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Case report

An atypical presentation of Kawasaki disease with thrombocytosis at the onset of the illness

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Summary

Kawasaki disease (KD) is a systemic acute vasculitis, characterized by fever, bilateral non-exudative conjunctivitis, redness of the tongue, lips and oral mucosa, changes in the extremities, cervical lymph node, and polymorphic exanthema. Administration of IVIG within the first 10days after onset of fever in combination with high dose aspirin reduces the risk of coronary artery damage in KD. There is currently no laboratory test for diagnosing KD. Rather, diagnosis is performed with reference to established clinical criteria. Unfortunately, atypical manifestations of KD appear to be on the rise decreasing the likelihood of timely diagnosis and appropriate treatment. Herein, we report an unusual case of KD: a 6-year-old boy presented with fever with desquamation of the fingers and thrombocytosis.

Background

The first case was described by Tomisaku Kawasaki in 1961[1]. Kawasaki disease (KD) is commonly seen in children below the age 5 years, frequently in association with coronary artery aneurysms [2]. The aetiology is unknown. Though suspected to have an infectious aetiology, this has not been proven[3]. Diagnostic criteria for KD include fever and at least 4 of the 5 following symptoms[1]:

- Bilateral bulbar conjunctival injections without suppuration
- Changes in the oropharyngeal mucosa
- Changes of the peripheral extremities
- Polymorphous rash
- Cervical lymphadenopathy without suppuration

Thrombocytosis is commonly seen after the first week of the illness and may be marked[4]. Incomplete KD should be considered in a child with unexplained fever exceeding 5 days, associated with 2 or 3 of the above symptoms. Diagnosis of incomplete KD is based on echocardiographic findings[1]

A six-year-old previously healthy boy presented with a history of high fever for 3 days duration which was documented as 102°F at home. Fever was not associated with any systemic symptoms except for irritability. He was admitted to a local hospital and transferred to Teaching Hospital on day 4 of illness for further management. His white blood cell (WBC) count was 12,890/cu mm (Neutrophils 77%, lymphocytes 21%), haemoglobin (Hb) level 13.8 g/dl, platelet count 243000/ cu mm at the local hospital for which he was commenced on intravenous (IV) penicillin. On arrival in our hospital, he had high fever with peeling of skin in both hands.

Examination revealed an adequately grown child with mild irritability with desquamation of skin in both hands with involvement of the palms without peripheral oedema. He had a 'strawberry tongue' but no cervical lymph node enlargement and no abnormalities found on conjunctivae. Cardiovascular, respiratory and abdominal examination revealed no abnormalities and his central nervous system examination was normal except for irritability. His WBC was 16,500/cu mm (Neutrophils 86%, lymphocytes 11%), Hb 12.8 g/dl, platelet count 503,000/ cu mm, C-reactive protein (CRP) 75mg/dl and erythrocyte sedimentation rate (ESR) 62 mm in 1st hour on day 4 of illness. He was started on IV cefuroxime because of the possibility of sepsis and fever subsided on 3rd day of hospital admission. A dermatologist who was consulted thought that the peeling may be due to allergy. He was discharged on oral cefuroxime for 5 days.

Five days later, he presented with high grade fever of one day duration with persistence of the peeling of skin. On examination, he had desquamation of the skin in both hands, palms, forearms and face, strawberry tongue and non-tender multiple cervical lymph node enlargement. Figure 1 shows peeling of the skin of the palm and Figure 2 shows the resolving strawberry

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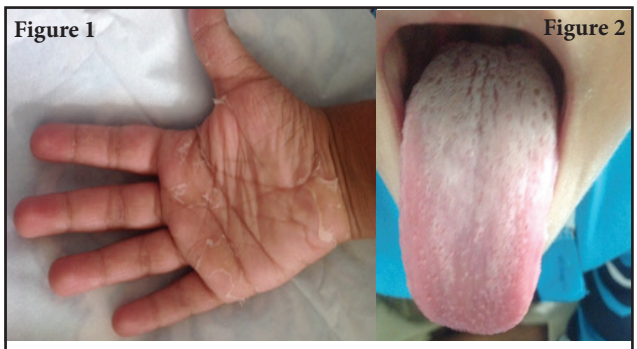

Figure 1 : Palm showing desquamation, Figure 2 : Resolving strawberry tongue

WBC was 20,410/cu mm (Neutrophils 81%, lymphocytes 8%), Hb 12.2 g/dl, platelet count 522,000/cu mm, CRP 75mg/L, ESR 106 mm in 1st hour, standard agglutination test (SAT) negative for salmonella infection, urine full report (UFR) normal, alanine transaminase (ALT) 15 U/L and aspartate transaminase (AST) 19 U/L. The blood and urine cultures were sterile and the blood film revealed features of viral infection and reactive thrombocytosis. Chest X-ray was normal with normal size heart. He was commenced on IV cefotaxime and the fever did not respond. KD was suspected and a 2D echo study was performed which revealed a mild focal dilatation of 3 mm in the left coronary artery with normal right coronary artery and cardiac function. He was given IV immunoglobulin 2g/kg and an anti-inflammatory dose of aspirin on 6th day of second admission. Fever subsided and he was given the anti-platelet dose of aspirin. Repeated 2D echo showed 2.2 mm dilatation of left coronary artery with normal right coronary artery and cardiac function. Differential diagnoses were sepsis, Staphylococcal scaled skin syndrome (SSSS) and Kawasaki disease.

Treatment

He was given IV immunoglobulin (IVIG) 2g/kg and an anti-inflammatory dose of aspirin on 6th day of second admission. Fever subsided and he was given the anti-platelet dose of aspirin for another 6 weeks.

Outcome and Follow-Up

He has made a complete recovery with the treatment and he was fever free on following day of IVIG treatment. Repeated 2D echo in 6-week time showed 2.2 mm dilatation of left coronary artery with normal right coronary artery and cardiac function. He was reviewed after 6 months of illness and he had normal examination findings including 2D echocardiogram.

Discussion

KD is the most common cause of acquired heart disease in children in developed countries[5,6]. Whilst KD is not so common in children under 3 months, or over 5 years, they are at greater risk for coronary artery aneurysms[1,5]. KD has an acute, a subacute and a convalescent phase. In the sub-acute phase, lasting from around 2-4 weeks, patients may have desquamation of the fingers and toes, arthritis and thrombocytosis[7]. In our case thrombocytosis was observed on the fourth day of illness. The pathogenesis of reactive thrombocytosis is unknown[8]. However, it is believed that thrombopoietin and IL6 contribute to thrombocytosis in Kawasaki disease[8]. For our best knowledge, there are no similar published cases of Kawasaki disease presented with thrombocytosis at the onset of illness though there are enough case reports with atypical presentation.

Learning points

- Kawasaki disease is the most common cause of acquired heart disease in children in developed countries
- Clinicians should be made aware of atypical cases to facilitate early diagnosis and treatment.
- Absence of sensitive diagnostic testing remains a major obstacle to correctly identify patients with Kawasaki disease.

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