Case Report

Severe desquamation in Kawasaki disease: Is it somehow protective?

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ABSTRACT

Kawasaki disease is a common vasculitis that typically affects children between one and five years of age. We report a 12-year-old boy who presented following a presumed diagnosis of pharyngitis associated with nondesquamating skin rash and conjunctivitis. Despite treatment with amoxicillin for seven days his fever persisted for ten days and then remitted. Two weeks later, he developed full thickness extensive desquamation of his palms and soles that mandated a visit to emergency department in our tertiary health centre. Physical examination revealed full thickness desquamation of his palms and soles with absence of erythema or swelling and he had unremarkable systemic examination. Laboratory tests showed thrombocytosis and high erythrocytes sedimentation rate. Throat culture and Anti-streptolysin-O titer were negative. Aspirin, anti-platelets dose, was initiated. Echocardiography was performed in the first visit and repeated three times later: at four weeks, six weeks and at three months of the illness revealing normal coronary arteries. Follow up complete blood count and sedimentation rate were normal after six weeks, therefore, aspirin was discontinued. Full thickness desquamation, not as it would be expected, might be somehow protective against the involvement of coronary arteries in Kawasaki disease.

Keywords:

Coronary, Desquamation, Kawasaki, Peeling, Saudi Arabia.

INTRODUCTION

The diagnosis of Kawasaki disease (KD) is based on clinical criteria. Typical KD is diagnosed based on fever lasting five or more days, in addition to four out of the following five criteria: bilateral non-purulent conjunctivitis; erythematous oral mucous membranes; cracked lips and strawberry tongue; cervical lymphadenopathy; polymorphous rash; and acral erythema or desquamation of palms and soles.

Atypical Kawasaki disease occurs in children with fever lasting five or more days and with maximum two or three of the previously mentioned criteria. Delayed or missed diagnosis of Kawasaki is very frequent in atypical (incomplete KD) cases. Epidermal desquamation is a frequent manifestation that prompts the diagnosis of missed cases of KD.

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We report a young boy who presented initially with incomplete features of KD that were fever, conjunctivitis and skin rash with a presumptive diagnosis of pharyngitis. However, he presented later with full thickness extensive desquamation of his palms and soles consistent with a diagnosis of KD with other criteria. We expected this patient to develop further complications given his severe clinical presentation. However, his illness recovered so dramatically with a standard treatment of KD without an involvement of his coronary arteries.

CASE REPORT

A previously healthy 12-year-old Saudi boy was first treated in the primary care with a week course of amoxicillin for a possible pharyngitis because of fever, conjunctivitis and a non-desquamating skin rash. The fever was high grade documented as 39°C and continued for 10 days then remitted, while other symptoms resolved. Two weeks later, he started to have desquamation, which rapidly progressed to a full thickness over his palms and soles when he presented to our centre. The child and his parents were really concerned and anxious about his skin peeling as it is their first time to see such abnormality. Physical examination revealed a well looking young boy, afebrile and normotensive. He has a full thickness desquamation of hands and feet with no erythema or swelling (Figure 1 & 2). Peripheral pulses were palpable and other systemic examination was unremarkable.

His blood tests revealed: Platelets 804 × 10^9/L (reference: 150-450 × 10^9 /L), Erythrocyte sedimentation rate (ESR) 61 mm/h (reference: 0-10 mm/h), C-reactive protein (CRP) 7.09 mg/L (reference: <3.5 mg/L) and Lactate Dehydrogenase (LDH) 281 g/L (reference: 125-220 U/L). Urinalysis, throat culture, and anti-streptolysin-O (ASO) titer were unremarkable. An echocardiogram was normal in the first visit. He was started on oral aspirin 81 mg daily and was followed in the clinic when he developed onycholysis over all fingernails. A repeat blood test in 6 weeks was normal. The echocardiogram was repeated 3 times later, at 4 weeks, 6 weeks, and 3 months when it showed no coronary arteries abnormality; hence, aspirin was discontinued.

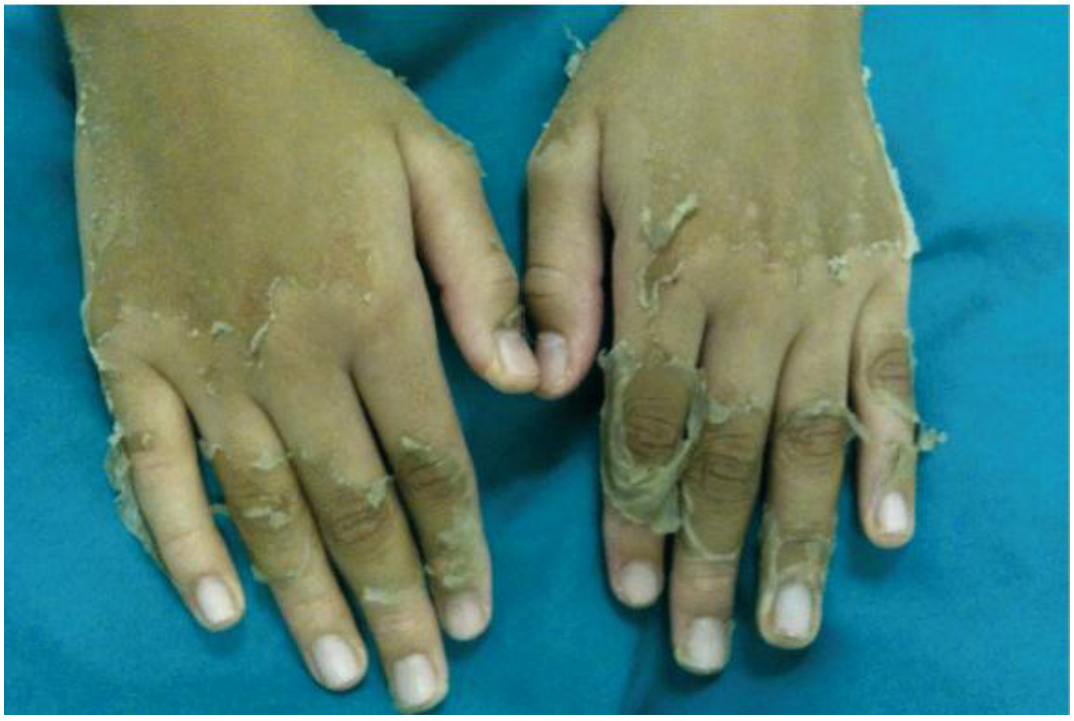

Figure 1- Severe desquamation of hands with no erythema or swelling.

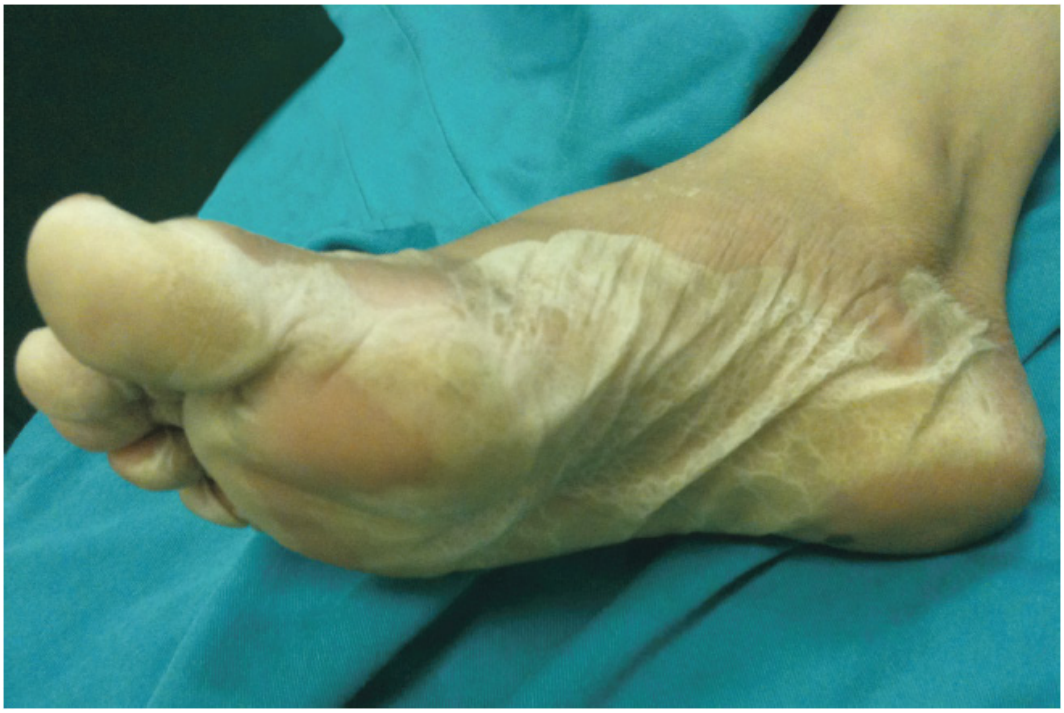

Figure 2- Full thickness skin desquamation of the sole in our patient.

DISCUSSION

Desquamation is very common in Kawasaki disease (KD). In the original report on KD from Japan, 49 out of 50 patients had desquamation and was also above 90% in two different series from the United States [1-4]. Although a lower rate was reported in Chinese population (83%) [5]. In an old report from Kuwait in 1990, 83% of 41 children with KD developed desquamation [6].

A full-thickness epidermal peeling is the hallmark of KD and often prompts the diagnosis in missed cases. Due to lack of confirmatory laboratory investigations, the diagnosis relies on high index of suspicion that is based on a set of clinical features [7]. One of these features is desquamation of the fingers after day 10 of the illness. The sensitivity and specificity of desquamation in KD are unknown. However, patients who did not peel were more likely to develop aneurysms interestingly suggesting somehow a protective role of skin peeling [4]. In our case the full-thickness desquamation was quite impressive and having this presentation in atypical age of Kawasaki patient with incomplete features, one would expect more risk of coronary involvement. However, deliberate follow up of this boy with multiple transthoracic echocardiograms did not show evidence of coronary arteries involvement.

A real estimate of desquamation in a patient with KD is difficult due to differences in the monitoring of patients in both in-patient and out-patient settings. Also, patient might be assessed by different physicians. Other factor that might affect accuracy is the dependence on parental history in some cases.

Among a cohort of 243 children, reported by Wang et al. and diagnosed with KD over a period of 5 years, only 78 (32%) of them did not have desquamation. In this cohort, cases without desquamation were significantly more likely to have coronary aneurysms despite treatment with intravenous immunoglobulin within the first 10 days after the onset of fever [4].

Moreover, Patients suffering from KD may have a single episode of skin desquamation or recurrent episodes [8,9]. Patients who have recurrence of peeling tend to have less peeling over time and they usually don't have significant coronary disease. Typical desquamation of Kawasaki disappears without specific treatment [8].

CONCLUSION

Although the mechanism is not well understood, severe desquamation in patients of KD, in contrast to non-desquamating patients who were more prone to coronary arteries involvement, might be somehow protective of coronary aneurysm in these patients.

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