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Case Series

Cutaneous clues to Kawasaki disease: observations from seventeen pediatric patients

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ABSTRACT

Kawasaki disease (KD) is an acute pediatric vasculitis and a leading cause of acquired heart disease. Cutaneous manifestations are central to its diagnosis, yet variability in rash morphology and distribution may delay recognition, particularly in incomplete cases. Objective of the study was to describe the frequency, morphology, and distribution of cutaneous manifestations in pediatric patients with KD at a tertiary care center in Puerto Rico. We conducted a retrospective case series of pediatric patients diagnosed with classic or incomplete KD at Hospital Episcopal San Lucas in Ponce, Puerto Rico, between January 2019 and December 2024. Patients meeting American Heart Association diagnostic criteria were included. Clinical data focused on rash morphology, distribution, timing, and desquamation. Seventeen patients were included. All patients presented with fever. A polymorphous rash was observed in 15 patients (88.2%), most commonly morbilliform in appearance (82.4%). One patient exhibited a scarlatiniform rash, and two patients (11.8%) did not develop a rash. Rash involvement most frequently affected the trunk and extremities (64.7%), followed by the face (41.2%). Inguinal or perineal involvement occurred in 17.6% of cases. Desquamation was documented in 41.2% of patients, predominantly as periungual peeling during the subacute phase. Twelve patients (70.6%) met criteria for complete KD, while five (29.4%) had incomplete presentations. Cutaneous manifestations were common and largely consistent with classic KD patterns. Recognition of early or subtle dermatologuc findings is essential particularly in incomplete KD to enable timely diagnosis and reduce coronary complications.

Keywords: Kawasaki disease, Rash, Pediatric dermatology, Mucocutaneous findings

INTRODUCTION

Kawasaki disease (KD) is an acute vasculitis of childhood and the leading cause of acquired heart disease in children in developed countries.[1] It classically presents with prolonged fever and a constellation of mucocutaneous findings.[1] Among its principal clinical criteria is a polymorphous skin rash, which occurs in the majority of cases. In fact, approximately 90% of children with KD develop an erythematous polymorphous exanthem which typically appears within the first 5 days of illness.[2] The rash is usually diffuse and maculopapular, giving a measles like or viral exanthem appearance.[1] It may initially be mistaken for a viral infection or drug eruption, however, the persistent fever along with other signs should raise suspicion of KD.[1]

The rash is classically described as polymorphous because it can present in several forms varying from macular to maculopapular, morbilliform, or scarlatiniform.[1] Importantly, bullous, vesicular, or petechial rashes are not typical in KD and should prompt consideration of alternative diagnoses.[3] In terms of rash distribution, it is usually first noticed in the trunk and then spreads to involve the extremities within days.[4] Facial involvement can occur, though often is less pronounced than on the

torso. When the rash involves the face, it tends to be around the eyes, nose and extending around the mouth, which forms a distinctive "T-shape".[5]

Moreover, patients' rash may involve the groin/perineal region which frequently undergoes early desquamation by the end of the first week, this is a clinical feature of KD which can be easily overlooked in the clinical evaluation, unless meticulously looked for.[6] Additionally, the palmoplantar erythema is part of the classic "extremity changes" which often includes swelling of hands and feet.[1] Often the latter symptoms occur in the acute phase but later in the subacute phase patients present with periungual peeling of the fingers and toes.[1] These cutaneous characteristics, especially when combined with KD's other features such as conjunctival injection, oral changes, extremity changes including lymphadenopathy, for the cornerstone of clinical diagnosis.[1]

Early recognition of these skin manifestations is crucial for timely diagnosis and management of KD. Skin rash can appear as early as in the first stage which is acute along with fever and conjunctivitis.[1] Prompt diagnosis allows for early treatment, typically with high-dose-intravenous immunoglobulin (IVIG) and aspirin, which if given ideally within the first 10 days of illness can reduce the risk of permanent coronary damage by about 75%.[7] Therefore, early recognition of the KD rash along with fever and mucosal changes enables physicians to initiate therapy before cardiac sequelae develop. In summary, understanding the frequency, morphology, and distribution of Kawasaki rash has direct implications for patients' outcomes as it serves as an invaluable diagnostic clue that should prompt immediate evaluation.

CASE SERIES

A total of 17 pediatric patients were included in the case series. The median age at presentation was approximately 1.8 years, with ages ranging from 10 months to 4 years 7 months. The majority of patients (15 out of 17) were under 4 years old. There was a notable male predominance, with 13 males and 4 females (76% and 24%, respectively).

All patients presented with fever as an initial symptom. The classic clinical features of KD were observed in most cases: a polymorphous rash in 15 patients (88.2%), bilateral non-exudative conjunctival injection in 15 (88.2%), changes of the lips and oral mucosa (cracked lips or strawberry tongue) in (70.6%), peripheral extremity changes (hand/foot edema or erythema) in 13 (76.5%), and cervical lymphadenopathy in 9 (52.9%). Additionally, 12 patients (70.6%) met the criteria for complete KD, while the remaining 5 (29.4%) had incomplete presentations (having fewer and fewer than four of the classic criteria).

Of the 15 patients who develop a rash, the majority (14 patients, 82.4%) exhibited a morbilliform eruption. One case (5.9%) demonstrated a scarlatiniform (scarlet-fever-like) rash pattern. Only two patients (11.8%) did not have any rash during the course of their illness. No other distinctive rash morphologies were noted in this series.

The initial distribution of the rash varied. Truncal and extremity involvement were the most common, each observed in 11 patients (64.7%) at rash onset. Facial rash was present in 7 patients (41.2%). Less frequently, the rash affected flexural creases (axillae or groin) in 3 patients (17.6%), the genital (inguinal/perineal) region in 3 patients (17.6%), and acral areas (palms and soles) in 3 patients (17.6%).

In many cases the eruption was initially generalized (64.7%), while in other cases stayed localized (11.8%) or started localized and later became generalized (11.8%) or not reported (11.8%).

The rash typically appeared early in the illness course, often within the first week of symptom onset and as by the time of diagnosis a rash was present in the majority of cases.

Desquamation was documented in 7 patients (41.2%). This finding usually manifested in the subacute phase of the illness (generally during the second week after fever onset). Periungual peeling of the fingers and toes was presumed to account for most instances as 6 patients presented with it (35.3%). No patient was reported to present with nail changes.

Table 1: Clinical and cutaneous characteristics of pediatric patients with Kawasaki disease (n=17).

Variable	N (%)
Demographics	
Male	13 (76.5)
Female	4 (23.5)
Age <4 years	15 (88.2)
Kawasaki classification	
Complete	12 (70.6)
Incomplete	5 (29.4)
Mucocutaneous findings	
Rash present	15 (88.2)
Conjunctival injection	15 (88.2)
Oral mucosal changes	12 (70.6)
Extremity changes	13 (76.5)
Lymphadenopathy	9 (52.9)
Rash morphology	
Morbilliform	14 (82.4)
Scarlatiniform	1 (5.9)
Rash distribution	
Face involvement	7 (41.2)
Trunk involvement	11 (64.7)
Extremity involvement	11 (64.7)
Flexural creases	3 (17.6)
Inguinal/perineal	3 (17.6)
Acral (palms/soles)	3 (17.6)

DISCUSSION

Our case series highlights that cutaneous manifestations are a central feature of KD, with a polymorphous rash observed in 88.2% of patients, closely matching the 80–90% frequency reported in large studies.[8] The demographic profile of our cohort (median age ~1.8 years, 76% male) reflects the known epidemiology of KD, which predominantly affects children under 5 years of age and demonstrates a male predominance.[1] Nearly all patients exhibited mucocutaneous findings, with rash typically appearing within the first 5 days of illness.[1] Notably, 29% of patients had incomplete KD, meeting fewer than four classic criteria; among these, two did not develop a rash, highlighting how incomplete presentations may lack hallmark features and require a higher index of suspicion, often relying on laboratory criteria to support the diagnosis. Importantly, this study provides region-specific data from a Puerto Rican pediatric population and underscores the diagnostic value of cutaneous findings, particularly in incomplete KD, where clinical features may be subtle.[9]

Most patients (82.4%) exhibited a maculopapular eruption, recognized as the most common KD rash pattern.[10] Only one case (5.9%) exhibited a scarlatiniform (scarlet-fever-like) rash, a less frequently documented morphology in the literature, which made the diagnosis challenging. The eruption appeared within the first week of illness in all affected patients.[2] The rash most often began on the trunk and subsequently spread to the extremities.[10]

Importantly, inguinal/perineal rash was observed in 17.6% of cases, lower than the ~67% frequency reported in some studies.[8] This discrepancy may reflect underrecognition, as these subtle findings can be missed without focused examination. Additionally, desquamation was documented in 41.2% of patients, predominantly as periungual peeling of the fingers and toes during the subacute phase, with onset typically occurring 2–3 weeks after fever onset.[8] Although periungual desquamation most commonly involves both upper and lower extremities, in our series, only 3 patients had desquamation limited to the fingers. No nail changes were reported, which may reflect variability in documentation or timing of follow-up.

When comparing our findings to other case series and reviews, the cutaneous profile of our KD patients aligns well with established patterns. For example, a national cohort in Switzerland reported rash in 85.4% of cases, comparable to our own rates. In our series 5 patients had incomplete KD and only 2 lacked rashes. This reflects the fact that many patients with incomplete KD still manifest cutaneous signs.

Recognizing dermatologic findings is particularly critical in incomplete KD. Even transient or localized features, such as perineal erythema or periungual desquamation, should raise suspicion when persistent fever is unexplained. As emphasized by Gupta and Singh, thorough skin examinations are essential in suspected cases, especially when diagnostic criteria are not fully met.[3] In our series, one-third of patients initially had incomplete presentations, and some were misdiagnosed with viral exanthems before KD was confirmed.

The cutaneous findings in our Puerto Rican pediatric KD population closely resemble international patterns, with minor variations, such as nail findings and perineal rash, likely attributable to documentation or population differences. Although KD remains relatively rare, with an estimated incidence of 0.02%, we encountered a considerable number of cases between 2019 and 2025.[11] This underscores the importance of clinician awareness of KD's dermatologic features in Puerto Rico.

CONCLUSION

In conclusion, early recognition of the frequency, morphology, and progression of cutaneous manifestations in KD is essential for timely diagnosis and intervention. Patients with incomplete KD often still present with a maculopapular rash, which may aid in clinical recognition and management. Identification of these findings enables prompt initiation of IVIG and aspirin therapy, significantly reducing the risk of coronary complications. Careful skin examination, including less obvious sites such as the groin and periungual areas, may provide critical and often subtle diagnostic clues. Further prospective, multicenter studies in larger and more diverse populations are needed to better characterize the full spectrum, variability, and diagnostic implications of cutaneous manifestations in KD across diverse clinical settings.

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