KAWASAKI DISEASE IN A TWO YEAR OLD NIGERIAN CHILD; FULL RECOVERY WITH SUPPORTIVE TREATMENT.

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ABSTRACT

Data from a two year old male patient managed at the University of Uyo Teaching Hospital, Uyo, southern Nigeria, is presented to highlight the sporadic occurrence of Kawasaki disease in a Nigerian child who was diagnosed as having a viral exanthema at the onset of illness, at a peripheral centre. His presentation at this tertiary hospital, coupled with the high clinical index of suspicion confirmed the evolving diagnosis. This report also highlights the associated challenges of therapy, especially non-availability of standard recommended treatment like the intravenous immunoglobulin in most clinical settings in Nigeria, and the clinical evidence, that supportive therapy is very helpful in this condition.

Key words: *Kawasaki, child, Uyo, southern Nigeria*

INTRODUCTION

Kawasaki disease is an autoimmune disease in which the medium sized blood vessels throughout the body become inflamed.[1-5] It is mostly seen in children below five years of age. It affects boys more than girls. People of Asian ancestry, particularly Japanese and Korean people are most susceptible. Also affects people of Afro-Caribbean ethnicity. The disease was rare in Caucasians until the last few decades and incidence rate fluctuates from country to country.[1-5] Currently, Kawasaki disease is the most commonly diagnosed pediatric vasculitis in the world. By far the highest incidence of Kawasaki disease occurs in Japan, with the most recent study placing the attack rate at 218.6 per 100,000 children less than five years of age (~1 in 450 children).[5] At this present attack rate, more than 1 in 150 children in Japan will develop Kawasaki disease during their lifetime.[3-6]

It affects many organ systems, mainly those including the blood vessels, skin, mucous membrane, and lymph nodes; however its rare but most serious effect is on the heart where it can cause fatal coronary artery aneurysms in untreated children. Without treatment, mortality may approach 1%, usually within six weeks of onset. With treatment, the mortality rate is 0.17% in the United States. [1,4,5] There is often a pre-existing viral infection that may play a role in its pathogenesis. The conjunctivae and oral mucosa, along with the skin, become red and inflamed. Oedema is often seen in the hands and feet. One or more cervical lymph nodes are often enlarged. Also, a recurrent fever, often with body temperature of 37.78°C (100°F) or higher, is characteristic of the acute phase of the disease. In untreated children, the febrile period lasts on average approximately 10 days, but may range from five to 25 days. The disorder was first described in 1967 by Tomisaku Kawasaki in Japan.[4-6] The diagnosis of Kawasaki disease which is rarely seen in Nigerian children, and the challenges of standard treatment, necessitated this report.

Kawasaki disease can only be diagnosed clinically (i.e. by medical signs and symptoms). There exists no specific laboratory test for this condition. It is difficult to establish the diagnosis, especially early in the course of the illness, and frequently children are not diagnosed until they have been seen by several health care providers.[1,5-7] Many children,

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especially infants, eventually diagnosed with Kawasaki disease do not exhibit all of the above criteria. In fact, many experts now recommend treating for Kawasaki disease even if only three days of fever have passed and at least three diagnostic criteria are present, especially if other tests reveal abnormalities consistent with Kawasaki disease.[1,5,7] In addition, the diagnosis can be made purely by the detection of coronary artery aneurysms in the proper clinical setting.

Classically, five days of fever plus four of five diagnostic criteria must be met in order to establish the diagnosis. The criteria are: (1) erythema of the lips or oral cavity or cracking of the lips; (2) rash on the trunk; (3) swelling or erythema of the hands or feet; (4) red eyes (conjunctival injection) (5) swollen lymph node in the neck of at least 15 millimeters.[1-3]

As the cause(s) of Kawasaki disease remain unknown, the illness is more accurately referred to as Kawasaki syndrome. Like all autoimmune diseases, the cause of Kawasaki disease is presumably the interaction of genetic and environmental factors, possibly including an infection. The specific cause is unknown, but current theories center primarily on immunological causes for the disease. Evidence increasingly points to an infectious aetiology, but debate continues on whether the cause is a conventional antigenic substance or a superantigen. [1-6]

Intravenous immunoglobulin (IVIG) is the standard treatment for Kawasaki disease and is administered in high doses with marked improvement usually noted within 24 hours. Salicylate therapy, particularly acetylsalicylic acid (aspirin), remains an important part of the treatment (though questioned by some). Except for Kawasaki disease and a few other indications, aspirin is otherwise normally not recommended for children due to its association with Reye's syndrome.[1,7]

With early treatment, rapid recovery from the acute symptoms can be expected and the risk of coronary artery aneurysms greatly reduced. Untreated, the acute symptoms of Kawasaki disease are self-limited (*i.e.* the patient will recover eventually), but the risk of coronary artery involvement is much greater.[1,3,4] We report the case of a two year old male with initial symptoms suggestive of a viral illness, later diagnosed to be Kawasaki disease at presentation to our centre.

CASE REPORT

ASG, a two year old male resident in Uyo, Akwa Ibom state but from the Idoma tribe of Benue State of Nigeria, presented with fever of five days duration and body rash of three days. The fever was high grade, continuous, associated with chills, no rigors. Rashes started two days later from the trunk, and later spread to the neck. There was bilateral redness of eyes, but no eye discharge. He had erythema of the mouth and rashes and desquamation of the neck. There was pruritus. He was taken to a private facility and given co-amoxiclav suspension which was discontinued after 24 hours as this was presumptuously, linked with the rash. Intravenous Ceftriaxone was then commenced by the third day of illness at the same private facility. With no relief of symptoms, parents were anxious, and he was brought to our tertiary health facility for further treatment.

The past medical history showed no previous admissions. He was on no prolonged medications. There is no family history of such illnesses. Child's pregnancy was carried to term. Mother delivered by caesarian section by choice. Puerperium was uneventful. Child had been fully immunized. Was on mixed feeding from birth, thereafter, was introduced to cereals and family diet. He developed anorexia following onset of illness.

Developmental milestones were normal for each stage. He is the second of two children. The first child, a four year old female was well. Both parents are public civil servants,

with tertiary level of education. The review of systems revealed significant findings in the digestive system, with redness of the oral mucosa and tongue, and anorexia. In the musculo-skeletal system, they were rashes on the neck and trunk, and redness of the palm and feet.

On examination, he was acutely ill, overweight, febrile (38.6°C) with bilateral conjuctival injection, redness of the lips and oral mucosa, polymorphic rashes on the neck and trunk, palmar and plantar erythema, and unilateral significant cervical lymphadenopathy measuring 2 – 3cm, firm, discrete and moderately tender. He was not jaundiced, not cyanosed, not pale, with no signs of dehydration, and there was no peripheral oedema.

Digestive system showed desquamation and cracking of the lips with redness of the tongue and oral mucosa. Abdomen was full, with an enlarged liver, 8cm from the right costal margin, firm, smooth and mildly tender, spleen was tipped. Bowel sounds were normoactive.

Musculoskeletal system showed a desquamating rash around the anterior part of the neck, the groin and the perianal region. A polymorphic rash was seen on the trunk, and there was erythema of the palms and soles. The respiratory system revealed a respiratory rate of 24 cycles per minute, he was not dyspnoeic. The chest movement was symmetrical, and the trachea central. There was normal tactile fremitus, resonant percussion notes and vesicular breath sounds. Cardiovascular examination, showed a pulse rate of 120 beats per minute, full volume, regular, with a normal first and second heart sounds. The child was conscious and alert, but weak. No signs of meningeal irritation, and no cranial nerve deficit. He had normal muscle tone globally.

A diagnosis of Kawasaki disease was made and he was admitted into the children emergency unit. Several investigations were requested for which included:

A full blood count showed anaemia with a packed cell volume of 26% and haemoglobin of 9.4g/dl. Red blood cell count – 3.6 x 10^{12}/L. Mean corpuscular volume -75fl. Mean corpuscular haemoglobin – 27pg. Mean corpuscular haemoglobin concentration – 36g/dl. Platelet count was normal - 200 x 10^{9}/L, with clumping of platelets on the peripheral film. There was leucocytosis - 14.9 x 10^{9}/L with neutrophilia of 83%, and severe toxic granulation of neutrophils .Lymphocyte was 16% and eosin 01%. The blood film on admission showed hypochromasia of red blood cells.

Malaria parasites were not seen on peripheral smear. Blood culture and urine culture grew no organisms.

Urinalysis showed deep amber clear coloured urine. A trace of protein, Bilirubin - +++, nil blood, nil glucose, specific gravity of 1.020, pH of 6.5, Nil ketones and Normal urobilinogen.

Electrolytes/Urea/Creatinine showed:

Electrolytes	**On admission**	**Three days later**
Sodium (mmol/l)	124	130
Pottasium (mmol/l)	3.8	3.5
Chloride (mmol/l)	94	93
Bicarbonate (mmol/l)	19	20
Urea (mmol/l)	2.0	1.7
Creatinine	331	245

Intravenous Ceftriaxone (Rocephin brand) was given to the child at a dose of 1g every 12 hours, and intravenous Gentamycin 30mg given 8 hourly, for one week. Also, 1250mls of intravenous dextrose 8% in one-fifth saline to run over 24 hours at 14 drops per minute was continued till child was able to take adequately per oris, which was on the third day into admission. He tolerated oral sips on admission, and semi-solid foods, two days later. The child remained continuosly febrile for the next five days after admission with temperature ranging between 37.8^0C and 39.1^0C. He received antibiotics for seven days, and was eventually discharged on father's request after one week of hospital admission. Child recovered fully at home, was seen at the follow-up clinic ten days later, and followed up forth-nightly for three months. He had no residual sequelae of the illness.

DISCUSSION

Kawasaki disease has its highest incidence among people of Asian ancestry. The rarity of its occurrence and/or reports from African children, more-so Nigeria, the diagnostic and therapeutic challenges in management, often low index of suspicion among many clinicians, necessitates this clinical report.[8-12] In Nigeria, and most of Africa, very few cases have been seen or reported. A case report each has been documented in a three year old female child in the western part of Nigeria[8] and also in an adolescent male in North-east Nigeria.[9] A single report from the southern part of Nigeria was documented twenty-five years ago.[10] These show the very sporadic nature and probably under reports of this disease in Nigeria.

The index child had all the five definitive criteria for the diagnosis of the disease and was very ill. He received only supportive treatment, as intravenous immunoglobulin was not available despite concerted attempts to procure it. Intravenous immunoglobulin has been shown to significantly reduce the risk of cardiac complications.[1,4-7] There was reluctance to use Acetyl salicylic acid (aspirin), because of the risk of Reye's syndrome in young children,[1,6,7] especially in a child one could take no further risk of a co-morbidity.

He did not manifest any obvious cardiac abnormalities on clinical examination. Echocardiography deferred and performed at the fourteenth day of onset of illness showed normal cardiac size, coronary vessels and blood flow. It was deferred to increase the chances of picking up any obvert or subtle findings, as documented in literature. The child however recovered fully with no cardiac manifestations despite the non-use of any conventional therapy. He was followed up closely for twelve weeks. We propose that early presentation and prompt diagnosis of index case, coupled with the dedicated supportive care, played a huge role in reducing morbidity in this affected child.

CONCLUSION

Kawasaki disease is rarely seen in Nigerian children, but nonetheless occurs sporadically. It is important for clinicians to maintain a high index of suspicion in children with symptoms of viral exanthema, and to give dedicated supportive treatment even with non-availability of conventional therapy like the intravenous immunoglobulin in resource – poor settings.

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