Atypical desquamation in a 2.5-year-old boy with Kawasaki disease: A case report

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Abstract
Kawasaki disease (KD) is a vasculitis that mostly affects children under 5 years of age. This article presents a 2.5-year-old boy who presented with 6 days of fever, generalized maculopapular rash, bilateral non-exudative conjunctivitis, cracked lips, right cervical lymphadenopathy, erythematous extremities, and perianal desquamation. Laboratory studies showed leukocytosis and sterile pyuria. Because diagnosis of KD was proved, oral acetylsalicylic acid with the anti-inflammatory dose and intravenous immunoglobulin were started for him. On the seventh day of admission time, he developed desquamation and erythema on the site of his right cervical lymphadenopathy as well as periungual scaling. About three weeks after starting the treatment, scaling of the cervical lymphadenopathy and periungual area stopped. Echocardiography was performed for him three times: at the time of diagnosis, four weeks, and 6 months later and revealed normal coronary arteries. We report this sign, desquamation on the site of cervical lymphadenopathy, as a new finding.

Keywords: Kawasaki disease, Cervical lymphadenopathy, Desquamation

1. Introduction
Kawasaki disease (KD) is a vasculitis of medium-size vessels. It was first described by Tomisaku Kawasaki in 1967 as infantile febrile mucocutaneous lymph node syndrome (1). The etiology of KD is relatively unknown, but an infectious insult in genetically predisposed patients has been found to play a role in this disease (2, 3). Innate and adaptive immune systems have been found to be involved in KD (4). It usually affects children under the age of 5 years old (5). KD is mostly a disease in Asian countries (6). There is no specific laboratory test for it, and its diagnosis is mostly based on its clinical criteria (7). KD has been responsible for acquired heart diseases in children of developed countries (8). Coronary artery aneurism is the most dangerous complication of KD (7). Diagnosis of KD is based on its clinical manifestations, which includes a high-grade fever that lasts five days with four of the following criteria: mucosal changes (erythema, strawberry tongue), bilateral non-exudative conjunctivitis, cervical lymphadenopathy, polymorphous systemic rash, and extremity changes (9). Extremity changes include erythema and edema of hands and feet in acute phase. This article presents a boy who had KD with a rare finding, which was desquamation on cervical lymphadenopathy in the subacute phase of the course of his disease.
2. Case presentation

2.1. History
A 2.5-year-old boy was admitted to Namazi Hospital of Shiraz in June 2014 with a history of high-grade fever from 6 days prior to admission. He had received acetaminophen, but no improvement was achieved. After 48 hours, he had developed generalized rash and conjunctivitis.

2.2. Clinical presentations
He was febrile on admission time (37.9 °C axillary). On physical examination, generalized maculopapular rash was found. He had right cervical lymphadenopathy (4 cm × 2 cm), bilateral non-exudative conjunctivitis, erythematous tympanic membrane, strawberry tongue, and dry cracked lips (Figure 1). His hands and feet were erythematous. Also, perianal desquamation was found.

Figure 1. Cracked lips and strawberry tongue.

2.3. Laboratory findings
On admission time, hematological studies showed leukocytosis: white blood cell count of 19700/mm³ (57% neutrophils, 34% lymphocytes, 9% mixed), hemoglobin of 11.9 g/dl, red blood cell count of 480000/mm³, mean corpuscular volume of 79 fl, and a platelet count of 406000/mm³. Erythrocyte sedimentation rate was 52 mm/h and C-reactive protein level was 34 mg/dl (negative<6 mg/dl). Urine analysis and urine culture were performed, and sterile pyuria was found (WBC: 25-30 and negative culture).

2.4. Treatment
Diagnosis of Kawasaki disease was proved by clinical criteria supporting laboratory findings. He received a slow infusion of a single dose of intravenous immunoglobulin (IVIG; 2 g/kg). Also, oral acetylsalicylic acid with the anti-inflammatory dose (80 mg/kg/day) was started for him.

2.5. Outcome
Echocardiography was done, showing normal coronary arteries and normal structure and function of the heart. Electrocardiogram was normal. After seven days of admission, he developed desquamation and erythema on the site of his cervical lymphadenopathy (Figure 2). Periungual desquamation was found, too. Three weeks after starting treatment for him, scaling on the site of the right cervical lymphadenopathy was stopped, but mild erythema persisted (Figure 2). Also, desquamation of his periungual area was stopped about three weeks after treatment was started.

2.6. Follow-up
He was followed-up with, and after one month of treatment, the erythema of his neck disappeared. His disease was clinically improved. Laboratory investigations were as follows: white blood cell count of 12000/mm, hemoglobin of 12.7 g/dl, and platelet count of 392000/mm³. Erythrocyte sedimentation rate was 18mm/h. Echocardiography was
done for him again at that time and showed no coronary artery disease. About six months later, echocardiography was repeated for him, and it was normal.

![Figure 2](http://www.ephysician.ir)

**Figure 2.** A. desquamation and erythema on the site of right cervical lymphadenopathy. B. disappearing of scaling and persistence of erythema.

3. **Discussion**

KD is an immunologic-based disease. KD may be difficult to diagnose when the patient presents with atypical manifestations. Also, when the patient does not fulfill all KD diagnostic criteria, he/she may become misdiagnosed (10). Desquamation in KD is expected on the perianal area at early stages of the disease period or on the periungual area after 2–3 weeks of the onset of fever (6). Our patient presented with scaling on the site of his right cervical lymphadenopathy as well as the mentioned routine sites. Shredding-type desquamation can be associated with disorders such as scarlet fever, leptospirosis, staphylococcal toxic shock syndrome, recurrent perineal erythema, and drug eruption (11). However, based on the clinical manifestations, laboratory findings, and the response to treatment, the only diagnosis for our patient was KD. Atypical sites of desquamation in KD have rarely been previously reported. One was a 21-year-old male Caucasian patient who presented with a diffuse maculopapular and erythema multiform-like eruption, non-exudative bilateral conjunctivitis, injected lips, strawberry tongue, and symmetrical widespread lymphadenopathy. He developed scaling of his face as well as his hands about 6 days after diagnosis (12). Another one was a 5-year-old girl presenting with the clinical manifestations of KD. She had developed periungual desquamation and fine scaling, on the skin over the dorsum of her feet (13). However, desquamation of the face or dorsum of her feet seemed to be an extension of the scaling of lips or periungual area, respectively. According to the fact that diagnosis of KD is mainly based on its clinical criteria, any validated novel clinical sign, which is associated with KD, can help us with early diagnosis. Based on our PubMed search, no previous article described a case with desquamation on the site of cervical lymphadenopathy as an associated sign.

4. **Conclusions**

Our patient presented with an atypical site of desquamation. We report this sign as a new finding. It is possible that, in the future, cervical lymphadenopathy becomes accepted as a site of desquamation beside the periungual and perianal area.

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**Conflict of Interest:**
There is no conflict of interest to be declared.

**Authors’ contributions:**
All authors contributed to this project and article equally. All authors read and approved the final manuscript.
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