

Kawasaki disease in Port Harcourt, Nigeria

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Abstract

Kawasaki disease (KD) is an acute febrile vasculitis of childhood, predominantly affecting medium-sized arteries with a predilection for coronary arteries and commonly occurring in children under the age of 5 years. Early recognition can be challenging; however, delayed diagnosis increases the risk of coronary artery abnormalities and death. We report a case of KD in a 19-month-old child who presented with prolonged fever, conjunctival congestion, skin rash and redness of lips and tongue. He was initially managed as a case of complicated measles, but with worsening symptoms, KD was suspected. Echocardiography revealed a left coronary artery aneurysm. He received two doses of intravenous immunoglobulin G, over a period of 24 h and responded remarkably to treatment. KD is rare in Port Harcourt, Nigeria, and children with prolonged fever and erythematous rash may be mistakenly managed for measles if there is no high level of suspicion amongst clinicians.

Keywords: Coronary artery aneurysm, immunoglobulin, Kawasaki disease, measles, prolonged fever, rash

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INTRODUCTION

Kawasaki disease (KD) is an acute self-limiting, febrile illness of childhood accompanied by vasculitis of medium-sized arteries.¹ Although the most common cause of acquired heart disease in developed countries,^{2,4} it is rare in developing countries.^{5,6} Some cases have been reported in Nigeria^{7,8} but none from Rivers State.

The disease presents as complete or incomplete KD. Echocardiography may reveal coronary artery aneurysm (CAA).¹⁻³ Making a diagnosis of KD in a developing country can be quite challenging as common febrile illnesses such as measles may have similar symptomatology.⁹⁻¹³

We, therefore, report this case of a 19-month-old male who met the KD diagnostic criteria.

CASE REPORT

A 19-month-old male, OP, presented with a 3-week history of fever, redness of the eyes and mouth of 18 days, rash and excoriation of the skin of 2-week duration, vomiting and passage of loose stools of 5 days. At onset of illness, he was taken to a health facility where a diagnosis of measles and malaria was made, and he received paracetamol, amoxicillin, calamine lotion, Vitamin A, oral rehydration therapy and antimalarial. With the persistence of symptoms, measles complicated by sepsis was suspected, he was admitted and intravenous antibiotics and fluid added to his treatment. With no clinical improvement, the managing medical

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team asked for a paediatrician to review by the 3rd week of the illness. His pregnancy was normal, and mother received only routine drugs. He received all immunisation including measles and Bacillus Calmette–Guerin (BCG) vaccines in infancy, according to the National Programme on Immunization in Nigeria.

Clinical examinations revealed an obese child (weight of 21 kg and height of 92.5 cm), febrile (temperature 38.7°C), mildly pale with angular cheilitis, bilateral conjunctival injection, non-pitting bilateral pedal and hand oedema [Figures 1 and 2]. The lips, oral cavity, tongue and tonsils were erythematous. There was generalised desquamation of the skin in sheets [Figures 2-5], sparing the upper abdomen and trunk. The skin of the perineum was erythematous and weepy. He also had periungual desquamation of the nails and was irritable. The knees were warm, tender and the right knee measured 1.5 cm more than the left (at the level of the mid-patella) and he

was limping. His lymph nodes were not enlarged; only the first and second heart sounds and vesicular breath sounds were heard on auscultation.

Investigation results showed some deranged laboratory indices compared to the normal for age (adapted for facility use).¹⁴ He had mild anaemia with haemoglobin of 10.4 g/dl (normal: 12–18 g/dL), leucocytosis of $13.7 \times 10^9/L$ (normal: $4.8\text{--}10.8 \times 10^9/L$), neutrophilia of 76% (normal: 54–62%) and normal platelet count of $237 \times 10^9/L$ ($150\text{--}400 \times 10^9/L$). He had elevated erythrocyte sedimentation rate (ESR) of 133 mm/h (normal: 5–7 mm/h) and C-reactive protein (CRP) of 153 mg/L (normal <10 mg/dL). His total serum protein (69 g/L) was normal, but serum albumin (32 g/L) was reduced (normal value of total protein is 62–80 g/L and albumin 36–55 g/L). His liver enzymes were elevated: alkaline phosphatase was 36 IU/L (normal 22–29 IU/L), alanine aminotransferase was 19 IU/L (normal <12 IU/L) and aspartate aminotransferase



Figure 1: Oedema of the hand and desquamation of the skin of the lower limb



Figure 2: Oedema and desquamation of the hand



Figure 3: Desquamation of the skin over the palm of the patient



Figure 4: Desquamation of the skin over the sole of the foot



Figure 5: A sheet of skin from the sole of the foot

phosphatase was 17 IU/L (normal <12 IU/L). Echocardiogram done in the 3rd week of the illness revealed a left CAA, 16 mm thick with unobstructed outflow. There was no evidence of myocarditis, pericarditis, pericardial effusion or valve disease. A follow-up repeat echocardiogram 2 months after the initial echocardiogram showed that the CAA had reduced to 9mm with a complete resolution of CAA seen in a 3rd echocardiogram 10 months later.

He received aspirin and intravenous immunoglobulin G (IVIG) at 2 g/kg in two divided doses 24 h apart. The fever subsided 6 h after the second dose of IVG. He was discharged and presently on follow-up with a paediatric cardiologist.

DISCUSSION

KD is a rare diagnosis made amongst children with febrile illness in Africa and other developing countries.^{15,16} Studies have shown that the incidence of KD is higher in developed countries.¹⁴ The American Heart Association guideline suggests that the diagnosis of KD should be made in children who meet up with the diagnostic criteria, which include fever of 5 or more days in addition to having four or five cardinal symptoms. These symptoms are polymorphous rash (but usually maculopapular in 95% of cases), oropharyngeal changes, oedema or redness of the palm and feet, bilateral non-exudative painless bulbar conjunctival injection and unilateral cervical lymphadenopathy.¹² Our patient had fever for 3 weeks in addition to having all the cardinal symptoms and signs except for cervical lymphadenopathy. He, therefore, met the criteria for making the diagnosis of complete KD. In addition to these, he had deranged laboratory results of anaemia, leucocytosis, elevated ESR and CRP and

hypoalbuminaemia, which are commonly found in children with KD.³

Echocardiography is an important diagnostic test to evaluate these children for CAA, a condition prevalent in about a quarter of them, especially when they are not treated with IVIG or occurs at a lower rate when immunoglobulin treatment is delayed.³ Pre-morbid echocardiography is not routinely done but may be helpful in comparing the development and progress of CAA. The echocardiography done 3 weeks after the onset of fever revealed that our patient had already developed CAA, a condition that is highly contributory to the case fatality in KD.^{1,2,8} The left CAA was 16 mm at the first echocardiogram but reduced to 9 mm at repeat; thus, the Z-score of coronary dilatation reduced from 34.18 to 16.75 (normal Z-score is <+2.5-->-2.5). This shows that the child had a severe and grossly abnormal coronary artery with some improvement following treatment.

Another finding in KD is arthritis, which may have been present in our patient since he had signs of inflammation of the knees swollen, warm, tender knees and was limping. Raised ALP may suggest hydrops of the gallbladder, but we could not ascertain if he had this since no abdominal ultrasound or computerised tomography scan was not done.

Febrile illnesses are quite prevalent amongst children under the age of 5 years, more so in tropical countries like Nigeria and infectious disease such as malaria commonly causative.^{9,10,17,18} It is, therefore, not surprising that the initial diagnosis made in our patient was malaria. Although the thick film for malaria parasite test was positive, it is not unusual to find malaria as a comorbidity with other febrile illnesses amongst children under the age of 5 years in tropical Africa.^{19,20} Our patient may have had malaria with KD since the malaria parasite test was positive in addition to having symptoms of malaria.^{9,10} Although the authors did not find any reported case of KD and concomitant malaria, Animasahun *et al.*¹⁵ described the management of five cases of KD who also received antimalarial. That report, however, did not explicitly mention if any test for malaria was conducted before the commencement of the antimalarial. The fever in malaria may be high grade like that of KD; it may come in bouts every 24, 48 or 72 h depending on the malaria parasite species involved. When appropriate antimalarial treatment is administered either orally or Parenterally, symptoms including fever, usually subside.

Measles or other viral exanthema is another diagnosis often mistakenly made amongst children with KD.^{1,3} Measles

is a viral vaccine-preventable disease commonly found amongst children under the age of 5 years in developing countries like Nigeria.^{21,22} Children with measles share some common symptoms of KD such as fever, maculopapular rash and conjunctivitis. In severe cases, measles can be complicated with diarrhoea and acute severe malnutrition with nutritional oedema of the feet and palms.¹³ It is, therefore, possible for the diagnosis of KD to be missed by attending physicians.¹⁴ Measles, however, differs from KD by the presence of Koplik spots, its pathognomonic feature, which appears on the 3rd–4th day before the appearance of rash. The conjunctivitis in measles is usually exudative unlike the non-exudative conjunctivitis in KD. The rash in measles characteristically begins behind the ears and hairline on the forehead before spreading to other parts of the body, and the desquamation is fine, extensive and usually spares the palms and soles of the feet, unlike what happens in KD where the palms and soles of the feet are involved, and the desquamation maybe is in sheets. The appearance of the rash in measles usually heralds the resolution of symptoms including the fever, but this may not be the case in KD. The laboratory features in measles usually include leucopaenia and normal platelet count, ESR and CRP, while the opposite is often seen in KD. The platelet count in this patient was, however, normal. A similar finding was reported by Animasahun *et al.*¹⁵ in Lagos about the case number two. BCG erythema and induration does occur in KD but not in measles.¹⁵ Our patient did not have BCG scar reactivation.

The mainstay of treatment is the use of IVIG and aspirin as anti-inflammatory agent.²³ It is reported that CAA develops in up to 25% of children not treated with immunoglobulin within 10 days and this risk decreases to 3%–5% if immunoglobulin is given early.^{1,2} Early recognition and administration of IVIG would thus improve the outcome of KD. Although the response to IVIG may be variable when given later than 10 days after the onset of illness, the fact that our patient became afebrile 6 h after receiving the second dose suggested that he responded to treatment. In addition, there was an improvement in the CAA as it decreased from 16 mm to 9 mm 2 months after the initial echocardiogram.

CONCLUSION

This case report highlights the fact that the diagnosis of KD should be entertained amongst children presenting with prolonged fever and maculopapular rash in our environment. This is important so that prompt treatment can be instituted to prevent cardiac complications and mortality.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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