

## Young girl with Kawasaki disease

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### ABSTRACT

Kawasaki disease (KD) is an acute multisystem necrotizing vasculitis of unknown etiology involving medium-size vessels. It usually occurs in young children between 1 to 5 years of age. The diagnosis of classic KD is based on the simultaneous presence of high fever for 5 or more days with at least four of the remaining five symptoms which includes non-purulent bilateral conjunctivitis, changes in the lips or oral cavity, cervical lymphadenopathy, polymorphous exanthema, desquamation of the peripheral extremities. We present a case of a 4 years old girl who presented with history of fever, pain in multiple joints, oral and lip ulcers and peeling off of her skin on her palms and soles. She was started on aspirin and intravenous immunoglobulins (IVIG) to which she responded very well and was asymptomatic within a week. She had initially been managed empirically elsewhere for infection for over 2 months. Her echocardiography was normal and despite having delayed diagnosis and management, did not develop any cardiac complications.

### Keywords:

Kawasaki disease, diagnosis, lip ulceration, skin desquamation, immunoglobulin.

### INTRODUCTION

Kawasaki disease (KD) is an acute multisystemic vasculitis with unknown etiology affecting medium and small-size vessels.<sup>1</sup> The disease affects 8.1/100 000 children under 5 years of age in the UK and has been the commonest cause of childhood acquired heart diseases in the developed countries.<sup>2</sup> Seen mostly in young children between the ages of 1 to 5 years, it presents with history of fever, bilateral conjunctivitis, changes in the lips or oral cavity, cervical lymphadenopathy, polymorphous exanthema and desquamation of the peripheral extremities.<sup>1-4</sup> Infections are close mimickers of KD and if not diagnosed early can lead to life threatening complications. The diagnosis is usually delayed due to rarity of the disease.<sup>5,6</sup> Our patient similarly went to numerous pediatricians and was diagnosed late due to obscure symptoms which mimicked various infections. The patient presented with typical symptoms and responded well to the currently recommended treatment with IVIG and aspirin.<sup>6-9</sup> This case report is a reminder that KD, despite being a rare disease in children, can be easily picked up and requires a high index of suspicion by the wary physician for early diagnosis and management.

A 4-year-old girl referred to rheumatology clinic with complaint of high-grade fever, on and off for 1 month associated with oral and lip ulcers, with erythematous rashes over her palms and soles and peeling off of her skin for 3 weeks. Her lips had cracked open from 2-3 sites and there was evident bleeding. She then developed pain and swelling in her wrists, knees and ankles. She also had bilateral red painful eyes and described having previously developed redness of tongue which had now settled. She initially went to different pediatricians who treated her with empirical antibiotics, with no response. She had an extensive workup done for various infections however they were all negative. On examination she was an ill, irritable looking young girl sitting on bed vitally stable. She had oral and lip ulcers with evident bleeding and red congested eyes. She had erythematous lesions present over her hands and feet with peeling off of skin over her digits. There were palpable, non-tender, discreet lymph nodes in sub mandibular and posterior cervical region. On musculoskeletal examination both her wrists were swollen, tender with painful range of motion. Rest of her systemic examination was unremarkable (Figure 1A and B). Her Hemoglobin was 11g/dl, total leukocyte count  $7 \times 10^9/L$ , platelets 494,000/L, erythrocyte sedimentation rate 55mm/hour, SGPT 10IU/L, serum creatinine 0.5mg/dl, C-reactive protein (CRP) 14.6

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### CASE REPORT



**Figure 1.** Case of Kawasaki disease before and after treatment. [A] Oral and lips cracks and ulcers with evident bleeding. [B] Desquamation of skin with erythematous skin lesions. [C] Resolution of oral and lip ulceration. [D] Improvement of desquamation

(<0.5). Urine examination was normal and blood culture showed no growth. Her anti-nuclear antigen (ANA), double stranded DNA (dsDNA) and extractable nuclear antigen (ENA) profile were negative; ASO titre <200IU, serum ferritin 1000ng/ml and serum lactate dehydrogenase (LDH) 248IU/litre (all within normal ranges). Chest X-ray (PA view) and echocardiography were normal. On the basis of typical clinical picture, patient was diagnosed to have Kawasaki disease and was started on intravenous immunoglobulin 2gm/Kg dose and high dose aspirin (40mg/Kg) on which she clinically improved remarkably (Figure 1C and D) Aspirin dose was later tapered off. At the time of discharge after 2 weeks, her echocardiography was normal. Her parents were counseled for regular echocardiography.

## DISCUSSION

Kawasaki disease (KD) also known as mucocutaneous lymph node syndrome, is an acute multisystem necrotizing vasculitis of medium and small-size vessels. It is a common cause of acquired heart diseases in developed countries. Its etiology remains unknown. The disease is usually seen in young children between the ages of 1 to 5 years.<sup>1-3</sup> Incidence rates as high as 60-150 per 100,000 children below 5 years of age have been reported from several countries.<sup>1-6</sup> It occurs more commonly in males, then females at a ratio of 1.5:1.<sup>5-8</sup> Diagnosis of KD frequently requires exclusion of other diseases like scarlet fever, staph infection, measles, drug reactions. However, it is important to keep a high index of suspicion for the possibility of KD. The diagnosis is based on sequential presentation of fever for 5 or more days with at least four

of the remaining five symptoms. This includes non-purulent bilateral conjunctivitis, changes in the lips or oral cavity, cervical lymphadenopathy, polymorphous exanthema, desquamation of the peripheral extremities.<sup>2,6,9</sup> Presently, KD continues to be a disease associated with several controversies.<sup>5,6,9</sup> The main difficulties lie in how to perform a timely diagnosis, how to prevent cardiovascular complications, and how to treat refractory forms. Hence, alertness is required for incomplete cases which lack all the classical signs or atypical cases which are diagnosed after characteristic coronary artery changes are found on echocardiogram. This case report describes a 4-years-old girl who developed sign and symptoms typical of KD. She was administered intravenous immunoglobulin (IVIG) and responded very well. In this case this 4-year-old girl was worked up and treated empirically for various infections, despite repeated cultures being negative. Due to failure to respond to various treatments, she was finally referred to the pediatric rheumatology clinic for further workup for an underlying autoimmune condition after having developed arthritic features. On detailed clinical review, she had classical manifestations of KD including fever, conjunctivitis, cracked lips, lymphadenopathy, skin changes along with desquamation noted over her peripheries. IVIG and aspirin are considered first-line therapy for the treatment of KD. 80-90% of treated patients show a clinical and biochemical remission to IVIG.<sup>6,8,9</sup> The patient responded well to IVIG and aspirin and was clinically improved within 1 week of treatment. Patient was feeling well and was up and about just after a week. Ideally IVIG should be given as earliest as possible to reduce cardiac complications. Most concerning feature of KD is coronary artery disease. Coronary artery aneurysms may occur in approximately 25-30% of untreated patients. This complication represents the most important adverse prognostic factor and is the leading cause of death in KD.<sup>6,8,9</sup>

As KD is a close mimicker of infection and particularly lack of awareness regarding it due to its

rareness often leads to a delay in diagnosis. A timely diagnosis and an early beginning of treatment represent a key clinical skill to prevent cardiac complications.<sup>6,8,9</sup> Present patient despite being diagnosed and referred to us late did not develop life threatening complications. She had repeated echocardiograms which were all normal. This case report serves as a reminder that KD, despite being rare, can be diagnosed by the wary clinicians keeping a high index of suspicion and can be successfully managed.

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