Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Dermatology

Langerhans Cell Histiocytosis in a Child: A Case Report

A. Moussa[1]*, H. Hassan[1], M. El Amraoui[1], Y. Zemmez[1], R. Frikh[1], N. Hjira[1]

[1]Department of Dermatology, Mohammed V Military Teaching Hospital, Rabat

DOI: https://doi.org/10.36347/sjmcr.2026.v14i05.017 | **Received:** 07.03.2026 | **Accepted:** 20.04.2026 | **Published:** 06.05.2026

***Corresponding author:** A. Moussa
Department of Dermatology, Mohammed V Military Teaching Hospital, Rabat

Abstract **Case Report**

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by an abnormal proliferation of Langerhans cells. We report the case of a 16-month-old girl presenting with diffuse papulo-crusted skin lesions associated with splenomegaly and hematological abnormalities. Histological examination and immunohistochemistry (CD1a, S100) confirmed the diagnosis. This case highlights the importance of considering LCH in the presence of atypical cutaneous lesions in children in order to ensure prompt and appropriate management.

Keywords: Langerhans Cell Histiocytosis, Pediatric Patient, Immunohistochemistry, Splenomegaly.

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by the abnormal proliferation of Langerhans cells. It is defined by the accumulation of these cells in various tissues and organs, leading to a wide spectrum of clinical manifestations. Despite extensive research, the etiology and pathogenesis of LCH remain poorly understood, making it a condition that can be challenging to diagnose and manage [1].

The peak incidence is observed in children aged 1 to 4 years, with a gradual decline thereafter [2–4]. However, the disease may occur at any age and has also been reported in elderly individuals [5,6]. It shows a slight male predominance, with a male-to-female ratio of approximately 3:2 [10,11].

The clinical presentation is highly variable, ranging from localized indolent lesions to multisystem disease [4]. Bone involvement is the most common manifestation (approximately 80% of cases), followed by skin involvement (33%). Extra-osseous involvement, particularly affecting the pituitary gland, liver, spleen, bone marrow, and central nervous system, is less frequent [2,7,8].

Cutaneous lesions are often the initial manifestation of the disease. They predominantly involve the scalp, retroauricular folds, external auditory canals, and intertriginous areas (particularly the diaper area and inguinal folds), as well as the trunk, sometimes showing a "vest-like" distribution. The limbs and face are less frequently affected.

Several dermatologic conditions may mimic the cutaneous manifestations of LCH, making the diagnosis challenging. These include seborrheic dermatitis, psoriasis, atopic dermatitis, and certain viral exanthems, due to their eczematous or papular appearance [9].

The diagnosis is based on a combination of clinical findings and is confirmed by histopathological examination with immunohistochemistry. Therapeutic management depends on the extent of the disease, its severity, and the organs involved.

In this report, we present a case of biopsy-confirmed Langerhans cell histiocytosis (LCH), initially presenting with crusted papules distributed in a "sleeveless vest" pattern in a 16-month-old child.

CASE PRESENTATION

A 16-month-old girl, born to a well-monitored full-term pregnancy and delivered vaginally, with no significant personal or family medical history, was admitted for evaluation of erythematous crusted skin lesions evolving for one month. The clinical course was associated with asthenia, absence of fever, and abdominal distension, without vomiting or bowel habit disturbances.

Physical examination revealed mucocutaneous pallor and multiple erythematous papular crusted lesions

of variable size, involving the scalp, retroauricular folds, trunk, abdomen, genital region (Figure 1), and back (Figure 2), with areas showing a "sleeveless vest" distribution. Some lesions were confluent, forming plaques, particularly on the scalp (Figure 3). Examination also revealed splenomegaly extending approximately two fingerbreadths below the costal margin.

Laboratory investigations showed anemia (hemoglobin: 5.3 g/dL), thrombocytopenia (platelet count: 83,000/mm³), and leukocytosis (13,000/mm³, with 72% neutrophils). C-reactive protein was elevated at 47 mg/L, and lactate dehydrogenase (LDH) level was 260 U/L.

Histopathological examination demonstrated a regular epidermis with a preserved stratum corneum. The superficial dermis showed a dense cellular infiltrate composed predominantly of histiocytic cells, associated with lymphocytes and plasma cells (Figure 4A).

Immunohistochemical analysis revealed positivity for anti-CD1a (Figure 4B) and anti-S100 protein (Figure 4C), confirming the Langerhans cell origin of the infiltrate. Imaging studies, particularly abdominal ultrasound, confirmed homogeneous splenomegaly.

Based on these findings, the diagnosis of Langerhans cell histiocytosis was established. The patient was referred to a pediatric hematology and oncology department for further evaluation and appropriate management.

Figure 1: Diffuse erythematous papules involving the trunk, abdomen, and genital region.

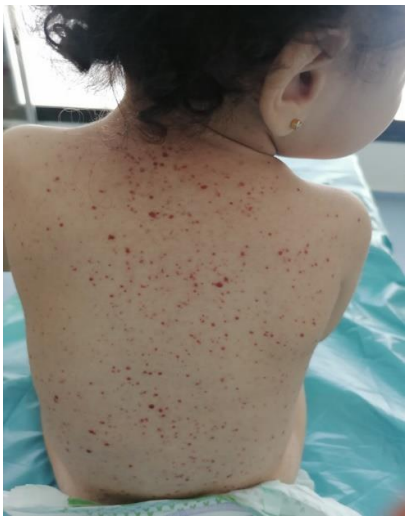

Figure 2: Diffuse erythematous crusted papules involving the back, displaying a "sleeveless vest" distribution, with sparing of the shoulders and limbs

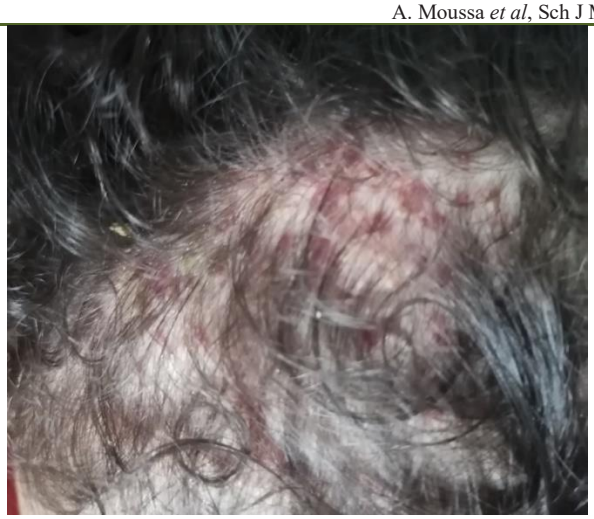

Figure 3: Erythematous scaly-crusted papules and plaques involving the scalp

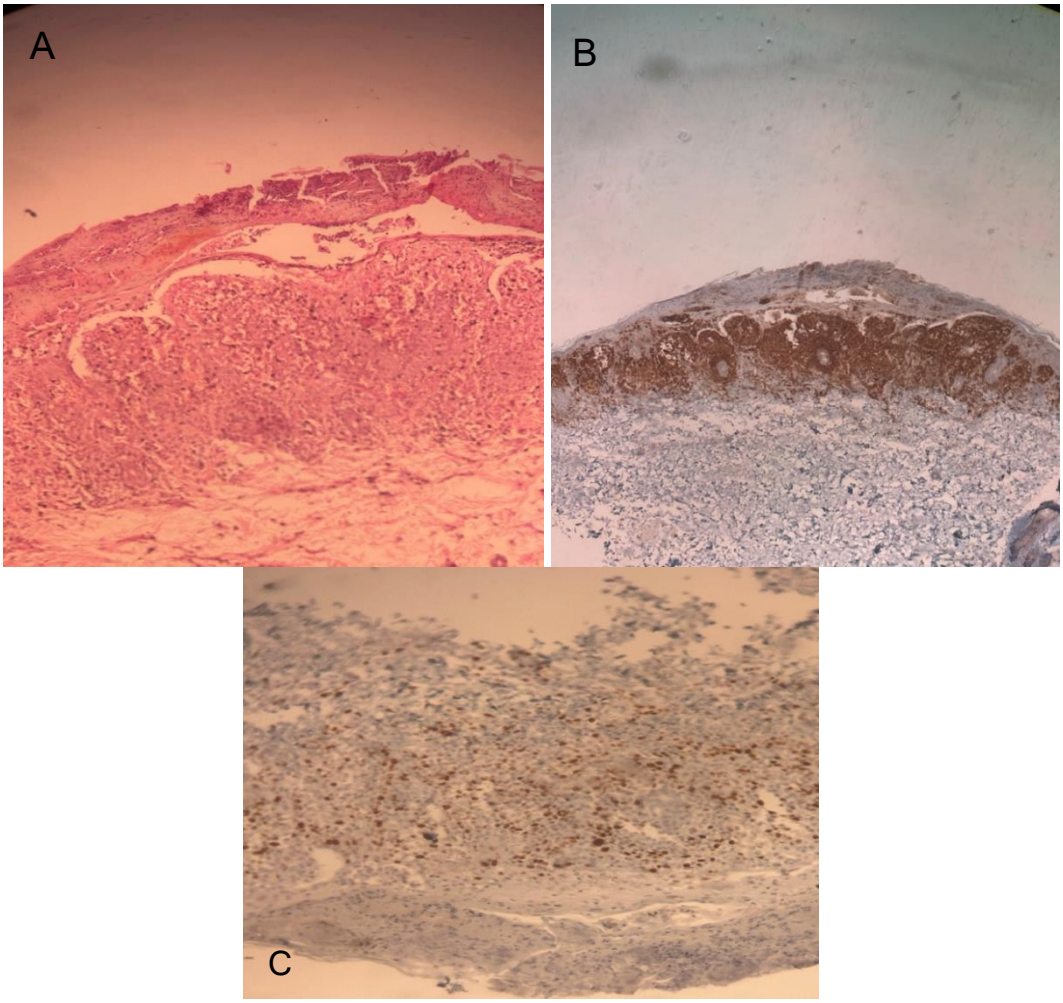

Figure 4: Histopathological examination showed a regular epidermis with a preserved stratum corneum. The superficial dermis exhibited a dense cellular infiltrate predominantly composed of histiocytic cells, associated with lymphocytes and plasma cells (A). Immunohistochemical analysis demonstrated positivity for CD1a (B) and S100 protein (C), confirming the Langerhans cell origin of the infiltrate

DISCUSSION

Langerhans cell histiocytosis (LCH) is an inflammatory neoplastic disorder that primarily affects infants and young children. It may present as a localized cutaneous disease or as a systemic condition with or without skin involvement [12]. The morphology of cutaneous LCH is highly variable and is frequently misdiagnosed as seborrheic dermatitis or eczema.

Although LCH can occur at any age, it predominantly affects the pediatric population [13].

Cutaneous involvement is observed in approximately 40% of patients with LCH [14]. The clinical manifestations are polymorphic and heterogeneous [15].

The most common presentations include widespread eruptions of closely grouped papules that may coalesce into scaly or brownish plaques, often covered with scales or crusts [14]. The most frequently affected areas are the scalp, retroauricular folds, external auditory canals, intertriginous regions (particularly the diaper area and inguinal folds), and the trunk, with a characteristic "vest-like" distribution [15]. The limbs and face are less commonly involved. The diagnosis is based on a combination of clinical findings and is confirmed by histopathological examination with immunohistochemistry [13,15].

The clinical course of LCH is unpredictable, according to the literature. Prognosis is closely related to the age at onset and the extent of organ involvement [9]. In very young children, involvement of the hematopoietic system—including the liver, spleen, and bone marrow—is common and is considered a poor prognostic factor [16].

Splenic involvement is relatively uncommon, occurring in approximately 15% of patients with LCH, and is mainly associated with multisystem disease [16]. The most characteristic radiological finding is splenomegaly, defined as an enlargement of the spleen extending more than 2 cm below the costal margin along the midclavicular line.

In our case, the patient presented with cutaneous lesions associated with anemia, thrombocytopenia, and homogeneous splenomegaly, which justified referral to a pediatric hematology and oncology department for further evaluation and appropriate management.

Conclusion

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by abnormal clonal proliferation of Langerhans cells. Its clinical manifestations are polymorphic, which may make the diagnosis challenging. When clinical suspicion arises, confirmation relies on histopathological examination complemented by immunohistochemical analysis.

The clinical course of the disease is unpredictable, warranting long-term, regular, and multidisciplinary follow-up to avoid overlooking potentially severe organ involvement, which may be life-threatening, particularly in children.

Therapeutic management depends on the organs involved, highlighting the importance of performing a comprehensive staging workup prior to treatment initiation.

References

1. Satter EK, High WA. Langerhans cell histiocytosis: a review of the current recommendations of the Histiocyte Society. Pediatr Dermatol. 2008 May-Jun ;25(3):291-5. doi : 10.1111/j.1525-1470.2008.00669. x.
2. Wang DS, Lu MY, Yang YL, Lin DT, Lin KH, Chang HH, Jou ST. Clinical outcomes of childhood Langerhans cell histiocytosis in Taiwan : A single-center, 20-year experience. J Formos Med Assoc. 2021 Jan ;120(1 Pt 3):594-601. doi: 10.1016/j.jfma.2019.12.019.
3. Gargan ML, Wong JL, Leidhin CN, Owens C, Byrne A. Langerhans cell histiocytosis in children under 12 months of age: The spectrum of imaging and clinical findings: Experience in an Irish tertiary referral centre. Eur J Radiol. 2021 Jan ;134 :109375. Doi : 10.1016/j.ejrad.2020.109375.
4. Krooks J, Minkov M, Weatherall AG. Langerhans cell histiocytosis in children: History, classification, pathobiology, clinical manifestations, and prognosis. J Am Acad Dermatol. 2018 Jun ;78(6):1035-1044. doi : 10.1016/j.jaad.2017.05.059.
5. Imanaka A, Tarutani M, Itoh H, Kira M, Itami S. Langerhans cell histiocytosis involving the skin of an elderly woman: a satisfactory remission with oral prednisolone alone. J Dermatol. 2004 Dec ;31(12):1023-6. doi : 10.1111/j.1346-8138. 2004.tb00648. x.
6. Stefanato CM, Andersen WK, Calonje E, Swain FA, Borghi S, Massone L, Kowalski JV, Bhawan J. Langerhans cell histiocytosis in the elderly: a report of three cases. J Am Acad Dermatol. 1998 Aug ;39(2 Pt 2):375-8. doi: 10.1016/s0190-9622(98)70395-0.
7. Stålemark H, Laurencikas E, Karis J, Gavhed D, Fadeel B, Henter JI. Incidence of Langerhans cell histiocytosis in children: a population-based study. Pediatr Blood Cancer. 2008 Jul ;51(1):76-81. doi: 10.1002/pbc.21504.
8. Laurencikas E, Gavhed D, Stålemark H, van't Hooft I, Prayer D, Grois N, Henter JI. Incidence and pattern of radiological central nervous system Langerhans cell histiocytosis in children: à population based study. Pediatr Blood Cancer. 2011 Feb ;56(2):250-7. doi :10.1002/pbc.22791.
9. Afonso C, Dias T, Teixeira C, Rodrigues D, Mira M. Cutaneous Manifestations of Langerhans Cell Histiocytosis in Pediatric Age : A Case Report. Cureus. 2024 Sep 25 ;16(9): e70141. doi: 10.7759/cureus.70141.
10. Guyot-Goubin A, Donadieu J, Barkaoui M, Bellec S, Thomas C, Clavel J. Descriptive epidemiology of childhood Langerhans cell histiocytosis in France,

2000-2004. Pediatr Blood Cancer. 2008 Jul ;51(1):71-5. doi: 10.1002/pbc.21498.

11. Yağci B, Varan A, Cağlar M, Söylemezoğlu F, Sungur A, Orhan D, Yalçin B, Akyüz C, Kutluk T, Büyükpamukçu M. Langerhans cell histiocytosis: retrospective analysis of 217 cases in a single center. Pediatr Hematol Oncol. 2008 Jun ;25(5):399-408. doi: 10.1080/08880010802107356.
12. Willman CL, Busque L, Griffith BB, Favara BE, McClain KL, Duncan MH, Gilliland DG. Langerhans'-cell histiocytosis (histiocytosis X) --a clonal proliferative disease. N Engl J Med. 1994 Jul 21 ;331(3) :154-60. doi: 10.1056/NEJM199407213310303.
13. Sobieraj J, Hatahet J, Menge T, Nasir W. Treatment of cutaneous Langerhans cell histiocytosis with ruxolitinib 1.5% cream. JAAD Case Rep. 2025 Jan 21 ;65 :171-173. doi: 10.1016/j.jdcr.2025.01.003.
14. Hussein MR. Skin-limited Langerhans' cell histiocytosis in children. Cancer Invest. 2009 Jun ;27(5) :504-11. doi: 10.1080/07357900802216452.
15. Luder CM, Nordmann TM, Ramelyte E, Mühleisen B, Kerl K, Guenova E, Dummer R. Histiocytosis - cutaneous manifestations of hematopoietic neoplasm and non-neoplastic histiocytic proliferations. J Eur Acad Dermatol Venereol. 2018 Jun ;32(6):926-934. doi: 10.1111/jdv.14794.
16. Cariello V, Lombardo P, Castelli L, Brillantino C, De Fusco C, Rossi A, Minelli R, Paviglianiti G, Grassi R, Rossi E. Integrated imaging of systemic Langerhans cell histiocytosis in an infant. Radiol Case Rep. 2022 Jun 2 ;17(8):2747-2753. doi: 10.1016/j.radcr.2022.05.016.