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Images in Rheumatology

Perianal skin peeling: An important clinical pointer toward Kawasaki disease

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A 10-month-old boy presented with a high-grade fever for 4 days. He also had bilateral, firm, tender anterior cervical node enlargement that was more prominent on the right side (Figure 1). No other lymph node groups were enlarged. Dryness, fissuring, and redness of the lips were noted at the time of admission. Skin desquamation with erythema was also noted in the perianal area (Figure 2), and hence, Kawasaki disease (KD) was considered a strong clinical possibility. There was no evidence of redness of the eyes, rash, erythema of the oral mucosa, or edema of the extremities. His blood count tests revealed leukocytosis (34.8×10^9 cells/L) with neutrophilic predominance (56% polymorphs), anemia (hemoglobin, 79 g/L), and platelet counts of 106×10^9 cells/L. Erythrocyte sedimentation rate (ESR) was 32 mm (1st hour) and C-reactive protein was 179.2 mg/L. Serum albumin was 2.3 g/dL. Echocardiography showed brightness of the left main coronary and left anterior descending arteries. There was, however, no dilatation. Blood cultures were sterile and anti-streptolysin O titers were normal. Intravenous immunoglobulin (IVIg) (Meglob; Synergy Diagnostics Pvt Ltd, Thane, Maharashtra, India) at 2g/kg infusion and aspirin at 50 mg/kg/day were initiated 5 days after onset. There was prompt defervescence of fever following IVIg therapy. Serial blood counts revealed progressive rise in platelet counts (251×10^9 cells/L and 778×10^9 cells/L on days 8 and 11 of illness, respectively). The baby subsequently developed periungual peeling over the fingers on day 10 of illness.

The American Heart Association (AHA) epidemiological definition for KD requires the presence of fever for at least 5 days and the presence of four of the following five clinical features: extremity changes, exanthematous rash, bulbar conjunctival injection, cervical adenopathy, and changes in lips and oral cavity (1). Apart from fever, the child had only three clinical features of the AHA Criteria for KD, namely anterior cervical adenopathy, lip changes, and periungual peeling at the subacute phase. As per the evaluation of suspected incomplete KD, the index child had additional features such as perianal peeling, coronary artery abnormalities in the echocardiogram, CRP of ≥3 mg/L, and ESR of ≥40 mm/h (1). Supplemental laboratory features for KD in the child were leukocytosis, anemia, elevation in platelet counts ≥450,000/mm^3 after 7 days, and hypoalbuminemia (1). Moreover, the clinical course of the illness with the development of

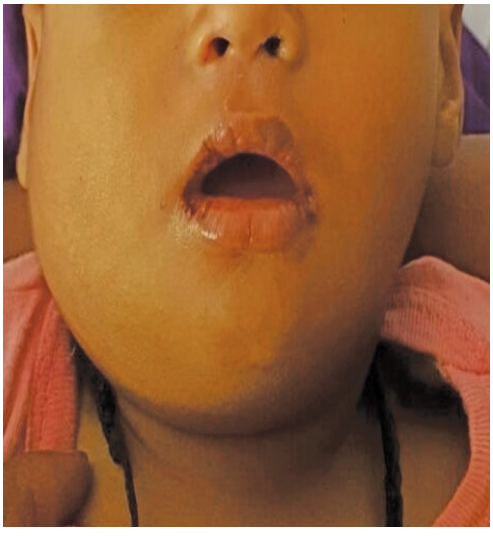

Figure 1. Cervical adenopathy in the child with Kawasaki disease (right > left)

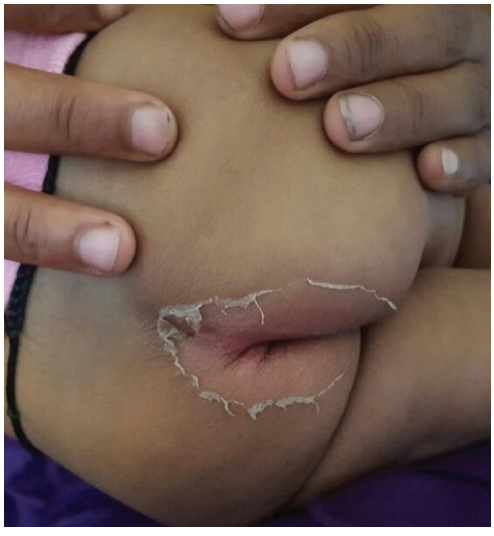

Figure 2. Circumferential perianal peeling with erythema noted in the perianal region in the index child

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typical periungual peeling and progressive rise in platelet counts observed in the subacute phase of illness strongly suggests KD (1). Incomplete presentation of KD is well known in infants and carries a high risk of coronary artery abnormalities (2).

Perineal desquamation, a key clinical feature of KD, can be easily overlooked in the clinical evaluation of febrile children, unless meticulously looked for (3). Perineal or perianal peeling may be noted as early as the 6th day of fever and occurs much earlier than the periungual peeling. The mechanism for desquamation in KD is not completely understood. Release of toxins or superantigens and excess cytokine production by the immune cells in the skin are some of the proposed hypotheses for the skin peeling (4). Perineal skin peeling is not a part of the AHA clinical criteria for the diagnosis of KD (1). Although differential diagnoses for fever and cervical adenopathy are many, perianal desquamation seen in the index child was a strong clinical indicator of KD and enabled us to promptly initiate IVIg (1). This report highlights the importance of perianal examination of a febrile child, in whom KD, an important medium vessel vasculitis that requires timely management, is suspected.

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