Case report

Kawasaki Disease presents as acute abdomen mimicking Appendicitis

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INTRODUCTION

Kawasaki disease (KD) is an acute febrile illness of childhood with the highest incidence occurring in Asian children. This is a vasculitis mainly affecting medium-sized arteries (1). The coronary arteries are the most vulnerable vessels, but occasionally popliteal, and brachial arteries can be affected. Usually, this vasculitis results in aneurysms in affected vessels. Sometimes thrombi formed within the lumen and obstructed the blood flow. In typical Kawasaki Disease, patients present with ocular manifestations such as bilateral non-exudative conjunctival injection with limbic sparing, nonsuppurative lymphadenitis, polymorphous cutaneous lesions, strawberry tongue, fissures of lips and body edema, and peeling of skin can be seen.

Sometimes there can be atypical presentations like acute abdomen which make it difficult to establish the diagnosis prompting more invasive and unnecessary interventions.

This case report describes a 5 year old girl presenting with acute abdomen and subsequently diagnosed to have Kawasaki disease.

CASE PRESENTATION

A 5-year-old girl presented to the surgical unit of Teaching Hospital Ratnapura with acute onset abdominal pain for a 1-day duration. The pain was mainly in the periumbilical region which a was continuous, aching type with no radiation. After admission, a clinical diagnosis of acute appendicitis was made clinically and confirmed ultrasonically. Appendicectomy was performed under general anesthesia on day 1 of admission and a minimally inflamed appendix was demonstrated histologically.

Since day 1 of admission, the child has been having high-grade fever spikes, the maximum being 102^0F documented. Despite broad-spectrum antibiotics and antipyretics, the fever was persistent. The mother also has noticed redness in the bilateral eyes which is non-purulent along with reddish lips and tongue since day 2 of fever and on the 3rd-day child has developed a generalized urticarial rash. On clinical suspicion of Kawasaki disease, a 2D echo was done and echogenic dilatation of LMCA[1] was detected.

After that the child was transferred to the pediatric medical ward on day 4 of fever.

Upon admission, the child was febrile and had a heart rate of 128bpm with BP-85/53mmHg and SPO_2 – 99% on air. Non-suppurative bilateral conjunctivitis with strawberry tongue was noted. She was found to have right-side non-tender cervical lymphadenopathy measuring 2*2 cm.

Laboratory findings revealed WBC-23.78*10^3, Neutrophils- 83.2%, Lymphocytes- 10.2%, Eosinophils – 5.9%, platelets – 420*10^3, ESR- 115 mm/hour, CRP-94 mg/l. Urine full report showed 6-8 pus cells with urine culture and blood culture were sterile.

Soon after transfer, the child was given IV Immunoglobulin 2g/kg dose as infusion and started on oral aspirin 80mg/kg divided into 4 doses every 6 hours. Over the next 24 hours, a marked clinical improvement was observed. Yet on the day following Intravenous immunoglobulin, the child developed bilateral mild subconjunctival hemorrhages with intact vision. On day 7 of illness child was found to have desquamation of fingers, a late manifestation of KD.

After 10 days in the hospital, the stay child was discharged with oral aspirin and the date for 2Decho was given. On follow-up, after 2 weeks the child was completely symptom-free and a repeat 2D echo was normal.

Figure 1: subconjunctival hemorrhages

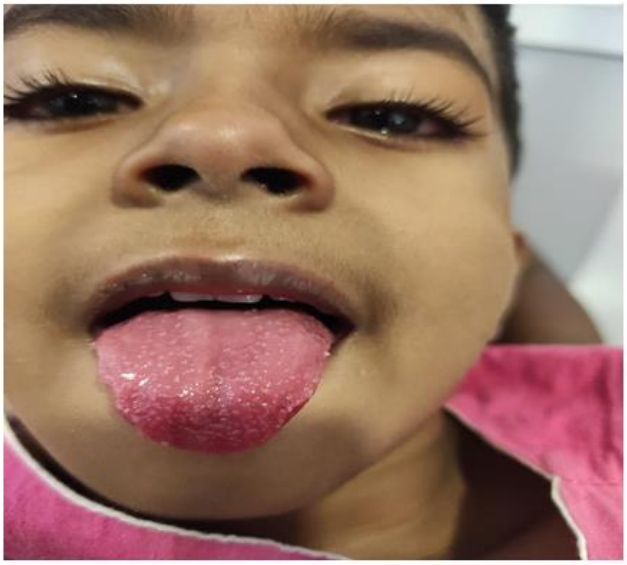

Figure 2: Strawberry tongue appearance

(Consent was taken from parents to publish photographs for academic purposes.)

DISCUSSION

Kawasaki disease is an acute febrile illness where vasculitis is the main pathology affecting medium-sized arteries and the most common cause of acquired heart disease in children. The etiology of Kawasaki disease remains unknown, but it could be infectious in origin due to some epidemiologic and clinical features. But no single infective agent has been successfully identified yet.

Characteristically high fever, lasting at least 5 days is one of the main clinical manifestations of KD[2]. This fever is usually unresponsive to antibiotics and antipyretics. In addition to fever, there are 5 other principal features of KD. These are; bilateral non-exudative conjunctival injection with limbus sparing, mucositis of the oral cavity with strawberry tongue, changes in extremities including erythema and edema, polymorphous exanthema, and unilateral non-suppurative cervical lymphadenopathy (>1.5cm). Classical KD presents with these usual symptoms and signs. But some may present with unusual symptoms and signs; known as atypical/ incomplete Kawasaki disease. For diagnosis, a patient must have a fever for more than 5 days plus 4/5 of clinical manifestations. For incomplete Kawasaki 2D echocardiogram is helpful for the establishment of diagnosis as to start treatment early to prevent coronary artery aneurysms before day 10 of illness as the risk of developing coronary artery aneurysms can be reduced to 5% from 20% if treated early (1)(9).

Some of the atypical presentations of Kawasaki disease are acute abdomen mimicking appendicitis, renal impairment, facial nerve palsy, testicular swelling, pulmonary nodules, and Gastrointestinal symptoms such as vomiting and diarrhea. (2)(3)(6)(7).

Acute abdomen could be the earliest symptoms of Kawasaki disease mainly in older children. The pathophysiology behind this presentation is vasculitis and some may have a clinical suspicion of appendicitis as a result of appendicular vasculitis. These children may initially undergo an X-ray abdomen, and ultrasonogram to look for evidence of pneumoperitoneum, and appendicitis and primarily to look for surgical causes of acute abdomen leading to accurate diagnosis easily overlooked. some children would undergo appendicectomy on the clinical ground but fever may persist raising the probability of KD (7).

Investigations wise neutrophilic leukocytosis, thrombocytosis, high CRP, and ESR raise suspicion of KD. Atypical biochemical parameters like deranged liver enzymes, hypoalbuminemia, hyponatremia, Ultrasonic evidence of hydrops Gallbladder are also seen. More importantly 2D echocardiogram may show early changes like increased echogenicity, and ectasia of coronary vessels before advancing into aneurysms.

Available treatment modalities are Intravenous immunoglobulin 2g/Kg (5). Aspirin 80 to 100mg/ Kg /Day until fever-free for 48 to 72hours flowed by 3mg to 5mg /kg for 6 weeks usually recommended. In resistant Kawasaki disease where there is no adequate response to Intravenous Immunoglobulin repeat dose of immunoglobulin and Intravenous methylprednisolone pulses, infliximab, cyclophosphamide, and methotrexate are also described in the literature. In addition to Aspirin other anticoagulants like clopidogrel, dipyridamole, warfarin, and heparin are also being used (4)(9).

Key words

Kawasaki disease, Acute abdomen, Acute appendicitis, atypical presentation

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