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Classic Kawasaki disease with a late presentation of post-Kawasaki arthritis

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Introduction

Kawasaki disease (KD) is an acute febrile illness seen worldwide, a medium vessel systemic vasculitis showing predilection for coronary arteries with a risk of developing Coronary Artery Abnormalities (CAA) subsequently (1). The etiology of KD remains unknown. Certain infectious triggers and genetics play a key role in acquiring the disease. Yet, not a single infectious agent has been identified nor isolated through years of research.

Patients present with high-grade remitting fever, poorly responding to antipyretics for at least five days or more and there are 5 principal criteria for diagnosis (1). B/L nonexudative conjunctival suffusion with limbal sparing (2). Erythema of the oral and pharyngeal mucosa with strawberry tongue and red, cracked lips (3). Oedema and erythema of the hands and feet (4). Rash of various forms (maculopapular, erythema multiforme or scarlatiniform) (5). Nonsuppurative cervical lymphadenopathy, usually unilateral, with node size >1.5 cm

Although not included in the principal criteria, perineal desquamation is common in the acute phase. In contrary, periungual desquamation of the fingers and toes begins 2 - 3 weeks after the onset of illness. This sometimes may progress to involve the entire hand and foot.

The incidence of KD in the US is approximately 25/100,000 in children below 5 years of age. Meanwhile in Japan the incidence is 10 times as in the US (2,3).

KD readily responds to Intravenous immuno--globulin (IG) therapy. Around 25% of the untreated patients and <5% of the treated, develop CAA (4). KD with a late onset small joint arthritis is a rarely seen phenomenon despite initial sound recovery (5). We present a case of Classic Kawasaki disease in an 18-month-old baby who initially responded well to IV IG treatment and later presented with small joint arthritis needing corticosteroid treatment.

Case presentation

An 18-month-old baby from an urban region in Sri Lanka who was previously well, presented with remittent fever of 7 days duration which followed the 18-month vaccination. He also had scrotal swelling, excessive crying and perineal desquamation. Child was markedly irritable with B/L non exudative conjunctival injection, generalised polymorphous rash, Cracked lips with strawberry tongue, oedema and erythema of hands and feet. He also had a left sided non suppurative cervical lymphadenopathy with a node size of 2 cm. Except for a fever induced tachycardia, child was haemodynamically stable and abdominal examination was unremarkable.

A full blood count showed a total white cell count of 19x10^3 /UL with neutrophils 79%, Hb 9.2 g/dL and platelet count of 372x10^3 /UL. CRP was 218 mg/dL, with an ESR of 74 mm within the 1st hour. Although he had sterile pyuria, urine and blood cultures were negative.

Fulfilling the clinical criteria for classic KD (5 out 5 clinical criteria) baby was given IV IG 2 g/Kg and anti-inflammatory dose of Aspirin (100 mg/Kg/day in divided 4 doses) was started simultaneously and was weaned down to antiplatelet dose (5mg/Kg/day) after the baby became afebrile in 48 hours. Echocardiogram done on D8 of illness was normal with no coronary artery abnormalities. USS of the Abdomen + KUB was normal with no gallbladder hydrops.

Baby was discharged on D13 of illness where he had good clinical improvement and normal inflammatory markers.

Two days following discharge (on D15 of illness) the baby was readmitted with a small joint arthritis of both hands and feet with diurnal worsening and restricted mobility (Figure 1). The arthritis was mainly involving proximal and distal inter-phalangeal joints of hands and feet with no large joint involvement. There was no fever, no rashes or hepatosplenomegaly. However, ESR on admission was 97 mm/hr and CRP was 78 mg/d. WBC on admission was 17x10^3/UL (Neut - 49%) and the platelet count was 905x10^3 U/L. A repeat echocardiogram was performed which showed no coronary artery abnormalities. The diagnosis of post Kawasaki arthritis was made and baby was started on oral corticosteroids (prednisolone) 2 mg/kg/day which was gradually tapered off within 15 days. A rapid recovery was noted with complete resolving of arthritis as well as the inflammatory markers.

On follow up, his antiplatelet dose of Aspirin was continued up to 6 weeks, and the repeat echocardiogram done at the end of 6 weeks was noted to be normal. The baby was thriving well with good weight gain and no complications noted thereafter.

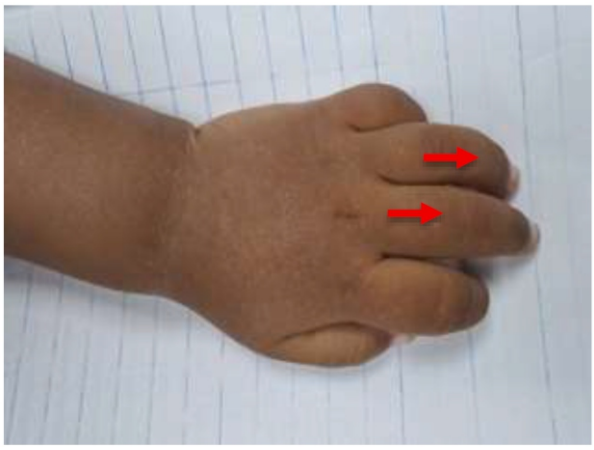

Figure 1: Arthritis of proximal & middle interphalangeal joints (red arrows)

Discussion

KD is the most common cause of acquired heart disease in young children. The intense inflammatory process has a predilection for the coronary arteries, resulting in the development of aneurysmal lesions, arterial thrombotic occlusion or, potentially, sudden death. There is no specific diagnostic test; however, treatment with immunoglobulin and aspirin effectively reduces cardiac complications from 25% to 4.7% in children (1, 4, 6). Inflammation of the myocardium, endocardium or pericardium can occur early in the disease and endothelial dysfunction along with abnormalities of myocardial blood flow may require continuing medication, interventional catheterisation or even cardiac surgery.

KD readily responds to IV IG in majority of cases (2). Arthritis and arthralgia are usually observed in the acute or subacute stage in 15% ~ 45% of KD patients. Arthritis in KD may occur early in the disease around 7-10 days from disease onset or later at around second or third week. Both small and large joint involvement is seen and the arthralgia may persist for several weeks, but is rare (1,5). However, it was recently reported that about 2% of KD patients experienced arthritis in the defervescent state following IVIG management, requiring corticosteroids or a second course of IVIG (8). Although the mechanisms of arthritis in KD have not been

obviously understood, arthritis is considered to reveal a local inflammation of joints triggered by the inflammatory cascade in KD. An inflammation which is noted in the synovial fluid of KD supports the inflammatory nature of KD arthritis. Interleukin IL-1, IL-2, IL-6, IL-8, interferon-γ, and TNF-α, have been demonstrated to be elevated in KD. Our patient developed small joint arthritis of both hands and feet with refusal to walk by around 2 weeks from disease onset with rising inflammatory markers. Mainstay of treatment was oral corticosteroids for 02 weeks, to which he responded well (5). This case report describes the timely identification and treatment of post Kawasaki arthritis without any further complications (5).

References

1. McCrindle BW, Rowley AH, Newburger JW, *et al.* Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. *Circulation*. 2017; **135**(17): e927-e999. DOI: 10.1161/CIR.0000000000000484.
2. Uehara R, Belay ED. Epidemiology of Kawasaki Disease in Asia, Europe, and the United States. *J Epidemiol*. 2012; **22**(2): 79-85. DOI: 10.2188/jea.JE20110131.
3. Makino N, Nakamura Y, Yashiro M, Ae R, Tsuboi S, Aoyama Y, *et al.* Epidemiological observations of Kawasaki Disease in Japan, 2013-2014: From the results of the 23rd nationwide survey. *J Epidemiol.* 2018; **28**(11): 427-32. DOI: 10.2188/jea.JE20170089.
4. Pediatrics: Kawasaki Disease. Nelson Textbook of Pediatrics, 21st Edition, Chapter 484.
5. Cimaz R, Sundel R. Atypical and Incomplete Kawasaki Disease. *Best Pract Res Clin Rheumatol.* 2009; **23**(5): 689-697.
6. Nakamura Y, Yashiro M, Uehara R, *et al.* Epidemiologic Features of Kawasaki Disease in Japan: Results of the 2007-2008 Nationwide Survey. *J Epidemiol.* 2010; **20**(4): 302-307. DOI: 10.2188/jea.JE20090110.
7. Furyk J, Steen S, Swann O. Kawasaki disease: A review. *Emergency Medicine International.* 2013; 645391. DOI: 10.1155/2013/645391.
8. Wood, LE, Tulloh, RM. (2009). Kawasaki disease in children. Heart (British Cardiac Society), **95**(10): 787-792. DOI: 10.1136/hrt.2008.143669.