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Case Report`

Kawasaki Disease Associated with Streptococcal Infection and Facial Nerve Palsy: A Case Report

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ABSTRACT

Background: Kawasaki disease (mucocutaneous lymph node syndrome), an acute febrile vasculitis of childhood that affects medium and small-sized arteries, is uncommonly reported in the West African sub-region. Its diagnosis relies on the presence of a constellation of clinical signs which could mimic or coexist with infectious viral or bacterial agents, thereby requiring a high index of suspicion. **Case presentation:** We report a two-year, ten months-old boy who presented with prolonged high-grade fever for over 3 three weeks; non-purulent conjunctivitis, unilateral cervical lymphadenopathy, cracked lips, reddish tongue, diffuse oedema, erythema of his palms and soles; skin desquamation over the tips of his digits and left-sided facial nerve palsy. He had leukocytosis, thrombocytopenia, and elevated inflammatory markers. Throat swab yielded *Streptococcus pyogenes species*; however, serial echocardiography was unrevealing. He was managed with aspirin, steroids, bed rest, and antibiotics (based on culture sensitivity), but intravenous immunoglobulin (IVIG) was not readily available and therefore was not administered. His inpatient clinical course showed fever persisting into the second week of inpatient management that gradually became undulating by the third week before lysing. **Conclusion**: The rare complication of facial nerve palsy is highlighted, while the evidence for acute bacterial infection posed further diagnostic challenges in this child with clinical features of Kawasaki disease. The unavailability of IVIG also portends a prolonged course for the acute stages, which are largely unresponsive to antipyretics and antibiotics. The importance of long-term follow-up for potential coronary artery aneurysms in the face of risk factors is further emphasized.

Keywords: *Kawasaki disease, Streptococcal infection, Facial nerve palsy*

Introduction

Kawasaki disease (KD), also called mucocutaneous lymph node syndrome, is a generalized vasculitis that affects medium-sized arteries. It was first described in 1967 by Dr. Tomisaku Kawasaki in Japan.[1] It is characterized by systemic inflammation that manifests as persistent fever, erythema of the mucous membranes, bilateral nonexudative conjunctivitis, rash, swelling and redness of the hands and feet, and cervical lymphadenopathy.[2] The disease most frequently occurs in children aged between six months and five years.[3] It is usually self-limiting but, in some cases, can result in a coronary artery aneurysm. Those who develop coronary artery aneurysms may also develop coronary thrombosis or stenotic lesions and are at risk of myocardial infarction, congestive cardiac failure, and sudden death.[4] Apart from cardiac complications, neurologic manifestations such as irritability, lethargy, and aseptic meningitis have also been reported.

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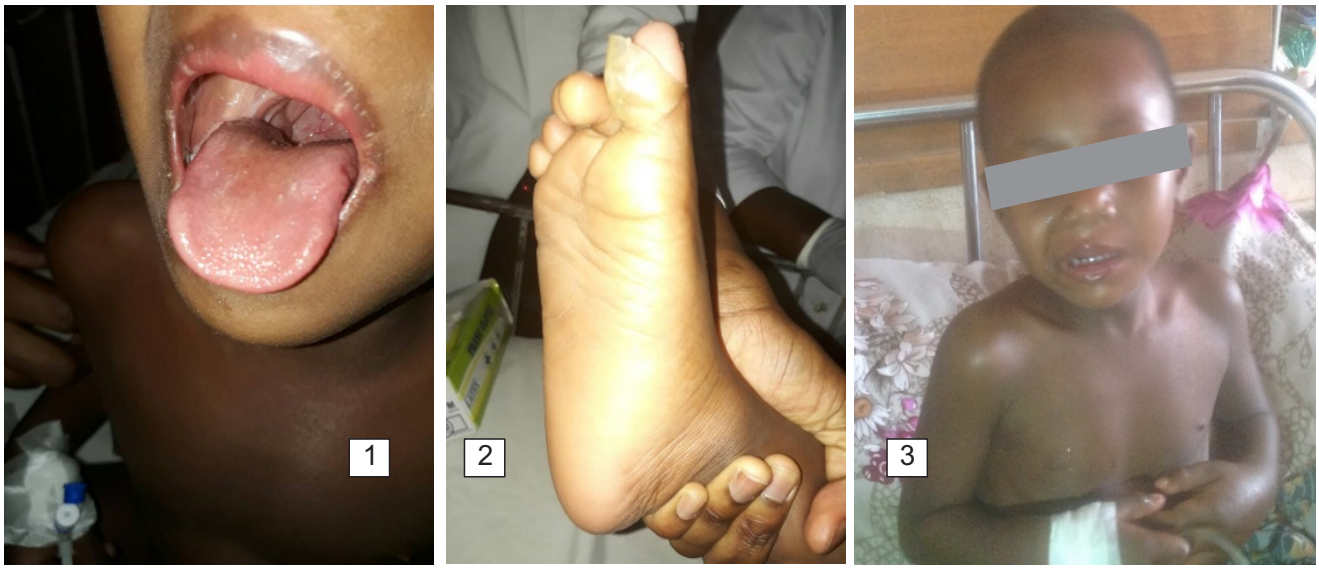
However, neurologic complications are uncommon. Facial nerve palsy (FNP) has been reported as a neurologic complication of KD since 1974.[5] It has been associated with a higher incidence of coronary artery lesions and a more severe disease.[5] The aetiology of KD remains unknown. Viral and bacterial cultures from patients with KD have failed to yield a consistent organism, nor have molecular methods identified a causative agent.[6,7] However, observed bimodal seasonal peaks in KD may suggest that different infectious diseases trigger the disease in winter/spring and summer.[8] Here, we report a case of Kawasaki disease associated with group A streptococcus infection and facial nerve palsy.

Case report

We report a two-year, ten months-old boy who presented with prolonged high-grade fever of over three weeks, irritability, and skin rashes. At the onset of the illness, he had a fever, rhinorrhea, and cough, for which he was seen and admitted to a private hospital. He was subsequently referred to our hospital on account of persistent fever and irritability despite antibiotics. On examination, he was noticed to be irritable and febrile with a temperature of 39.9°C, non-purulent conjunctivitis, unilateral cervical lymphadenopathy, cracked lips, reddish tongue, diffuse oedema, erythema of his palms and soles; skin desquamation over the tips of his digits, enlarged tonsils and left-sided facial nerve palsy (FNP). A detailed central nervous examination did not yield any further abnormality. Cardiovascular and respiratory examinations were essentially normal.

Laboratory investigation findings revealed anaemia, leukocytosis of 15.4 x 10^9/L with 78% neutrophils, thrombocytopenia, and elevated erythrocyte sedimentation rate (ESR). Throat swab culture yielded *Streptococcus pyogenes species*; however, Chest X-ray, electrocardiogram, and serial echocardiography were unrevealing. A clinical diagnosis of Kawasaki disease was made. He was managed with aspirin 80 mg/kg/day, bed rest, and Ceftriaxone at 100 mg/Kg antibiotics (based on culture sensitivity), but intravenous immunoglobulin (IVIG) was not readily available. His inpatient clinical course showed fever persisting into the second week of inpatient management that gradually became undulating by the third week before lysing. The facial nerve palsy significantly improved before his discharge after three weeks on admission. Serial echocardiography done at six months, one year and two years were essentially normal. We followed him up for two years and lost to follow-up during the covid-19 pandemic.



Pictures (1) : Reddish tongue and cracked lips, (2) : Desquamation of the feet and (3) : Facial nerve palsy and oedema of the hands

Discussion

Kawasaki disease is an acute vasculitis of medium-sized arteries which mainly affects children less than five years. Its diagnosis is mainly clinical based on symptoms that meet the classic diagnostic criteria, including fever of >5 days, polymorphous exanthema, cervical lymphadenopathy, non-purulent conjunctivitis, and changes in the lips, oral cavity, and extremities.[9] Early diagnosis is difficult for the initial attending physician as KD symptoms are similar to other conditions, not all symptoms appear simultaneously, and there are no specific diagnostic tests. It, therefore, needs a high index of suspension to diagnose and should be suspected in any child with a history of high fever for more than a week with rash and lymphadenopathy. Our patient had all five principal features in addition to the fever that lasted more than five days at the time of presentation in our hospital.

The dreaded complication of Kawasaki disease is coronary artery disease; however, neurologic complications such as aseptic meningitis, ataxia, focal encephalopathy, cranial nerve palsies, cerebral infarction, and transient hemiplegia have been reported.[10,11] Our case is the first to be reported with facial nerve palsy in our centre. Facial nerve palsy results from ischaemic vasculitis affecting the arteries supplying the nerve and immunologic mechanism. Although FNP in Kawasaki disease is reported to resolve spontaneously within three months, its presence is thought to be associated with a more severe clinical progression and higher incidence of coronary artery disease, which may reflect late diagnosis, more severe inflammation, and vasculitis.[5,12] Even though our patient presented late and had facial nerve palsy, no coronary artery disease was noted. This is in contrast to earlier reports that showed the presence of FNP is associated with severe disease progression and the development of a coronary aneurysm.[12] Although early administration of intravenous immune globulin (IVIG) has been shown to reduce the risk of coronary artery lesion and hasten resolution of FNP[13,14], it was not given to our patient due to non-availability. However, aspirin was given to the index patient even though it has not been reported to reduce the risk of a coronary aneurysm or hasten the resolution of FNP. Despite this limitation, our patient did not develop coronary artery disease, although FNP took more than three months to resolve completely.

The exact cause of KD is unknown, but several studies have associated the inflammation in the disease with several infectious agents in genetically susceptible individuals.[15,16,17] Streptococcal and staphylococcal super antigens have also been implicated.[18]

Conclusion

Diagnosis of Kawasaki disease should be suspected in any child with a history of prolonged high fever, body rashes, and lymphadenopathy. This case highlighted the rare complication of FNP in a child whose clinical features satisfied the diagnostic criteria for KD in the presence of a severe streptococcal infection. The unavailability of IVIG also portends a prolonged course for the acute stages, which are largely unresponsive to antipyretics and antibiotics.

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