



A narrative of a cohort of patients with Kawasaki disease seen at the Lagos State University Teaching Hospital

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Abstract: Kawasaki disease (KD) is an acute febrile illness affecting young children. It was first described by Tomisaku Kawasaki in Japan (1961). Also referred to as mucocutaneous lymph node syndrome because of its involvement of lymph nodes, skin, the nasal, oral and pharyngeal mucous membranes. It is characterized by prolonged fever, exanthema and a tendency to develop coronary arterial lesions leading to aneurysms and thromboembolism, a vasculitis syndrome which can lead to mortality from coronary artery aneurysm (CAA) in a small percentage of patients. The cause is unknown. Although, there is the possibility that it might be related to an infectious disease which triggers abnormalities of the immune system. There has been no evidence of the disease transmission from one person to the other. A possible role of environmental factors like toxins, dust mite has been considered as triggering factors, but not proven. It is a rare disease that mainly affects children aged less than five years old. The cases discussed were of good prognosis, in spite of the presence of aneurysmal dilatation in one of the children. Also, the possibility of missed diagnosis is very common with Kawasaki's disease because of poor awareness of the disease among health care practitioners and its resemblance to other childhood illnesses such as measles which are commoner and are vaccine preventable. These cases are reported to increase the awareness of its existence in our environment and hence, increase the index of suspicion and prompt diagnosis that will reduce morbidity and mortality in affected children.

Keywords: Kawasaki disease (KD); coronary artery dilatation; echocardiography; case series

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Introduction

Kawasaki disease (KD), being a vasculitis illness, typically presents as a syndrome with multi-systemic manifestations including a prolonged fever, irritability, eye changes like non-exudative bilateral conjunctivitis and anterior uveitis, perineal erythema, erythematous swelling of the hands and feet, oral changes like 'strawberry tongue' and lip fissuring, unilateral solitary non-suppurative cervical lymphadenopathy, myocarditis, pericarditis, sterile pyuria, hepatic, renal and gastro-intestinal dysfunction (1).

The disease is also known as mucocutaneous lymph node

syndrome in reference to involvement of lymph nodes, skin, the nasal, oral and pharyngeal mucous membranes (2). It commonly affects children less than four years old and was first described by Tomisaku Kawasaki in Japan (1961) (3).

Kawasaki disease is rare with possibility of missed diagnosis. These cases are reported to increase the awareness of its existence in our environment and hence, increase the index of suspicion and prompt diagnosis that will reduce morbidity and mortality in affected children.

We present the following case in accordance with the CARE reporting checklist (available at <http://dx.doi.org/10.21037/aoi-19-12>).

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Case presentation

Case 1

OG, a 2-year-old male toddler was referred to LASUTH on 9/5/16 following a referral from a private hospital with a history of fever of 9-day duration, swelling of hands and feet, redness of eyes and skin eruptions of 4 days duration. Fever was high grade, continuous, temporarily relieved with paracetamol. Five days later, he developed redness of both eyes associated with milky discharge but no associated itching. He also had sores in the oral mucosa which subsequently dried up but bled with little pressure especially with crying. No history of straw berry tongue. No history of bleeding from any orifice or history of recent travel or contact with corpse suspected to have died of a viral haemorrhagic fever. Nil history of contact with any one with similar problem and no history of bone pains. He developed erythematous maculopapular rashes around the perineum and medial thigh which later dried up with resultant excoriation. His hands and feet were swollen and erythematous. No history of swelling elsewhere, cough, bluish discoloration of lips and skin, urinary symptoms. His genotype was AS. In the course of the illness presented at two private hospitals where he was admitted for 3 days and 6 days respectively and was placed on intravenous fluid, ceftriaxone and artesunate but due to persistence of symptoms was referred to LASUTH. At presentation in CHER (Children Emergency Room) examination revealed an acutely ill looking child, conscious, irritable, febrile (temperature 38 °C), pale, anicteric, bilateral conjunctivitis, hyperaemic ulcerated lips, oedema of hands and feet with right cervical and submandibular lymphadenopathy. He was tachycardic, tachypneic and had hepatomegaly. A clinical diagnosis of Kawasaki disease was made.

Laboratory results revealed neutrophilic leucocytosis with a normal platelet level. A low haematocrit level of 19.2% which necessitated transfusion with packed cells. Within 72 hours of admission he had developed thrombocytosis. Echocardiography was found to be normal and blood culture showed no growth. Liver function test showed elevated AST while protein and Albumin levels were low.

He was commenced on intravenous meropenem, intravenous Immunoglobulin at 2 g/kg to run over 10 to 12 hours. He was also given oral Aspirin at 100 mg/kg/day,

later reduced to 3–5 mg/kg/day when patient was afebrile for 48 to 72 hours for a total of 6 weeks.

The following are the laboratory results PCV 24%, Hb 7.8 g/dL, WBC 22,400/Lit, neutrophils 66%, lymphocytes 24%, others 0%. On 10/5/16 ESR 90, HB 7 g/dL, WBC 17,370/Lit, neutrophils 57.7%, lymphocytes 26.5%, monocytes 14.6%, eosinophils 1.0%, basophils 0.2%, platelets 377,000/Lit. On 12/5/16 WBC 15,640/Lit, Hb 5.7 g/dL, HCT 19.2%, neutrophils 68.1%, lymphocytes 19.4%, monocytes 11.8%, eosinophils 0.4%, basophils 0.3%,

On 13/5/16 WBC 13,090/Lit, Hb 8.5 g/dL, HCT 26.5%, platelets 233,000/Lit, neutrophils 44.6%, lymphocytes 41.2%, monocytes 11.2%, eosinophils 2.8%, basophils 0.2%.

On 17/5/16 WBC 10,000/Lit, neutrophils 20.8%, lymphocytes 69.5%, monocytes 7.7%, basophils 0.0%, eosinophil 2.0%, Hb 8.3 g/dL, PCV 23.8%, platelets 519,000/Lit.

The fever persisted after the first course of the Immunoglobulin, hence a repeat dose was given after which the patient made remarkable clinical improvement and was discharged. Has been on follow up. Serial echocardiography on follow up has been normal. Repeat complete blood count has also been normal.

Case 2

A.L, a 19-month-old female toddler admitted in LASUTH on 17/06/2016 with complaints of fever of a week, redness and swelling of the extremities of 5 days and irritability of 5 days. She was referred from a private hospital where she had been on admission on account of the symptoms above. Fever was high grade, intermittent and relieved with the use of antipyretics, there was decreased appetite and irritability. Redness of the mouth was noticed about 2 days later with associated blisters formed. Redness subsequently progressed to involve the eyes, upper and lower limb and the perineum. There was also swelling of both hands and feet. Redness and swelling was observed few hours before the commencement of drugs given from the private hospital. No reduction in urine output, no past history of swelling. Genotype was AA.

Examination revealed an irritable child who was pale and febrile (38.2 °C), reddish roughened tongue (strawberry tongue), swollen feet and legs, hyperaemic bilateral non-discharging conjunctiva, ulcers on the lips and angles of the mouth, desquamation of the palm and excoriation in the groin. There was hepatomegaly which was not tender.

A diagnosis of Kawasaki disease was made. She was admitted, commenced on high dose oral aspirin at 80 mg/kg/day, intravenous immunoglobulin 1 g/kg. Echocardiography was done which revealed left coronary dilatation. Electrolyte, urea and creatinine essentially normal. Full blood count showed mild anaemia (PCV-27%) and leukocytosis (WBC-20,600 cells/Lit).

Fever subsided by the 4th day on admission and remained stable for 3 days later when mother requested for discharge. Low dose Aspirin was commenced and patient remained stable at follow up. The low dose aspirin was given for 6 months in this patient. Follow up echocardiography has been normal.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

Discussion

Several risk factors linked with increased susceptibility to Kawasaki disease have been identified and they include age less than five years, male sex and Asian or Pacific Island ancestry (particular children of Japanese and Korean descent) (2). It can however affect children of all ethnic groups (4). Tizard *et al.* reported an incidence of 134–135 per 100,000 children (5). The mortality figures documented in literature vary from 0.08% in Japan to 3.7% in the UK (5). The cause of KD is unknown, however it is not contagious. Different models and theories have associated development of the disease to infectious agents like bacteria and viruses and also to environmental factors, but these stand to be proven unequivocally. Genetic loci which increase susceptibility to KD have been identified (2).

The American Heart Association diagnostic criteria includes the presence of a fever (often high grade) for more than five days with any four of the following clinical features; changes in the peripheral extremities (reddening or oedema of the palms and soles, periungual desquamation, and Beau lines), polymorphous (but non-vesicular) rash (generalised or limited to groin or lower extremities), Oropharyngeal changes (erythema, fissuring and crusting of the lips, strawberry tongue etc.), Bilateral, non-exudative, painless bulbar conjunctival injection, non-purulent cervical lymphadenopathy (unilateral) (1,2).

Kawasaki disease follows a clinical course namely; acute, sub-acute and convalescent (3). The first phase consists symptoms and signs such as high grade fever, conjunctivitis (non-suppurative), skin rash in the trunk and genital areas, strawberry tongue, swelling and redness of the palms and soles, unilateral cervical lymphadenopathy and irritability (2). In the second phase, the child develops desquamation of the skin of hands and feet, particularly in the periungual areas. Joint pains, diarrhoea, vomiting and abdominal pains are other symptoms noted (2). In the third phase, the resolution of the signs and symptoms are noted except if complications develops (2). KD is a cause of acquired heart disease and cardiac complications include vasculitic changes of the coronary arteries, myocarditis and valvular damage (2). Tizard reported the occurrence of coronary artery aneurysm (CAA) in 25% of children with untreated KD, a small number develop coronary thrombosis, myocardial infarction and death. Most aneurysms develop 6–8 weeks from the onset of the disease (5). A study done over a 9-year period (1980–1988) in the United Kingdom revealed that coronary arterial lesions were present in 28% and persisted in 23% of children (5).

At least three successive echocardiography examinations should be carried out (at time of diagnosis of KD, at two and then at six weeks after onset) to screen for the presence of CAA. There is no specific diagnostic laboratory investigation for KD, although most patients have elevated serum levels of the acute phase reactants (erythrocyte sedimentation (ESR), C-reactive protein (CRP), etc.). Urine protein products Meprin A and Filamin C have been demonstrated to be more useful than other acute phase reactants like ESR and CRP as biomarkers in the diagnosis of KD (1).

The administration of Intravenous gamma-globulin (IVIG) in the first ten days of the onset of the illness has been shown to remarkably reduce the incidence of CAA from up to 25% in untreated KD as mentioned earlier to about 2%, in keeping with results from a recent Cochrane review assessing IVIG use in KD (4,5).

Conclusions

Kawasaki disease in the cases discussed above which fulfilled the criteria for diagnosis may be rare in our environment, but the presentation of the two toddlers discussed above

presenting in an interval of five weeks is a reason to raise awareness about its occurrence in the Tropics.

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Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at <http://dx.doi.org/10.21037/aoi-19-12>

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at <http://dx.doi.org/10.21037/aoi-19-12>). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

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