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Annular Pustules in Kawasaki Disease: A Further Case Indicating the Association With Psoriasis?

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We report the case of a 4-year-old girl with Kawasaki disease (KD), or mucocutaneous lymph node syndrome, who presented with an annular pustular eruption. Targetlike erythematous and scaly patches were observed after resolution of the pustules. A biopsy of the skin was performed, and results showed spongy pustules not associated with the intraepidermal eccrine duct. Generalized pustular eruption, including pustular psoriatic lesions, has been described in KD. However, to our knowledge, this is the first report of annular pustular eruption mimicking annular pustular psoriasis in KD.

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Awasaki disease (KD) is an acute systemic vasculitis of unknown etiology that frequently affects children younger than 5 years.^{1,2} Diagnostic criteria for KD presented by the American Heart Association include fever of 5 or more days' duration, without other explanation, and at least 4 of the following 5 criteria: (1) bilateral nonexudative conjunctival injection; (2) injected or fissured lips, injected pharynx, or red strawberry tongue; (3) polymorphous exanthema; (4) acute

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nonsuppurative cervical lymphadenopathy; and (5) acral eruption.² Cutaneous manifestations of KD are polymorphous and include macules, papules, urticaria, scarlatiniform erythroderma, morbilliform rash, and even targetlike lesions that resemble erythema multiforme.¹ Extensive small pustules in KD have rarely been described in the English literature.³⁻⁵ We report a case of KD with an unusual annular pustular manifestation.

Case Report

A 4-year-old girl was referred to our department with a 5-day history of intermittent fever, sore throat, and cervical lymphadenopathy. The findings from the physical examination revealed small pustules in an annular arrangement on the trunk and extremities (Figure 1). Three days later, the pustules resolved with annular targetlike scaling and mild erythematous patches (Figure 2). In addition, fissures of the lips, nonpurulent conjunctivitis, and swelling of the hands and feet were observed. Laboratory studies yielded the following values: hemoglobin level, 9.0 g/dL (reference range, 12.0–16.0 g/dL); platelet count, 447×10³/µL (reference range, $150-400 \times 10^{3}/\mu$ L); white blood cell count, 33.8×10^{3} /µL (reference range, 3.5– 11.0×10^{3} /µL); antinuclear antibody, 1:160 (nucleolar pattern); and antistreptolysin O, 287 IU/mL (reference range, <200 IU/mL). A diagnosis of KD was made. No microorganisms were cultured from the pustules.

Microscopic examination of a skin biopsy specimen revealed multifocal, subcorneal, and intraepidermal spongy pustules unrelated to the intraepidermal eccrine duct in serial sections.

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Figure 1. Multiple annular pustules on the upper limb.



Figure 2. Targetlike erythema resembling erythema multiforme appeared after the pustules resolved.

Treatment with a single dose of intravenous immunoglobulin G (32.5 g) and high-dose aspirin (1300 mg/d) achieved an excellent response, with rapid resolution of the fever and the pustules. Six months later, 2-dimensional cardiac sonography did not detect dilation of the coronary artery.

Comment

KD, a disease of unknown etiology, occurs predominantly in infants and children younger than 5 years.¹⁻³ Clinical manifestations of KD include a polymorphous exanthema, such as urticarial plaques, maculopapular or morbilliform exanthema, targetlike lesion (erythema multiforme–like), erythema marginatum, and diffuse scarlatiniform erythroderma.¹ Generalized pustular eruption has rarely been reported in KD.³⁻⁸ However, in a Japanese study, 24 of 372 patients (6.5%) with KD developed generalized pustules during the course of the disease.⁸ The pustules appeared primarily on the knees, buttocks, and thighs; persisted for 1 to 5 days; and were arranged irregularly. Microscopic findings of the biopsy specimens showed subcorneal and spongy pustules in the epidermis and mild perivascular

mononuclear cell infiltration in the dermis. Because of the high incidence, the Japan Kawasaki Disease Research Committee included small pustules as an additional cutaneous manifestation in their revised diagnostic guidelines for KD in 1984.⁹

Our case is unique in the annular presentation of the pustules, which, when accompanied with the scaling patches and microscopic features, suggests the diagnosis of annular pustular psoriasis. Psoriatic eruptions, including 8 cases with typical scaly patches and 3 cases with generalized pustules, have been recently described in patients with KD.^{5,10} The 2 diseases were thought to be related because of the low incidence of psoriasis in infancy and early childhood.⁵ The annular pustular eruption mimicking pustular psoriasis in our case may provide additional evidence of the relationship between the 2 diseases.

Some investigators postulated that psoriasis might be "uncovered" through profound cytokines released in KD.⁵ However, no cases of patients developing chronic psoriasis after the convalescent phase of KD have been reported. We believe that the psoriatic eruption of KD may not represent true psoriasis but a psoriasislike cutaneous reaction. In addition, the subsequent targetlike erythema and only slight scaling are consistent with the reported erythema multiforme–like eruption of KD rather than the usual pustular psoriasis.

In conclusion, our case provides more evidence of the recently reported link between psoriasis and KD. The clinical and pathologic features of KD are similar to annular pustular psoriasis. Studies on the immunologic abnormalities of KD and pustular psoriasis may help clarify the etiology of the psoriasislike eruption in KD.

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