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An infant with persistent high-grade fever: Having a low threshold for suspecting and treating atypical Kawasaki disease

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Summary:

We report a previously healthy 8-month-old infant who presented with a 6-day history of high-grade fever. Initial clinical examination revealed no fever, but two classical features of Kawasaki disease were observed in bilateral conjunctivitis and polymorphous rash. Initial investigations revealed anaemia, and high CRP, white cells, platelets and neutrophil count. As chest x-ray showed right mid-zone consolidation, he was treated for a chest infection. However, he was still pyrexial with high inflammatory markers after a 5-day course of intravenous antibiotics. While he did not fulfill the criteria for classic Kawasaki disease, a diagnosis of atypical Kawasaki disease was considered. After discussion with Rheumatology, he was treated as atypical Kawasaki disease with IV immunoglobulin and high-dose aspirin. The case identifies how an important diagnosis can manifest differently and needs a high index of suspicion to make this diagnosis in a timely fashion to prevent serious sequelae.

Background:

Patients with atypical or incomplete Kawasaki Disease (KD) do not fulfill all of the diagnostic criteria of classic KD. For a diagnosis of atypical KD, the patient must have:

• Fever for 5 or more days.

• Two of five clinical features for typical KD (which comprises bilateral non-purulent conjunctivitis, changes in upper respiratory tract mucous membranes such as strawberry tongue, erythema/oedema/desquamation of hands and feet, polymorphous erythematous rash on trunk and limbs and cervical lymphadenopathy>15 mm).

• C-reactive protein (CRP) greater than 3.0 mg/L and/or Erythrocyte Sedimentation Rate (ESR) greater than 40 mm/h.

• Compatible laboratory findings (at least 3 of the following: albumin, ≤ 3.0 g/dL; anaemia for age; elevation of alanine aminotransferase; platelets ≥ 450,000 µL after seventh day of illness; white blood cell count ≥ 15,000 µL; urine ≥ 10 white blood cells/high-powered field).

• Positive echocardiography findings for coronary artery aneurysms/dilatation.

• No other reasonable explanation for illness.

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Since there is no singular sensitive test to confirm KD, it is challenging to establish a definitive diagnosis quickly, and failure to treat early often lead to coronary artery complications occurring in children with KD. In the developed world, KD is the biggest cause of acquired heart disease in children. The mainstay of treatment of KD remains Intravenous Immunoglobulin (IVIG) infusion within 10 days of onset of the disease. IVIG can decrease the risk of coronary artery abnormalities from 25% to 5%. A detailed history and physical examination are required to reveal the cause of other features of KD, but atypical KD should still be considered when all classical features of KD are not present.

Case Presentation:

An 8-month-old White Eastern European male was referred to Paediatrics with a 6-day history of persistent high-grade fever (39-40˚C) and 2-day history of bilateral non-purulent conjunctivitis and widespread, maculopapular, non-urticarial blanching rash. There was no cough or coryzal symptoms, or history of recent illness in the household. Two days before admission, he was started on oral amoxicillin in primary care.

The patient was born by normal vaginal delivery at 41+2 weeks gestation with a birth weight of 3.4 kg. He had remained fit and well in the past and did not take any regular medications. He was not known to have any allergies. His immunizations were up to date.

On examination, the patient was alert but irritable. He showed no symptoms or signs of respiratory pathology. His temperature was settled at 37.8˚C with Paracetamol. He had tachycardia (heart rate 150 beats per minute) and good peripheral perfusion. Anterior fontanelle was noted to be soft. He had a widespread maculopapular blanching rash on torso, arms and legs. He did not have reactivation of his BCG scar. His throat was mildly congested with pus spots on tonsils. He did not have cervical lymphadenopathy.

Investigations:

Initial Full Blood Count (FBC) revealed a haemoglobin concentration of 90 g/dL, a Mean Corpuscular Volume (MCV) of 74.3 fL and a White Cell Count (WBC) of 12.7*109 /L with 6.2*109 neutrophils, and 4.4*109 lymphocytes. The platelet count was initially 458*109/L and CRP was elevated at 101 mg/L. Liver function tests revealed a total bilirubin of 2.9 mg/dL, aspartate aminotransferase of 17 U/L, alkaline phosphatase of 148 U/L, total protein of 68 g/dL, albumin of 39 g/dL and globulin of 29 g/dL. Urinary microscopy showed normal white cell count. Urine and blood cultures did not grow pathogens. Chest x-ray showed a right mid-zone consolidation.

Echocardiography showed normal appearance of aortic arch and proximal coronary arteries with no evidence of atrioventricular valve regurgitation or pericardial effusion. Biventricular function was normal and right and left ventricular outflow tracts had unobstructed flow.

The CRP increased further during admission to 121 mg/L, while WBC count increased to 16.5*109, platelets count rose to 774*109 and ESR was 37. Fibrinogen was elevated at 7.10 g/dL, and troponin was elevated at 5.8 ug/L.

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Differential Diagnosis:

As this patient presented during the COVID-19 pandemic, Paediatric Inflammatory Multisystem Syndrome Temporally associated with SARS-CoV-2 (PIMS-TS) was ruled out based on clinical and biochemical criteria. He was covered for potential Sepsis till urine and blood cultures were reported to be negative. Further to the chest x-ray demonstrating a right mid-zone consolidation, causes of atypical pneumonia were also considered. Respiratory viral PCR sample and nasopharyngeal PCR sample were both negative for respiratory viruses. Cytomegalovirus and Epstein-Barr virus screen was negative. Antistreptolysin-O (ASOT) was below 200 ruling out recent group A streptococcus infection. Therefore, in the absence of any other plausible cause of illness, the patient was treated for Atypical KD.

Treatment:

The patient was given Intravenous Immunoglobulin Infusion (IVIG) in a dose of 2 g/kg and high-dose oral aspirin 12.5 mg/kg on his fifth day of admission and eleventh day of illness. He also completed a course of intravenous antibiotics and oral antibiotics during inpatient admission.

Outcome and Follow-up:

Following IVIG administration, the patient improved both clinically and biochemically and was not noted to be irritable. He went on to develop desquamation of his toes and fingers. He was discharged after 9 days of admission when his CRP had decreased to 22 and he had remained afebrile for 24 hours. His parents were advised to contact Paediatric Assessment Unit if he spikes fever within 48 hours of discharge. A local clinic follow up was arranged and a formal referral to tertiary Cardiology was made for urgent outpatient review.

Discussion:

While the exact cause of KD remains unclear, it is characterized by largely self-limiting systemic inflammation of medium-sized arteries. However, it constitutes a diagnostic conundrum as its principal presentation can mimic several other diseases, including measles, scarlet fever, and juvenile idiopathic arthritis. Current NICE guidelines recommend having a low threshold for considering KD especially in infants younger than 1 year, who often do not fulfill the criteria for KD at presentation and hence have delayed diagnosis and treatment, putting them at higher risk of experiencing coronary artery complications.6 Furthermore, principle features of KD, as shown in this case, may appear and disappear at different points in the clinical course of the disease and maybe subject to factors such as parental recall and clinician subjectivity. As such, it is recommended that diagnosis of KD should be considered in all cases of fever lasting 5 days or more; the presence of cardinal KD features make the diagnosis more likely but their absence should not rule out KD.

Learning points/take home messages:

- Consider KD in infants with prolonged fever, irritability and unexplained or culture-negative shock/meningitis/lymphadenitis, unresponsive to antibiotics.
- Suspicion of KD should involve early discussion with Rheumatology and Cardiology colleagues to initiate IV immunoglobulin and high-dose aspirin.
- IV immunoglobulin is most effective when given within first 10 days of illness, and reduces risk of coronary artery complications from 25% to 5%.

Biography

Roshan Bharani is working as a ST4 Paediatric trainee at Ipswich NHS Foundation Trust (NHS East Suffolk and North Essex).