

## A Rare Case Report of Non-Langerhans Cell Cutaneous Histiocytosis in a 6-Month Old Infant

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**Abstract :** INTRODUCTION: Hemophagocytic lymphohistiocytosis (HLH) is a severe, potentially fatal syndrome in which there is excessive immune activation. The disease is seen in children and people of all ages, but infants from birth to 18 months are most frequently affected. HLH is a sporadic or familial condition that can be triggered by various events that disturb immunological homeostasis. In cases with a genetic predisposition and sporadic occurrences, infection is a frequent trigger. Because of the rarity of this disease, the diverse clinical presentation, and the lack of specificity in the clinical and laboratory results, prompt treatment is essential, but the biggest obstacle to a favorable outcome is frequently a delay in identification. CASE REPORT: Here we report a case of a 6-month-old male infant who presented to the dermatology outpatient with disseminated skin lesions present over the face, abdomen, scalp, and bilateral upper and lower limbs for the past month. The lesions were insidious in onset, initially started over the abdomen, and gradually progressed to involve other body parts. The patient also had a history of fever which was moderate in grade, on and off in nature for 1 month. There were no significant complaints in the past, family, or drug history. There was no history of feeding difficulties in the baby. Parents gave a history of developmental milestones appropriate for age. Examination findings include multiple well-defined monomorphic erythematous papules with a central crater present over bilateral cheeks. Few lichenoid shiny papules present over bilateral arms, legs, and abdomen. Ultrasound of the abdomen and pelvis showed mild degree hepatosplenomegaly, intraabdominal lymphadenopathy, and bilateral inguinal lymphadenopathy. Routine blood investigations showed anemia and lymphopenia. Multiple X-rays of the skull, chest, and bilateral upper and lower limbs were done and were normal. Histopathology features were suggestive of non-Langerhans cell cutaneous histiocytosis. CONCLUSION: HLH is a fatal and rare disease. A high level of suspicion and an interdisciplinary approach among experienced clinicians, pathologists, and microbiologists to define the diagnosis and causative disease are key to diagnosing this case. Early detection and treatment can reduce patient morbidity and mortality.

**Keywords :** histiocytosis, non langerhans cell, case report, fatal, rare

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