

CASE REPORT

A rare cutaneous manifestation of Kawasaki disease

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ABSTRACT

Kawasaki disease (KD) is the second common vasculitis in childhood following Henoch–Schönlein purpura. The common skin manifestations in KD are polymorphic exanthema, epidermal desquamation, erythema of the palms and soles and edema over the extremities. Skin erythema at the Bacille–Calmette–Guérin (BCG) vaccination site has been reported frequently in patients with KD. Skin ulceration after BCG vaccination in a context of KD was also reported but not as a part of the disease manifestation. We report a 14-month-old child who presented with clinical criteria for KD and developed left forearm ulcer that did not respond to antimicrobial therapy but responded well to immunomodulatory therapy.

KEYWORDS

BCG; Cutaneous manifestation; Kawasaki disease; Ulcer.

INTRODUCTION

Kawasaki disease (KD) is considered as an acute systemic vasculitis of unknown etiology, which predominantly affects children between the age of 1 and 5 years. KD is a common pediatric vasculitis that is diagnosed mainly based on clinical criteria, which are presence of fever for 5 days or more, beside four out of five principal features: Bilateral non-exudative conjunctival injection, oral mucosal changes, polymorphous rash, changes in extremities and unilateral cervical lymphadenopathy [1]. The common skin manifestations in KD are polymorphic exanthema, epidermal desquamation, erythema of the palms and soles and edema over the extremities [2]. To our knowledge, skin ulceration, as a cutaneous feature of KD, has not been previously reported except after Bacille–Calmette–Guérin (BCG) vaccination [3,4]. We report here a 14-month-old child who presented with clinical criteria for KD and developed left forearm ulcer that did not respond to antimicrobial therapy but responded well to immunomodulation, including intravenous immunoglobulin and steroid therapy.

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CASE REPORT

A 14-month-old boy presented to our Emergency Department in a specialised Children Hospital with a 1-week history of fever (40°C) and symptoms of upper respiratory tract infection. He was initially admitted in a private hospital for 1 week. During that admission, he received Ceftriaxone though he continues to spike high grade fever and found to have an increase in liver enzymes; namely, alanine aminotransferase was 100 U/l (Normal range: 5–55 U/l) and aspartate aminotransferase was 98 U/l (Normal range: 5–34 U/L) and a decrease in platelets count, which was $110 \times 10^9/l$ (Normal range: $150\text{--}450 \times 10^9/l$). Also, his inflammatory markers were elevated, including: C-reactive protein 88 mg/l (Normal range: <3.50 mg/l) and Erythrocyte sedimentation rate 50 mm/hour (Normal range: 0–15 mm/hour). His white cell count (WBC) was $29 \times 10^9/l$ (Normal range: $6\text{--}16 \times 10^9/l$) and therefore, he was referred to our hospital for specialised services. His past medical history was only significant for atopic dermatitis.

Interestingly, he developed a painful, red, progressively swollen skin on his left arm that had descended to his forearm. Moreover, there was a necrotic lesion and hardening of subcutaneous tissue over the distal forearm, which evolved to a deep ulcer in the distal forearm (Figures 1 and 2). The differential diagnosis of such a cutaneous lesion, includes eczema herpeticum, cellulitis or thrombophlebitis. Nevertheless, there was no IV line in the same hand to suggest extravasation, not in support of the latter differential diagnosis. Left arm ultrasound showed diffuse subcutaneous soft tissue edema over distal forearm. Additionally, an MRI of the left forearm reported mild deep subcutaneous inflammation with involvement of superficial layer of extensor muscle group but no osteomyelitis or drainable collection. Antibiotic coverage was expanded to include Vancomycin. Yet, the skin ulcer did not improve after he received antibiotics and he continued to spike fever. However, he had improved significantly after the administration of intravenous immunoglobulin (IVIG) therapy. The patient received two doses of IVIG 2 g/each and received pulse methylprednisolone 30 mg/kg due to a relapse of his fever. Thereafter, he was maintained on methylprednisolone 2 mg/kg/day and aspirin 40 mg daily.

Continuing his care, an echocardiography was repeated after 2 weeks of his illness, reporting mild dilatation (2.6 mm) over the left coronary artery. Moreover, as advised by the plastic surgeon a wound care nurse did a daily dressing for his wound. The left hand ulcer, leukocytosis and thrombocytosis all significantly improved after the IVIG dose; the platelet was $313 \times 10^9/l$ (Normal range: $150\text{--}450 \times 10^9/l$) and the WBC was $12.79 \times 10^9/l$ (Normal range: $6\text{--}16 \times 10^9/l$). After 2 weeks, hand swelling had resolved, and a repeat echo showed no disease progress. Hence, we discharged the patient from our hospital with a treatment of Aspirin 40 mg daily, prednisolone 5 mg oral twice per day and daily dressing with Fucidin for the skin ulcer.

DISCUSSION

To our knowledge, skin ulceration in a clinical context of KD has not been described previously



Figure 1. Erythematous ulcer with black plaque and edema around the lesion.

in the literature as a manifestation of the disease. Uda and Hataya [4] have reported skin ulceration only in relation to the BCG vaccination in a patient with KD. Several case reports, thereafter, have also shown skin reactions at the site of BCG vaccination in patients with KD [3–6]. Although the causation is not yet clear, it is thought that the BCG vaccine proteins play a role in the reaction, which is called hyperantigenemia phenomenon [7].

Our patient had the swelling and subsequently skin ulcer on the left side and the policy in Saudi Arabia is that BCG vaccine is given usually on the left arm. Therefore, we suggest the possibility that our patient had distal ulcer due to BCG vaccination, which was triggered by his KD.

The skin ulcer did not improve with antibiotics as he was given Ceftriaxone and Vancomycin but there was no improvement until the patient had received IVIG. This supports the postulation of the role of KD in the development of the

skin ulcer and demonstrates that it is not just secondary to an infection. More in favour of this is the fact that our patient had marked thrombocytosis and leukocytosis that also improved with IVIG and steroid treatment, typical of KD. In a study done by Lai et al. [3], they reported that KD patients with a skin reaction to BCG had significant thrombocytosis and leukocytosis and are more associated with coronary artery abnormalities. Our patient also had mild coronary aneurysm dilatation that regressed over time and fortunately left no sequelae.

CONCLUSION

Skin ulcer with KD is a rare incidence that we have reported here. Cutaneous ulcer in clinical context of KD should raise the suspicion of soft tissue inflammation, as a manifestation of KD, once the infectious causes are ruled out. IVIG and systemic steroid should be sufficient in treating cutaneous ulcer related to KD.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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None.

ETHICS

Signed informed consent for participation and publication of medical details was obtained from the parents of this child. Confidentiality of patient's data was ensured at all stages of manuscript preparation. The Ethics Committee of our institute; King Abdullah International Medical Research Center (KAIMRC) granted ethics clearance and approval of the study.



Figure 2. Skin sore with irregular borders without erythema during recovery phase.

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