to AHA recommendations [8]. The diagnosis based on high platelet count would be too late for the prevention of coronary lesions although it may prevent further deterioration.

Conflicts of interest

There are no conflicts of interest.

References

Case report


