



Palatal Ulcer in Leukocyte Adhesion Deficiency: an Unusual Occurrence

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To the Editor,

A 5-week-old girl presented with swelling and redness around umbilicus. She was born of non-consanguineous marriage to a primigravida, was delivered at term, and was exclusively breast-fed. Her umbilical cord separated at 3 weeks of life. Physical examination revealed erythema around umbilicus with no discharge and a well-defined perforation over hard palate (Fig. 1). Investigations revealed total leucocyte count of $127.95 \times 10^9/L$ with 60% polymorphonuclear leucocytes. With history of delayed cord fall, presence of omphalitis and marked neutrophilic leukocytosis, clinical diagnosis of leukocyte adhesion deficiency (LAD) was considered. CD18 positivity on neutrophils was markedly reduced (1.48 vs 99.97% in control) and confirmed diagnosis of LAD.

Leukocyte adhesion deficiency is a primary immunodeficiency disorder (PID) with a defect in phagocyte function. Patients with LAD have polymorphonuclear leukocytosis, even in the absence of infections [1]. Absence of pus at site of infection is characteristic [2]. They may present with delayed cord separation, umbilical cord sepsis, and non-healing ulcers classically in perianal region. To the best of our knowledge, however, a palatal ulcer with perforation has never been reported in this condition.

Key message: Leukocyte adhesion deficiency can present with non-healing ulcers and the absence of pus is characteristic.

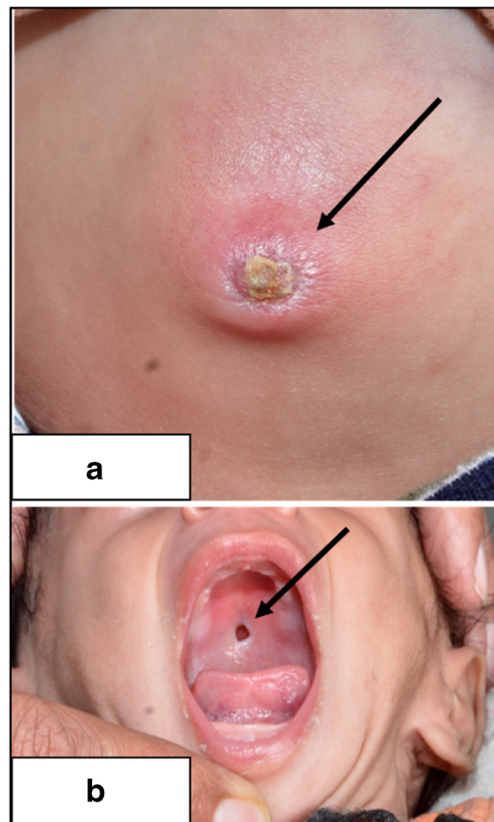


Fig. 1 a Figure showing redness and swelling around umbilicus. b Figure showing ulcer at the junction of the hard and soft palate

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Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

References

1. Etzioni A. Defects in the leukocyte adhesion cascade. *Clin Rev Allergy Immunol.* 2010 Feb;38(1):54–60.
2. van de Vijver E, van den Berg TK, Kuijpers TW. Leukocyte adhesion deficiencies. *Hematol Oncol Clin North Am.* 2013 Feb;27(1):101–16.