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Case and Review

This "*mite*" Surprise You: Scabies Masquerading as Langerhans Cell Histiocytosis – A Case Report

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Scabies · Langerhans cell histiocytosis · Pediatric dermatology

Abstract
Introduction: Scabies is a common parasitic infestation caused by the mite *Sarcoptes scabiei*. Scabies can mimic other entities clinically, resulting in misdiagnosis. The presence of a mite in the stratum corneum on biopsy specimens is diagnostic of scabies. However, there are instances when mites are not visible, and immunohistochemical (IHC) staining may be misleading. An example is when IHC demonstrates Cluster of Differentiation 1a and S100 positivity. The main differential diagnosis for this finding is Langerhans cell histiocytosis, a group of idiopathic disorders of bone marrow-derived Langerhans cells, with manifestations ranging from isolated to life-threatening multisystem disease. ***Case Presentation:*** We present a case of a patient who was diagnosed with Langerhans cell histiocytosis based on histological findings, further review with a repeat reading and deeper sectioning of her biopsy revealed a mite in the stratum corneum, altering the diagnosis, course, and management. She subsequently developed persistent post-scabietic nodules, an underreported entity that may occur following infestation and persist for up to a year. These lesions are self-limiting and do not require repeated courses of treatment. ***Conclusion:*** Langerhans cell hyperplasia may be seen in a multitude of entities, including scabies. Familiarity with this phenomenon is crucial to avoid unnecessary invasive investigations, aggressive management and alleviate patients' concerns.

Sara Al Janahi and Raghda Al Maashari contributed equally to this work.

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Introduction

Scabies is a common parasitic infestation caused by the mite, *Sarcoptes scabiei*. The classic clinical presentation is polymorphous lesions, including burrows, papules, and nodules with excoriations. Bullous presentations have also been reported [1]. Scabies is known to mimic common diseases clinically and has been misdiagnosed as a variety of entities, including Langerhans cell histiocytosis (LCH) [2–8], pityriasis rosea, and dermatitis herpetiformis [1, 9]. Misdiagnosis results in delayed resolution of the condition. However, the most important diagnosis to exclude is LCH as it carries the risk of systemic involvement and may have a poor prognosis, requiring more investigations (laboratory, radiologic) and significantly altering the course of management. There have been reports of children receiving systemic chemotherapy for LCH before ultimately being diagnosed with scabies. The two entities may overlap clinically, as LCH also has a variable presentation, including crusted, papulo-nodular lesions. The overlap may also be noted histologically, with both entities containing Cluster of Differentiation (CD)1a and S100 positive expressing cells. The presence of mites in the stratum corneum confirms the diagnosis of scabies. We present a case of a 3-month-old female, diagnosed with LCH based on clinical and histological findings. Repeat reading with deeper sectioning of her biopsy specimen revealed a mite in the stratum corneum, altering the diagnosis, course, and management. Appropriate anti-scabietic therapy resulted in resolution of her condition. However, she subsequently developed post-scabietic nodules. These lesions are not indicative of active mite infestation and may persist for up to 1 year [10, 11]. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000536369).

Case Presentation

A 3-month-old girl with Fitzpatrick Skin Type V, of African and European ethnicity, was referred to our tertiary hospital for further workup of newly diagnosed LCH. History was obtained from her mother, who reported the appearance of erythematous, crusted papules, and nodules that began on the right upper extremity, 1 month prior to presentation (Fig. 1). The eruption subsequently spread to the scalp, face, upper and lower extremities, trunk, inguinal folds, and toe web-spaces.

There was no improvement with emollients or topical hydrocortisone acetate 0.1% cream. At the referring facility, a 3 mm punch biopsy was performed from a lesion on the left upper arm. The biopsy was significant for a lymphohistiocytic infiltrate with abundant eosinophils. Immunohistochemical staining revealed that the infiltrate was positive for CD1a, CD68, CD20, CD3, and Langerin (CD207). A diagnosis of LCH was made, and the patient was referred to our tertiary care center to determine the extent of the disease. She underwent laboratory investigations such as a complete blood count and basic metabolic panel, a skeletal survey, and abdominal ultrasound, which were all unremarkable. The pathology slides were requested for a confirmatory reading as well as genetic studies to determine the presence of BRAF-V600E/MEK aberrations. As these somatic mutations are the most common genetic abnormality associated with LCH, and often associated with multisystem involvement, and poor prognosis [12]. A second reading of the biopsy slides was performed at our facility, which confirmed the presence of cells co-expressing CD1a and S100 (Fig. 2a).

However, in deeper sections, a burrow containing the edge of a mite was noted in the stratum corneum, suggesting scabies rather than LCH (Fig. 2b). Further inquiry revealed that the patient's family members experienced nocturnal pruritus, which they attributed to

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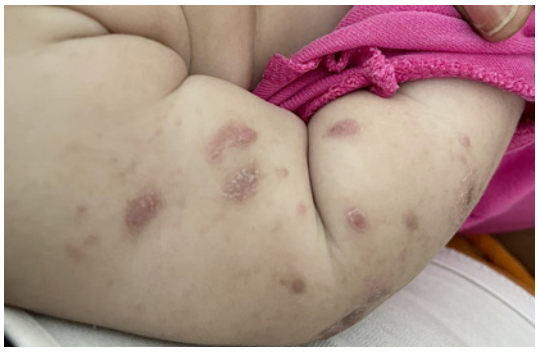

Fig. 1. Erythematous and hyperpigmented papules and nodules, some with thick overlying scales, on the bilateral upper extremities.

moving to a new home. We advised that the patient and her family members receive two treatments with permethrin 5% cream, 1 week apart, as well as decontamination of clothing, bedding, and household items. Six weeks later, the patient demonstrated significant improvement, with complete resolution of inflammatory skin lesions. However, she developed new erythematous nodules on the upper extremities. These nodules were erythematous, firm, non-tender, and lacked scales (Fig. 3). She was retreated with the same regimen of antiscabietic therapy; however, these nodules persisted, making them consistent with post-scabietic nodule [10, 11] lesions that are not reflective of active mite infestation.

Discussion

Scabies is a common parasitic infestation caused by the mite *Sarcoptes scabiei*. The classic clinical presentation is polymorphous lesions, including burrows, papules, and nodules with excoriations. Lesions may be impetiginous, folliculitis-like or have eczematous changes [1]. The presentation amongst children and adults is also variable. For example, infantile scabies has a predilection for the dorsal foot, palm, scalp, and face, and excoriations may be absent. Whereas in adults, lesions favor intertriginous or genital sites and often spare the scalp and the face. Nocturnal pruritus and excoriations are diagnostic clues for scabies in adults [13]. Scabies can be diagnosed clinically based on history and presentation; however, skin scrapings or dermoscopy may aid in confirming the diagnosis [14]. Histopathologic findings include an inflammatory cell infiltrate consisting of numerous CD1a, S100 positive staining histiocytes in the dermis, with histopathologic identification of mites being diagnostic [3]. Langerhans cell hyperplasia (correlating with CD1a and S100 positivity) has been reported in scabies, occasionally even in the absence of mites [3]. This histologic finding may lead to the misdiagnosis of LCH [3, 4, 8], atypical contact dermatitis, pityiriasis lichenoides et varioliformis acuta [15], or lymphomatoid papulosis [16] or molluscum contagiosum [17]. The most important differential diagnosis to exclude in this case is LCH; as patients with suspected LCH require prompt and thorough workup for systemic involvement. LCH is a group of idiopathic disorders of bone marrow-derived Langerhans cells. Cutaneous presentations of LCH include widespread scaly, erythematous patches with petechiae, or crusted lesions or papulo-nodular lesions [4], which may be clinically similar to those seen with scabies. Manifestations range from isolated disease with spontaneous resolution to life-threatening multisystem disease, involving the liver, spleen, and bone marrow [18]. The most common genetic abnormality is somatic mutations in BRAF-V600E, associated with multisystem involvement, and poor prognosis [12].

Burch et al. [4] reported cases of two unrelated young boys diagnosed with LCH who were treated with systemic chemotherapy. Upon further review and re-assessment, both

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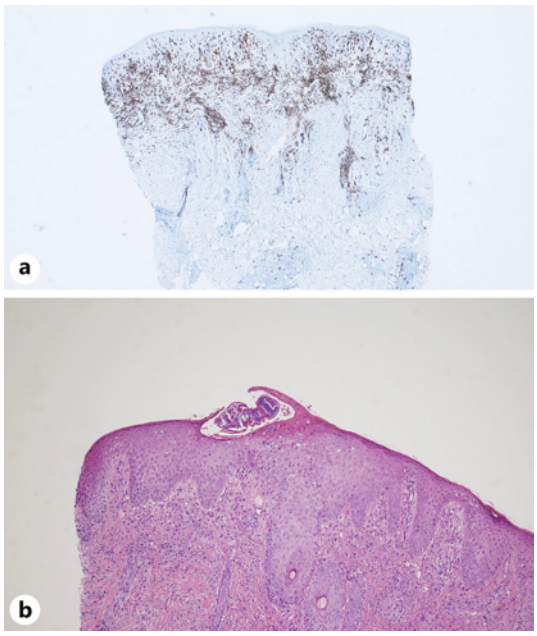

Fig. 2. a CD1a positively staining infiltrate, extending to the deep dermis. **b** A burrow containing the edge of a mite in the stratum corneum.

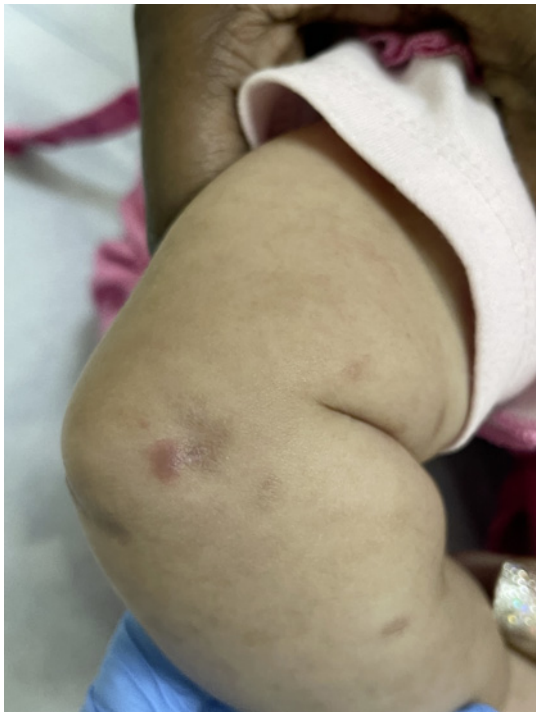

Fig. 3. Firm pink nodules on the upper extremity after scabies treatment.

patients were diagnosed with scabies, which improved after receiving appropriate therapy. The reported cases in which scabies were misdiagnosed as LCH are summarized in Table 1.

The development of post-scabietic nodules, with a waxing and waning course, and lesions that may persist from several months to over 1 year, have been described [10, 11]. Hashimoto et al. [10] reported eight cases of nodular scabies that resulted in post-scabietic nodules. It was proposed

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Table 1. Reported cases of scabies misdiagnosed as LCH

Reference	Clinical presentation	Histopathologic findings	Outcome
Aterman et al. [2] (1978)	13-month-old boy, with erythematous papules on the trunk	An infiltrate in the upper dermis, a perivascular and peri-appendicular infiltrate in the deep dermis, consisting of lymphocytes, eosinophils, and histiocytes	Managed with gamma benzene hexachloride
Talanin et al. [6] (1994)	5-month-old boy with multiple red papules, papulovesicles, and nodules on the chest, extremities, scalp, palms, and soles	Superficial and deep perivascular dermal infiltrate of lymphocytes, eosinophils, and atypical histiocytes	Received permethrin 5% cream, no relapse occurred over the next 6 months, and the skin gradually cleared
Tidman et al. [7] (2003)	27-month-old boy, with a 2-week history of a non-pruritic erythematous papules and nodules on the back and neck	Cellular infiltrate in the upper and deep dermis in a peri-appendageal and perivascular distribution. The infiltrate was comprised predominantly CD1a and S100 positive histiocytic cells, lymphocytes, and eosinophils	Treated with ×2 applications of 5% permethrin cream and shampoo - pruritus and cutaneous eruption cleared completely over the course of 3 months
Janik-Moszant et al. [5] (2003)	8-month-old boy with purple-red maculae and papules on the trunk	Irregular, locally abundant infiltration of the dermis, consisting of histiocytes (CD1a positive), lymphocytes, eosinophils, and polymorphonuclear granulocytes	5% sulfate ointment combined with chloramphenicol, led to a complete cure
Burch et al. [4] (2004)	Case I: 12-year-old boy with a pruritic rash on his torso, scrotum, and penis	Case I: mixed infiltrate consisting predominantly of S100 positive histiocytes and eosinophils, with a few scattered lymphocytes	Case I: at the 6-month follow-up, the patient remained completely clear of lesions
	Case II: 4-month-old boy with apple-jelly-colored papules over the medial upper arms and supra-umbilical	Case II: histiocytic proliferation in the superficial and mid-dermis with eosinophils and lymphocytes. The histiocytes in the infiltrate were CD1a and S100 positive	Case II: after treatment with permethrin 5% cream, the skin lesions cleared
Yang et al. [8] (2015)	6-month-old girl with erythematous to brownish papules and nodules on her trunk	Cellular infiltrate within the upper dermis with a perivascular distribution consisting of CD1a and S100 positive histiocytes, lymphocytes, and eosinophils	Received ×2 treatments with crotamiton 10% cream and was relapse free at 8 months

that these persistent nodules represent a prolonged response to mite antigens. Post-scabietic nodules can lack the scale and crusting typical of scabies infestation and can arise in a non-scabietic distribution, even after successful mite eradication. Familiarity with these lesions is crucial to avoid unnecessary repeated courses of treatment and alleviate the patient's concerns.

Conclusion

Langerhans cell hyperplasia and the presence of CD1a, S100 staining histiocytes are less commonly known entities in scabies. In this case, the diagnosis of scabies was supported by a multitude of findings, including clinical presentation, involvement of family members, and resolution of lesions with anti-scabietic therapy. However, the review of histopathology with deeper sections, leading to the identification of mites, was the key to diagnosis. Familiarity with the possible development of post-scabietic nodules is essential, as the lesions can arise even after successful eradication of the mite; they often lack the secondary crusting and scale of scabietic lesions. Recognition and correct interpretation of these lesions would avert unnecessary patient anxiety and overly aggressive management [11].

Acknowledgments

We would like to extend our gratitude to our patient's mother, who kindly permitted us to share this case for educational purposes.

Statement of Ethics

Ethical approval was not required for this study in accordance with the local or national guidelines. Ethical approval was not required for the publication of case reports for patients treated at Sheikh Khalifa Medical City, Abu Dhabi. Written consent for publication was obtained. Written informed consent was obtained from the patient's mother for publication of details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors declare no conflicts of interest.

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Author Contributions

Sara Al Janahi and Raghda Al Maashari contributed equally to this manuscript. Both physicians were directly involved in the patient's care. Tausif Saleem reviewed histopathology slides and ultimately made the diagnosis; he also contributed to the manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in the references section of this article. Further inquiries can be directed to the corresponding authors.

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