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Case Report / Olgu sunumu

Infantile and Adult Scabies mimicking Langerhans Cell Histiocytosis Clinically and Histopathologically

Klinik ve Histopatolojik Olarak Langerhans Hücreli Histiyositozisi Taklit Eden İnfantile ve Yetişkin Skabiyezi

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Abstract

Scabies is an infestation caused by the Sarcoptes scabiei burrowing into the epidermis. Histopathologically scabies infestation may reveal Langerhans cell hyperplasia which might cause misdiagnosis of Langerhans cell histiocytosis in some cases. We presented an infant and an adult who had a misdiagnosis of Langerhans cell histiocytosis (LCH) histopathologically and responded well to antiscabietic treatments. Therefore, awareness of this phenomenon might help clinicans to differentiate these two diseases with distinct prognosis and treatments.

Keywords: Langerhans Cell Histiocytosis, scabies, infant, adult

Öz

Skabiyez, Sarcoptes scabiei'nin epidermiste tüneller açarak ilerlediği bir enfestasyondur. Histopatolojik olarak skabiyez enfestasyonu Langerhans hücre hiperplazisini ortaya çıkarabilir ve bazı durumlarda Langerhans hücreli histiyositoz tanısının yanlış konulmasına neden olabilir. Histopatolojik olarak Langerhans hücreli histiyositoz (LCH) yanlış tanısı alan ve antiskabiyetik tedavilere iyi yanıt veren bir infant ve bir yetişkin skabiyez hastalarını sunduk. Bu nedenle, bu fenomenin farkındalığı, klinisyenlerin bu iki hastalığı farklı prognoz ve tedavilerle ayırt etmelerine yardımcı olabilir.

Anahtar kelimeler: Langerhans hücreli histiyositoz, skabiyez, infant, yetişkin

INTRODUCTION

Human scabies is a contagious disease caused by the mite Sarcoptes scabiei burrowing into the epidermis.[1] Infestation with the scabies results in severe pruritus which is usually worst at night. Pruritus is due to host hypersensitivity to the mite. Classical cutaneous findings are small erythematous and excoriated papules. Burrows may be visible as serpiginous lines. Differential diagnosis is broad in both pediatric and adult populations. Clinical differential diagnosis of scabies in pediatric population includes LCH. Histopathology of scabies infestations include hyperkeratosis, acanthosis, spongiosis and vesiculation. The dermal changes consist of perivascular and diffuse cell infiltrates, mainly mononuclear cells, and sometimes eosinophils.[2] Cases usually show numerous histiocytes in the infiltrate. Scabietic mites can also be observed in majority of patients. Immunohistochemical characterization of the inflammatory infiltrate showes predominantly CD3+ T lymphoctes and scattered CD20+ B cells. Many CD1a + and S100 + cells were seen in the superficial dermis in a perivascular and interstitial pattern and these cells are have medium size nuclei and delicate dendritic cytoplasm differing from the Langerhans cells in LCH. Since Langerhans cell hyperplasia can also be seen routinely, these findings might lead to misdiagnosis. Diagnosis challenges are

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common in infants as scabies has several atypical clinical presentations.[3-8] Herein two case of scabies, an infant and an adult mimicking Langerhans cell Histiocytosis that was diagnosed by histopathologica examination will be presented.

CASE

Patient 1

A three months old healthy male infant presented with widespread eruption of pruritic papules with fine white scales, pustules and partly with eczematous plaques. He and his family members had a history of treatment with permethrin %5 lotion applied one week apart which did not cured his symptoms and rash after two months of application. Skin lesions of the baby were evaluated by dermoscopic and native examination and no scabies finding was found. Then, his lesions were biopsied with the preliminary diagnosis of Langerhans cell histiocytosis, mastocytosis and eosinophilic pustular dermatosis. His biopsy was compatible with Langerhans cell histiocytosis revealing perivascular infiltrate in superficial dermis rich of histiocytes with irregular nucleus and groove formation accompanied by eosinophils, neutrophils and lymphocytes. Immunohistochemistry showed histiocytes positively stained with CD1a and S-100. Mildly increased mast cells were demonstrated by CD117, giemsa and toluidine blue. Due to presence of previous reports revealing scabies mimicking Langerhans cell histiocytosis both clinically and histopathologically, scabies treatment was repeated and 2 months later, lesions of the patient cleared completely. Histopathological evaluation was and findings were interpreted as reactive Langerhans cell hyperplasia. Follow up of the patient revealed no recurrence of the lesions and physical examination and laboratory evaluations were within normal limits.

Patient 2

Patient with a history of scabies infestation five months ago was referred to our dermatology clinic with a suspicious histopathological diagnosis of Langerhans cell histiocytosis. He had an erythematous plaque with a diameter of two centimeters on the left inguinal area and a erythematous papule located on penis. His skin biopsy demonstrated dense dermal infiltrate which are positively stained with LCA, CD3, CD68 and S100. S100 positive cells had broad cytoplasm. Permethrin %5 lotion applied one week apart to him and his family members. Lesions of the patient cleared after one month of follow up.

DISCUSSION

In a series of six infants and two adult cases of nodular scabies, patients developed multiple papulonodular lesions persisted from several months to over a year. Skin biopsies revealed heavy perivascular and periappendageal lymphohistiocytic cell infiltration, compatible with Langerhans cells which were immunopositive for CD1a and S100, but lacked Birbeck granules on electron microscopy. It has been proposed that these persistant nodules could represent a prolong response to mite antigens.[9] Bhattacharjee and Glusac demonstrated that immunohistochemical labeling showed florid CD1a and S100 positivity in most cases, indicative of Langerhans cell hyperplasia in a series of 16 cases of scabies.[6] Therefore, presence of significant Langerhans cell hyperplasia is not rare and it can be considered as a routine feature of scabies.

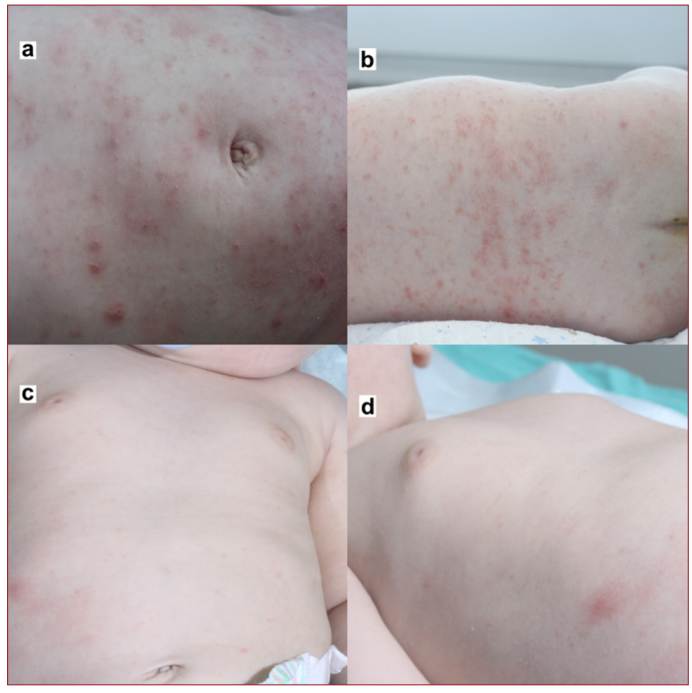

Figure 1. (a-b) Before and (c-d) After scabies treatment

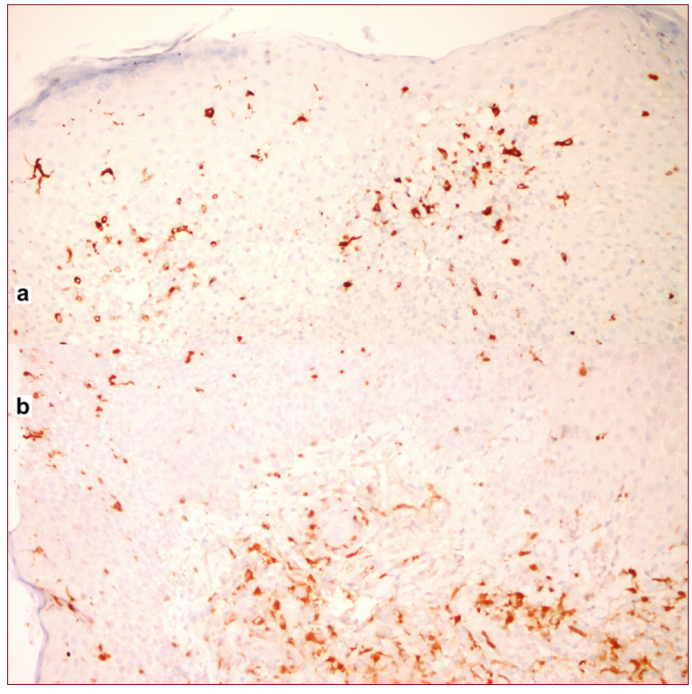

Figure 3. (a) Langerin- and (b)S100 An increase in langerhans cells stained with S100, CD1a and Langerin is observed in dermal infiltration.

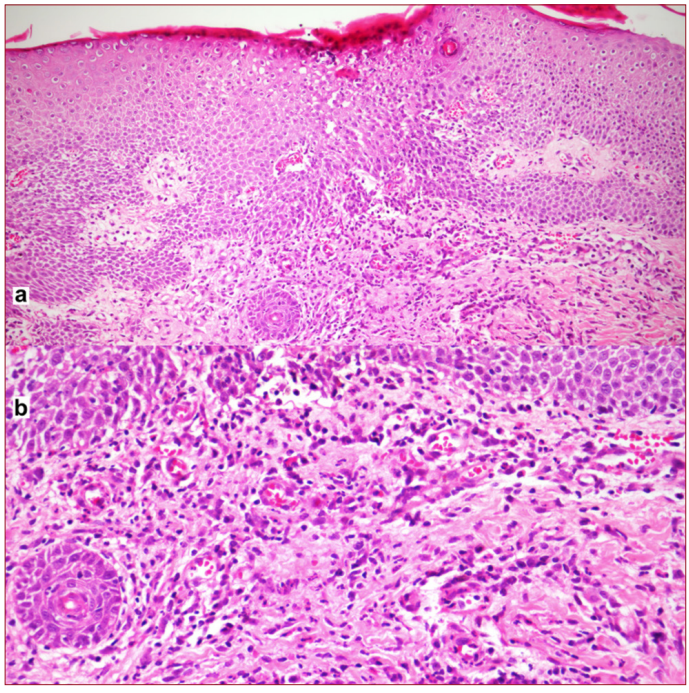

Figure 2. (a) Irregular acanthosis, spongiosis and lymphocyte exocytosis in the epidermis, as well as inflammatory cells in the superficial dermis. HEx200 (b) Inflammation rich in histiocytes in the superficial dermis. Langerhans cell histiocytosis raises suspicion.

Since infantile scabies can present as atypical skin nodules, vesicles, and pustules, it can mimic LCH clinically. Accordingly, histopathological demonstration of Langerhans cell hyperplasia can lead to misdiagnoses both clinically and histopathologically causing even consequences of treatment with systemic chemotherapy5. Misdiagnosis of scabies as LCH is not limited to pediatric population. Atypical presentations such as crusted scabies can cause such misdiagnosis. In a study by Kartono et al hospitalized patient with a hyperkeratotic skin eruption followed for years as LCH and patient receiving cheumotherapetics as a treatment, had a diagnosis of crusted scabies. Her lesions were completely cleared after treatment with 12 mg of oral ivermectin.[10]

CONCLUSION

As a result, scabies must always be ruled out in infants and adults with eczematous eruptions and inflammatory infiltrates that include histiocytes on histologic examination. Since it can cause serious complications such as unnecessary treatments with chemotherapeutics.

ETHICAL CONSIDERATIONS

Informed Consent: Written informed consent was obtained from all participants who participated in this study.

Status of Peer-review: Externally peer-reviewed.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

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Author Contributions: All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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