A case of Kawasaki disease masked by pustular type psoriasiform eruptions

Kimya Hassani-Ardakani, BSc, a Thusanth Thuraisingam, MD, PhD, b Duc-Vinh Thai, MD, c Rayan Alkhodair, MD, b Van-hung Nguyen, MD, c Sarah Campillo, MD, d Barbara Miedzybrodzki, MD, b and Fatemeh Jafarian, MD b
Montreal, Canada

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INTRODUCTION
Kawasaki disease (KD) is one of the most common vasculitides of childhood and typically presents with mucocutaneous lesions, thus highlighting the essential role of dermatologists in its early diagnosis. The typical cutaneous manifestations of KD are polymorphous and include maculopapular or morbilliform rashes in 70% to 90% of patients.1 The diagnosis is made by the presence of fever lasting 5 or more days in addition to 4 of 5 clinical signs: oral mucous membrane changes, bilateral conjunctivitis, peripheral extremity changes, polymorphous rash, and cervical lymphadenopathy.2 KD requires prompt diagnosis and treatment, as up to 25% of nontreated patients go on to have cardiovascular complications.1 However, patients may present with atypical or incomplete KD that does not meet the full diagnostic criteria, resulting in delays in diagnosis and treatment.

CASE REPORT
An 8-year-old boy presented with a 5-day history of fever and an acute onset of generalized pruritic rash, which had progressed from the trunk to the upper thigh and groin. His perinatal and medical history was unremarkable. The patient was taking melatonin for sleep daily since age of 6. He had not taken any new medications, and there was no change in his current medications. All vaccinations were up to date.

On physical examination, the patient had diffuse erythematous plaques with pustules involving the trunk, buttocks, upper thigh, and genitals (Fig 1). The face, hands, and feet were spared with no signs of edema or arthritis. The oral mucosa and conjunctiva were normal. Lymphadenopathy was not detected. Cardiac, lung, and abdominal examinations were unremarkable.

A skin punch biopsy was consistent with acute generalized exanthematous pustulosis (Fig 2). Histologic examination found spongiotic dermatitis with elongated rete ridges, superficial dermal edema, and subcorneal neutrophilic collection. The inflammatory infiltrate consisted of lymphocytes, neutrophils, and minimal eosinophils.

The absence of culprit drugs and minimal eosinophils on histology made the diagnosis of acute generalized exanthematous pustulosis unlikely. Based on the clinical and histopathologic findings, the diagnosis of pustular psoriasis was made, and the patient was treated with mid-potency topical

Abbreviation used:
KD: Kawasaki disease

From McGill University Faculty of Medicine a; Division of Dermatology, b and Department of Pathology, c McGill University Health Centre; and the Division of Pediatric Rheumatology, d Montreal Children’s Hospital.

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Correspondence to: Dr Fatemeh Jafarian, MD, Division of Pediatric Dermatology, McGill University Health Center, Montreal Children’s Hospital, 1001 Boulevard Decarie, Montreal, Canada, H4A 3J1. E-mail: fatemeh.jafarian@mcgill.ca.
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corticosteroids. Significant improvement was noted at 6-day follow-up with resolution of pustules and decreased erythema. At the 2-week follow-up, the patient still had a low-grade fever and a red and fissured lip with a strawberry tongue. Striking periungual desquamation appeared on the fingers and toes (Fig 3). The patient lacked cervical adenopothy and conjunctivitis. Laboratory tests found that the erythrocyte sedimentation rate and platelet count were elevated.

In light of the new clinical findings and the reactive thrombocytosis, atypical KD was diagnosed. Fortunately, electrocardiography and echocardiography did not detect any cardiovascular abnormalities. Given the delay in the diagnosis of KD, the patient was not a candidate for intravenous immunoglobulin and was treated with low dose salicylic acid.

DISCUSSION

KD is a systemic vasculitis of unknown etiology. It is a pleomorphic disease and can have varying clinical findings including atypical cutaneous lesions.3 Recently, several published case reports reported simultaneous presentation of KD and psoriasis in a pediatric population with no history of psoriasis.4-9

Since the first case of KD associated with psoriasis was published in 2000 by Han et al, 31 were described.6 Various forms of psoriasis, including plaque, guttate, and pustular psoriasis are reported to occur during different phases of KD. To date, including this report, 6 cases of KD associated with pustular psoriasis have been published (Table I).4,5,7,9 Five of the 6 patients had pustular type psoriasis in the acute phase of KD. In contrast to pustular psoriasis, most cases of plaque psoriasis associated with KD are observed in the convalescent phase of KD. In all patients, psoriasis resolved after several months, possibly in accordance with the down-regulation of proinflammatory cytokines and...
T-cell response. Given the significant difference in the timeline of the onset of psoriatic eruptions, we postulate that different inflammatory mechanisms are involved in patients presenting with pustular versus plaque-type psoriasis in association with KD. This report presents a novel case of atypical KD initially presenting solely with fever and acute onset pustular psoriasis.

Two models potentially explain the pathophysiologic association of KD and psoriasis. The first hypothesis suggests a common pathogenic role of superantigen-producing bacteria in the activation of KD and psoriasis. The second theory proposes that psoriasis is uncovered in susceptible patients because of the localization of antigen-independent activated T cells to the cutaneous tissue in individuals with KD. However, these hypotheses do not explain the variable time course of presentation of psoriatic eruptions in different individuals. Further research is required to identify the exact pathophysiology that links KD and psoriasis.

Given the increasing prevalence of incomplete KD, the current diagnostic criteria should be viewed as a guideline, and a new diagnostic tool that incorporates both clinical and laboratory values is required to improve the diagnosis of KD. A high index of suspicion is important because all children with untreated KD, regardless of their clinical presentation, are at high risk of cardiac complications. This case report shows that pustular psoriasiform eruptions can be an initial sign of KD. Pustular psoriasis is an uncommon presentation of KD; however, KD should be considered as part of the differential diagnosis for patients presenting with pustular psoriasis in the context of prolonged fever.

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