Case Report

Unusual Presentation of Pustular Eruption in Kawasaki Disease Shock Syndrome: A Case Report

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The present study case report described a 46-month-old girl with Kawasaki disease shock syndrome (KDSS) who presented with five days of fevers, unilateral cervical lymphadenitis, pustular eruption, maculopapular rashes, erythema of palms and soles, conjunctivitis, cracked lips, and shock. Laboratory results showed elevated ESR & CRP, leukocytosis, normocytic anemia, and transaminitis. Pustular eruption Gram and Wright stains demonstrated numerous neutrophils. Echocardiogram showed normal results. Fluid resuscitation, broad spectrum antibiotics, inotropic drug, IVIG, and high dose aspirin were given. Diagnosis of Kawasaki disease was supported by clinical and laboratory features at the acute phase, in conjunction with periungual peeling of fingers and toes and thrombocytosis at the subacute phase. The patient made a complete recovery.

The present study case showed an unusual pustular eruption in KDSS. Clinicians should consider these presentations to the diagnosis of KDSS and timely prescribed IVIG, to prevent coronary artery aneurysm.

Keywords: Pustular eruption; Kawasaki disease shock syndrome

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Shock is an unusual presentation during the acute phase of Kawasaki disease (KD), however, "Kawasaki disease shock syndrome" (KDSS) has been studied more recently. The characters of KDSS include atypical presentation, difficulty for early recognition, more severe inflammatory markers, intravenous immunoglobulin (IVIG) resistance, and severe coronary artery involvement[1-9]. KDSS patients were also reported to have more serious skin rashes[9].

Cutaneous manifestations of KD are variable. Pustular eruption is an atypical presentation[10-16]. The present report showed a KDSS patient with the unfamiliar presentation of both shock and pustular eruption. To prevent coronary artery damage, a diagnosis of KDSS and definitive treatment with IVIG should not be delayed[10].

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Case Report

The present case occurred in November 2018 at Burapha University Hospital in Chonburi, Thailand. The case report was approved by the Burapha University Institutional Review Board, approval number 025/2563.

A 46-month-old girl presented with high-grade fever, a painful right neck mass, red eyes, and maculopapular and pustular rashes for five days. Four days before the hospital visit, she was treated for the right swollen neck by ceremonial traditional medicine, written magic alphabets at the neck mass, and was given amoxicillin. Later, there were red rashes spreading from her neck to her trunk, hands, and feet. The other symptoms were ongoing with an appearance of pustular eruptions, involving swollen neck mass, submandibular area, and neck flexure (Figure 1). Two hours after admission, she developed hypotension as 70/40 mmHg, tachycardia at 130 per minute, and weak pulse. Two doses of normal saline solution (NSS) of 20 mL/kg over 15 minutes, intravenous cefotaxime plus clindamycin and dopamine of 10 mcg/kg/minute were initiated as a septic shock treatment. During physical examination, she had fever at 38.4°C and looked very lethargic. The right anterior cervical lymph node was painful, enlarged to 4×4 cm^2, not fluctuated, and covered with erythema and multiple tiny micropustules. Multiple tiny pustules were also involving the entire neck, neck flexure, and confluent

Table 1. Laboratory results on different days after disease presentation

Day of disease	Peak temp	ESR (mm/hour)	CRP (mg/L)	Hb (g/dL)	WBC (/mm^3)	Neutrophils/ lymphocytes (%)	Platelets (/mm^3)	AST/ALT (U/L)	Albumin (g/dL)
Day 5	38.4	120	200.4	10.8	21,980	93/4	258,000	32/70	2.6
Day 10	37.3	72	32	10.2	16,400	62/31	690,000	23/40	3.3

ESR=erythrocyte sedimentation rate; CRP=C-reactive protein; Hb=hemoglobin; WBC=white blood cell; AST=aspartate aminotransferase; ALT=alanine aminotransferase

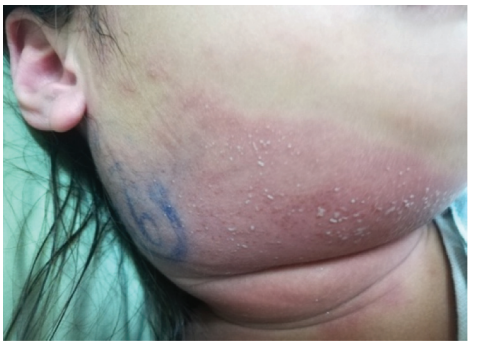

Figure 1. Representative clinical picture of pustular eruption in KDSS patient; micropustular rashes overlying cervical lymphadenitis and both sides of the neck.

at gluteal folds. Multiple erythematous papules, macules, and 0.5 to 1.5 cm patches were discrete on the trunk, bilateral symmetrical on palms, soles, axillae, antecubital fossae, popliteal fossae, and perineum, with generalized mild itching erythema on the back. Excoriation marks appeared at the lower back. Bilateral bulbar conjunctivitis without discharges, cracked lips, and strawberry tongue were found. BCG vaccination site at the left shoulder was not erythematous or enlarged. No other systemic organs were involved.

Pus from pustules at neck area was collected for Gram and Wright stain and showed sterile pustules with white blood cell infiltration of 30 to 50 per oil power field, 100% neutrophils. Tzanck smear showed no multinucleated giant cells. Laboratory results are shown in Table 1. She also had hyponatremia with 129 mmol/L sodium, hypokalemia with 3.2 mmol/L potassium, and metabolic acidosis with 12 mmol/L HCO_3. Cardiac enzymes, coagulation studies, kidney function tests, urinalysis, and chest radiograph appeared normal. No bacteria grew from pus and blood cultures. Serology for Measles, Rubella, and Epstein-Barr virus turned negative.

Eventually, the presentation of classic features of mucocutaneous lymph nodes and laboratory test results supported KD, so the diagnosis of KDSS was made. IVIG at 2 g/kg and aspirin at 100 mg/kg/day were given. Her symptoms rapidly improved, fever subsided, vital signs were stable. At day 3 after IVIG, the pustular rashes and erythematous rashes disappeared and the cervical lymph node gradually reduced in size. Echocardiographic examination was normal. The diagnosis was confirmed by the presentation of periungual desquamation and thrombocytosis 10 days after first symptoms (Table 1). Beau's line appeared 14 days after first symptoms. Aspirin was reduced to 5 mg/kg/day. Echocardiographic result at week 8 was normal, and aspirin was discontinued. The patient had been followed for two years without relapse of pustular eruption or other complications.

Discussion

KDSS has recently been identified and is exceedingly rare. The atypical presentations make diagnosis challenging. In KD, the pustular eruption is a rare presentation. In KDSS, no previous case of pustular rash has been reported. The patient presented with pustular rash, shock, and multisystem involvement in critical states. Although difficult to diagnose, urgent diagnosis and treatment are necessary.

This is the first case report of pustular rashes in KDSS. The patient was diagnosed based on her symptoms and was treated until recovery. KDSS is a severe condition of KD. In 2009, Kanegaye et al first introduced the term "KDSS"[1]. Since then, many studies on KDSS reviewed clinical features that show multisystem involvement and atypical presentations, such as more serious skin rashes, abdominal pain, and neurological symptoms. These were more severe inflammatory markers, high neutrophil:lymphocyte ratio, anemia, and hypoalbuminemia. KDSS patients required an inotrope treatment. They also had a higher risk of IVIG resistance and coronary artery aneurysm[1-9].

Pustular rashes were occasionally reported in KD[11-16]. They appeared on the erythematous areas and symmetrically arranged on the extensor surface of the extremities, axillae, genital area, and buttocks[13,14].

Some KD patients with pustular rashes required two doses of IVIG[14,15]. It is unknown if atypical serious skin manifestation may reflect more severe inflammatory process of KD.

After searching the literature with appropriate keywords, the authors did not find any pustular eruption reported in KDSS, although Ma et al had demonstrated that most KDSS patients had more serious skin rashes, as scarlatiniform rash, BCG erythema, and erythema annulare[9]. The present case report emphasized on pustular rash presentation in KDSS, the authors found a distinct distribution of pustular rashes involving first on the enlarged cervical lymph node, then neck flexure and gluteal fold with widespread erythematous maculopapular rashes. No IVIG resistance and no coronary artery aneurysm occurred. It is unknown if atypical serious skin manifestation may reflect more severe inflammatory process of KD. In the present case, sterile pustular eruption, which is an uncommon presentation in KD, may lead to recognition of the serious subtype of KD, KDSS.

The important differential diagnosis of wide-spread erythematous rashes with shock is Toxic shock syndrome (TSS). Antibiotics were started to cover TSS. The clinical presentations after the follow up on the fourteenth day of the disease showed periungual desquamation and thrombocytosis (Table 1), confirming the diagnosis of KD. The clinical presentations of KDSS and TSS are quite similar. Pustular eruption is not usually found in either TSS or KDSS. Unilateral cervical lymphadenopathy in an acute phase, characteristic periungual desquamation, and thrombocytosis in a subacute phase of KDSS are helpful to differentiate KDSS from TSS. The non-infectious diseases of sterile pustular eruption with fever were differentially diagnosed as Acute Generalized Exanthematous Pustulosis and generalized pustular psoriasis.

In KDSS patients, pustular rash can be a cutaneous manifestation. Clinicians should consider this presentation to avoid delays in diagnosis and treatment. The presence of periungual peeling of the fingers and toes, as well as thrombocytosis in the subacute phase, aid in the diagnosis of KD.

Conclusion

A 46-month-old girl with KDSS presented with pustular eruptions on cervical lymphadenitis, the entire neck, and the gluteal folds with maculopapular rashes, together with shock and all clinical findings that match the diagnostic criteria for KD. She responded well to IVIG, shock rapidly improved, no recurrent symptoms occurred, and echocardiogram results were normal. The pustular rash is an unusual presentation in KD. KD presented with shock is also unfamiliar to doctors. It is thus, difficult to make a correct diagnosis at the early stage, delayed treatment with IVIG may result in permanent coronary damage.

What is already known on this topic?

KDSS patients frequently exhibited more multisystem involvement and atypical presentation, including more serious skin rashes. Pustular rash is rarely seen in KD. The presentation of shock, which is indicative of KD, is also unfamiliar to pediatricians.

What this study adds?

This is the first case report of pustular rashes in KDSS.

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Conflicts of interest

The authors declare no conflict of interest.

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