

Case report:

A 3 Month Old Child with Atypical Kawasaki Disease

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Abstract:

Background: Kawasaki Disease (KD) is an autoimmune vasculitis of unknown etiology that occurs predominantly in infants and young children with special predilection to coronary arteries. **Case Presentation:** We report the case of a 3-month-old child with an atypical form of KD, characterized by prolonged fever and non-specific symptoms, who developed an aneurysm of the left anterior descending coronary artery. **Conclusion:** This case report underlines the challenges in recognizing atypical forms of the illness in young infants with minor features, who are at higher risk of long term cardiac complications.

Keywords: Kawasaki disease; Fever; Coronary aneurysm; Thrombocytosis; Case report

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Introduction: Kawasaki disease (KD) or infantile polyarteritis nodosa, is an acute febrile illness of childhood associated with medium vessel vasculitis.¹ After Henoch Schonlein Purpura, it is the second most prevalent cause of vasculitis in children and leading cause of pediatric acquired heart disease.²⁻³ About 85% of children having Kawasaki disease belong to the age group of less than five years and the peak age is from 18-24 months. The disease can also present before 6 months or after 5 years of age but the incidence is less and these affected children are at increased risk of coronary artery aneurysm with long term sequale.^{2, 4-6} Regarding the cause of Kawasaki disease, it is still not well explained, but there are some genetic and environmental factors (viral and bacterial infection) which have an impact in the pathogenesis.¹ Currently no specific diagnostic test is available for confirming KD. So diagnosis is done based on the combination of clinical criteria and laboratory findings. Presence of prolonged

fever (≥ 5 days) along with at least four out of five principal clinical features (change in extremities, polymorphous exanthema, bilateral bulbar conjunctival injection without exudate, changes in lips and oral cavity, cervical lymphadenopathy) are diagnostic for KD.⁷ Supportive laboratory evidence are anaemia, neutrophilic leukocytosis, thrombocytosis, positive C-reactive protein, raised erythrocyte sedimentation rate (ESR), elevated serum transaminase level and hypoalbuminemia etc.⁸ Treatment in Kawasaki disease is mainly targeted to decrease the inflammation of coronary arterial wall and to inhibit platelet activation. The American Heart Association (AHA) recommends standard treatment with Intravenous Immunoglobulin (IVIG) and aspirin in classic and complete KD cases whereas in resistant cases use of methylprednisolone and other immunosuppressive agents such as cyclosporins, cyclophosphamides are advocated.⁹

Atypical Kawasaki is one of the variant of KD which

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usually present before 6 months of age and may lack some of the eye and oral mucosal changes.⁹⁻¹⁰ As affected infants don't present with all classic features, diagnosis may be delayed in these cases.¹¹ So timely diagnosis of this special group remains challenging.

Here we report an unusual case of KD: a 3 month old baby girl who presented with features of acute gastroenteritis which is a common outdoor problem in our daily practice.

Case Presentation: A full term, outborn, previously healthy 3 month old female baby presented to our Emergency department with history of high grade intermittent fever for 10 days along with cough for 5 days. Two days before admission she also had vomiting for 5 times and loose, mucoid stool for several times. She was getting oral antibiotic prescribed by a local doctor. Baby had poor feeding, irritability and less urine output. On physical examination, she was sick looking, afebrile (Temperature 98.6°F), euglycemic and there was no sign of dehydration (capillary refill time <3 sec). Her other vital signs included heart rate of 132/min and respiratory rate of 36/min. Physical examination of heart and abdomen was normal. Blood tests showed Hb-8.9 g/dl, total WBC was high 21600/cmm with thrombocytosis (platelet- 610000/ cmm). CRP was also high 167.4 mg/L. Routine examination of stool revealed plenty of pus cell. It was clearly a case of sepsis with gastroenteritis. We started broad spectrum antibiotics along with other supportive measures. Temperature gradually lowered down but did not touch base line. To exclude other septic focus we did widal test, urine routine and culture, Ultrasonography of abdomen but all reports were normal. With treatment, frequency of loose stool and vomiting decreased, CRP and total white cell count also decreased. But after 3 days of slow improvement with 100°F temperature once daily, child became toxic with increased peak of fever (102°F). We thought about a new infective episode and repeated septic work up. There was mild anaemia, leucocytosis (25900/cmm) with thrombocytosis (743100/cmm). ESR was high; 60 mm in 1st hour though C-reactive protein was low than before (26 mg/L). Serial blood count revealed progressive increase in platelet count (868100/cmm → 916100/cmm). It struck our mind and we did an Echocardiogram which revealed mildly

dilated coronary arteries and aneurysmal dilatation of the left anterior descending coronary artery and findings were suggestive of KD. Without any delay we counseled parents about the disease pattern and started intravenous immunoglobulin (IVIG) 2 gm/kg on 6th day of hospitalization. After 48 hours fever subsided with subsequent improvement in general wellbeing, appetite and laboratory parameters. The child was on maintenance dose of aspirin. But repeat Echo after 2 weeks showed similar findings. So she got re-admitted and received IVIG followed by inj. Methyl prednisolone. Patient improved gradually and still in follow up.

Discussion: Kawasaki Disease is an autoimmune vasculitis of unknown etiology that occurs predominantly in children less than 5 years of age.¹² Clinical criteria and laboratory findings are used to diagnose KD as no clear diagnostic test is available. Patients who fulfill the principle clinical findings according to case definition, are termed as classic or typical KD. But all patients do not have symptoms that meet the classic diagnostic criteria of KD.¹³

Atypical KD is mostly seen in young infants less than 6 months of age, who are unfortunately at the greatest risk of developing coronary disease. These patients may have only prolonged fever as a sole clinical feature or sometimes associated with subtle clinical signs along with fever. Presence of certain lab criteria may raise the clinical suspicion of KD. Identification of such cases can be quite challenging, and fatal outcomes have been reported¹⁴.

In our case KD diagnosis was extremely difficult as no classical manifestation was present except one criterion that is prolonged fever and features of gastroenteritis. But in atypical KD, only prolonged fever may be present with no eye and oral mucosal change that we didn't suspect initially. Vomiting, diarrhea, abdominal pain, hepatitis are common gastrointestinal findings which may be found in these cases.⁹ Our patient's initial lab parameters were anaemia, leucocytosis, raised ESR and thrombocytosis which can be present in any inflammatory condition. Raised C- reactive protein and plenty of pus cell in stool were also supportive for sepsis. At that point, features were more in favor of gastroenteritis and we had no basis to perform

ECHO and to start IVIG because fever peak was decreasing with slow clinical improvement. But the second peak of fever which was non-responsive to broad spectrum antibiotics and persistent laboratory parameters guided us to do Echocardiogram.

As soon as we confirmed KD, we started treatment with IVIG. Intravenous Immunoglobulin is the mainstay of therapy in KD. How IVIG exactly works in KD is not clearly understood. Some possible mechanisms are modulation of cytokine production, anti-inflammatory effects and regulation of T-cell activity.¹⁵ Timely initiation of IVIG can reduce the incidence of coronary artery aneurysm from 25% to 4%.⁹

Our patient already presented after 10 days of illness and we had a delay in diagnosis. So resistance to treatment was expected for which 2nd dose of IVIG and methyl-prednisolone were also

required and patient responded. Steroids have a promising effect in IVIG resistant cases evidenced by many clinical trials.¹⁶⁻¹⁷

So KD should be considered in the differential diagnosis of prolonged fever in infants. Many pediatricians who are experienced in the diagnosis and treatment of KD at different medical centers, have got patients in whom prolonged fever was virtually the only manifestation of KD¹⁸. Age of the child is another important factor which will help us to assess the severity of the disease.

Conclusion:

Atypical Kawasaki Disease should be considered particularly in younger children less than 6 months old, presenting with prolonged fever, with non-specific clinical features and if empiric antibiotics have proven ineffective.



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