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We would like to extend our gratitude to the Department of Dermatology & Venereology of Dr. Sitanala Hospital for permitting the preparation of this case report.

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Case Report

Histoid leprosy mimicking lichen planus

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Abstract

Background: Histoid leprosy is a rare variant of lepromatous leprosy (LL), characterized by unique clinical, histopathological, and microbiological features. This type of leprosy is caused by multidrug therapy (MDT) drug resistance, train mutation of *Mycobacterium leprae,* or genetic factors.
Case Illustration: A 21-year-old Indonesian woman with a family history of histoid leprosy complained of multiple hypo-esthetic erythematous partly flat-topped papules around the lesion on the face and bilateral superior and inferior extremities for the last two years. A Slit-skin smear examination revealed acid-fast bacilli with a bacterial index (BI) of 4.17+ and morphological index (MI) of 1%. Histopathological examination on hematoxylin & eosin (H&E) stained revealed epidermal atrophy, Grenz zone, and bundles of thin spindle-like histiocytes with Virchow cells. Ziehl-Nielsen stain showed copious acid-fast bacilli. Therefore, the diagnosis of histoid leprosy was established.
Discussion: Lichen planus (LP) was considered because LP typically presents as pruritic, polygonal, violaceous flat-topped papules with symmetric distribution on the flexural surfaces of the forearms, wrists, and ankles, as well as the dorsal surface of the hands and shins. However, the face is rarely affected in LP. The patient's slit-skin smear and histopathological examinations presented strong evidence for histoid leprosy. Treatment with an MDT regimen resulted in clinical improvements as the erythematous lesions transformed into hyperpigmentation after weeks of treatment.
Conclusion: Histoid leprosy mimicking LP lesions in this patient developed without any prior administration of agents. Additionally, the patient had a high bacillary load, indicating a potential reservoir of the infection as the patient had a family history of leprosy.

Keywords: *histoid leprosy, lichen planus, MDT*

Background

Leprosy, also known as morbus Hansen (MH), is an infectious disease caused by *Mycobacterium leprae*. These bacteria affect the human skin and nerves. Leprosy is diagnosed if there are at least one of the three cardinal signs, including hypopigmentation or red patches with loss of skin sensation (hypoesthesia), peripheral nerve thickening, and identification of acid-fast bacilli (AFB) in the slit skin smear examination or histopathological examination.[1] It is usually transmitted through close contact with a leprosy patient, when the patient coughs or sneezes, or through prolonged direct contact.[2] In 2019,202,256 new cases of leprosy were reported in 118 countries. The highest contributors were India, Brazil, and Indonesia.[3] The Ridley-Jopling system classifies leprosy as tuberculoid leprosy (TT), borderline tuberculoid (BT), mid borderline (BB), borderline lepromatous (BL), and lepromatous leprosy (LL).[4] This case report will deeply discuss histoid leprosy, a variant of LL. Histoid leprosy is a rare condition characterized clinically by multiple painless, succulent, discrete, smooth, globular, skin-colored to yellowish-brown cutaneous and/or subcutaneous nodules and papules with apparently normal skin surrounding the lesions.

The lesions are usually located on the posterior and lateral aspects of the arms, buttocks, thighs, dorsal surface of the hands, lower part of the back, and over the bony prominences, especially over the elbows and knees. A slit-skin smear examination will reveal abundant bacilli and elongated fusiform histiocytes. The leprosy type is thought to be caused by various factors, combining abnormal immune response, inadequate treatment or relapse, and genetic factors. It is often present in patients who have lepromatous leprosy (LL) that either relapses after monotherapy treatment with dapsone, dapsone-resistant patients, or, rarely, a *de novo* infection, of which histoid leprosy develops without evidence of other leprosy types. Clinical differential diagnoses for this condition include sarcoidosis, dermatofibroma, cutaneous metastasis, and angiosarcoma.[5] In India, the incidence of histoid leprosy varied from 2.79% to. 3.60% among all leprosy patients.[6] We report a patient with histoid leprosy whose lesions mimicked lichen planus (LP). LP is an inflammatory skin disease that typically presents as pruritic, polygonal, violaceous flat-topped papules and plaques. The papules often have a dry and shiny surface with a symmetric distribution.[7] The diagnosis of histoid leprosy is established based on the patient's history, physical examination, and histopathological examination.

Case Illustration

A 21-year-old woman visited our clinic at Dr. Sitanala Hospital, Tangerang, Banten, with multiple red dermal lesions on both extremities, ears, and face for the past two years. The lesions first appeared on the left lower extremities and extended to the upper extremities, ears, and face progressively (Figure 1-4). The patient had no experience of discomfort, pain, or itchiness around the lesions. She had no history of similar disease, nor had she taken any prior medications. Her father had leprosy with similar lesions ten years prior but did not take leprosy medications regularly. The patient's family signed an informed consent form to publish the patient's data and photographs.

General physical examination was within normal range. On the face, auricula, as well as bilateral superior and inferior extremities, there were multiple well-defined discrete erythematous partly flat-topped papules, lenticular in size, with some confluence lesions. There was also a thickening of the auricularis magnus and ulnar nerves. Routine laboratory tests were within normal range. A Slit-skin smear examination with Ziehl-Nielsen staining revealed AFB with a bacterial index (BI) of 4.17+ and morphological index (MI) of 1%.

The treatment recommended by the World Health Organization (WHO) for multibacillary leprosy, which consisted of clofazimine, dapsone, and rifampicin, was administered. The patient showed clinical improvement as the erythematous histoid lesions became hyperpigmented after two weeks of treatment (Figure 5-6). The treatment was continued for a total of 12 months. Histopathological examination showed significant epidermal atrophy, Grenz zone, and bundles of thin spindle-like histiocytes with Virchow cells (Figure 7). Ziehl-Nielsen staining showed copious bacilli (Figure 8). The diagnosis of histoid leprosy was established based on the patient's history as well as clinical and histopathological findings.

Discussion

Histoid leprosy, first described by Wade in 1963, is a distinct and rare variant of lepromatous leprosy. It is characterized by unique clinical, histopathological, and microbiological features. This variant is common in patients receiving dapsone monotherapy or having irregular treatment, although it can sometimes arise as a *de novo* infection.[8] A Brazilian study reported that histoid leprosy constitutes 1.12% of all leprosy cases; however, studies on histoid leprosy remain limited.[9] The condition usually affects men between 30 and 50 years old. In this case report, the patient was a 21-year-old woman, which makes this presentation unusual. Most patients have low socioeconomic status and a history of contact with leprosy patients.[10]

Histoid lesions commonly appear as smooth, shiny, hemispherical, dome-shaped, non-tender, soft-to-firm nodules, which might be superficial, subcutaneous, or fixed deeply under the skin. They can also appear as plaques or pads on normal-looking skin.[10] The papules in this patient developed on the extensor surface of the upper and lower extremities and the face. Erythematous shiny papules were the morphological pattern with a significant proportion in this patient, arising from apparently normal skin.

The lesions were firstly thought as mimicking LP because LP typically presents as pruritic, polygonal, violaceous flat-topped papules, with symmetrical distribution on the flexural surfaces of the forearms, wrists, and ankles, as well as the dorsal surface of the hands and shins. The face is rarely affected in LP cases.[7] However, other differential diagnoses such as lichen nitidus, keratosis lichenoides chronica, and erythema dyschromicum perstans (ashy dermatosis) should also be considered.

A slit-skin smear examination was performed to differentiate histoid leprosy and other diagnoses. The results showed abundant acid-fast bacilli, occurring as a single entity or tightly clustered. The histiocytes and macrophages were packed with leprosy bacilli, characteristically longer than ordinary leprosy bacilli with tapered ends.[9] BI in histoid leprosy might be 3+ to 6+, and MI might be 0 to 10% or very high. The enormous bacillary population in histoid lesions is suggested to be due to focal loss of immunity.[11] This theory fits the patient's condition: her BI and MI were 4.17+ and 1%, respectively.

Histopathological findings in histoid leprosy include epidermal atrophy and dermis with a Grenz zone and circumscribed collections of fusiform histoid cells distributed in a storiform pattern.[11-13] The histopathological findings in the current patient showed prominent epidermal atrophy, Grenz zone, spindle-like histiocytic bundles, and Virchow cells (foamy appearance). Microscopic examination with Ziehl-Nielsen staining showed abundant bacilli, suggesting a BI of 6+. Based on the patient's history and physical examination, supported by laboratory, histopathological, and bacteriological examinations, we diagnosed this patient with histoid leprosy. We gave the patient multidrug therapy (MDT) according to WHO recommendation, consisting of clofazimine, dapsone, and rifampicin. The patient was followed up after two weeks of treatment. Clinical improvement was observed as the erythematous histoid lesions became hyperpigmented. Continuous treatment for 12 months is necessary for optimal bacterial clearance.

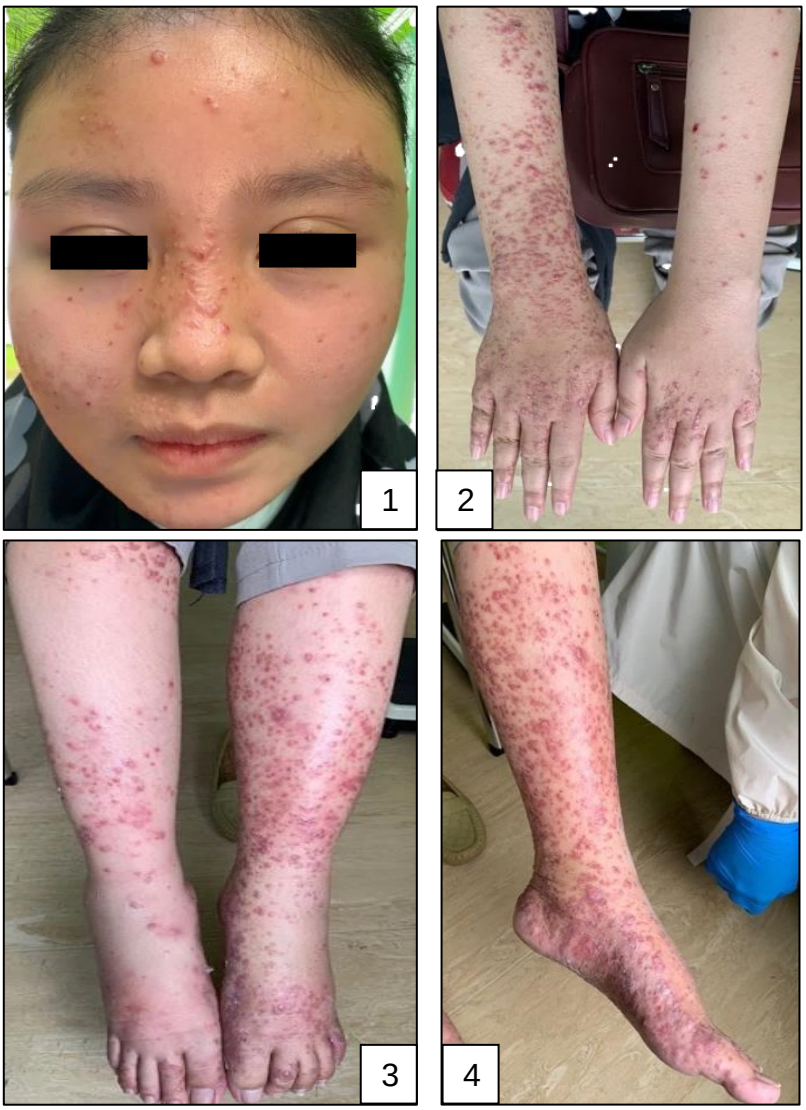

Figure 1-4. Multiple well-defined, discrete, erythematous, partly-flat-topped papules, lenticular in size, with some confluent lesions on the face and the extremities

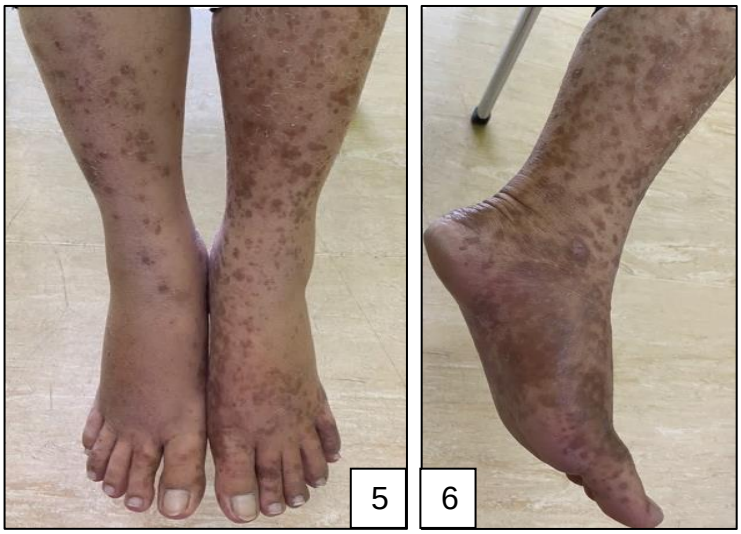

Figure 5-6. After two weeks of treatment, improvement was observed in multiple well-defined, discrete, hyperpigmented, partly flat-topped papules, lenticular in size, with some confluent lesions

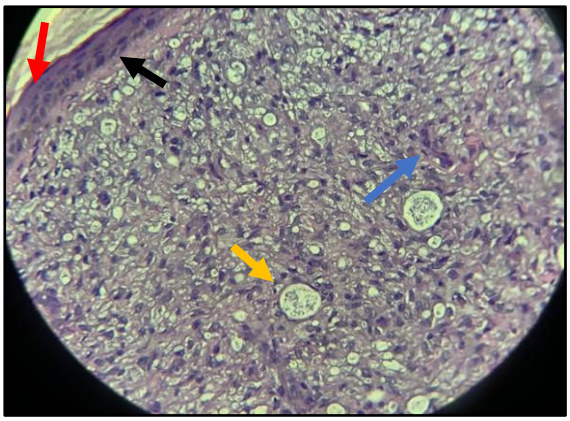

Figure 7. High-power (400x) view with hematoxylin & eosin (HE) staining showing showed prominent epidermal atrophy (red arrow), Grenz zone (black arrow), spindle-like histiocyte bundle (blue arrow), and Virchow cells (yellow arrow)

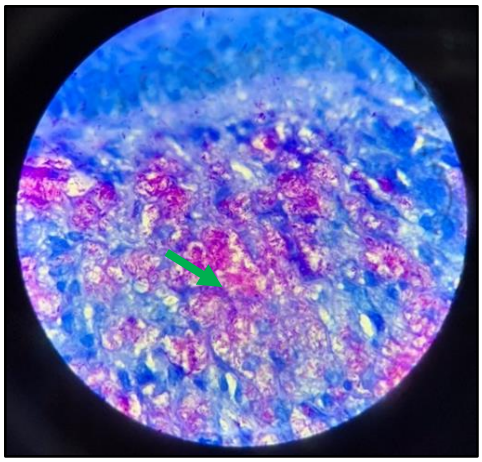

Figure 8. High-power (400x) view with Ziehl-Nielsen staining showed macrophages with abundant acid-fast bacilli (green arrow)

Histoid leprosy has been reported generally to manifest in patients after long-term dapsone monotherapy, irregular or inadequate therapy, developing as relapse after successful treatment, or even appearing *de novo* without a prior history of any antileprotic agents. *De novo* denotes that the lesions occurred without any previous history or treatment and, therefore, without the possibility of being a relapse case.[8] The current patient did not have any previous history of leprosy, nor had she taken dapsone before, making this case unique and distinctive.

The pathogenesis of this unusual variant has not been fully understood. It is thought to be the result of multiple factors, combining genetic factors, abnormal immune response, and inadequate treatments. Despite adequate numbers of macrophages, it has been claimed that they lack the functional property to kill high numbers of bacilli in histoid lesions.[11] In this patient, genetic factors may contribute, as she had close contact with her father, who also had leprosy with similar lesions 10 years prior. However, her father did not take any antileprotic agents. There is no clear recommendation regarding the treatment regimen for histoid leprosy. As some clinicians consider it a variant of LL, the condition is treated as multibacillary leprosy.[12,13]

Conclusion

Histoid leprosy is a rare type of multibacillary leprosy with characteristic clinical, bacterial, and histopathological features. It is very important to perform a slit-skin smear examination to differentiate histoid leprosy and lichen planus. Histopathological features such as epidermal atrophy, Grenz zone, diffused histiocytic bundles, and Virchow cells are important findings in histoid leprosy.

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Author Contributions

All authors contributed equally to the study preparation, data collection, case analysis, and the writing of the manuscript.

Conflict of Interest

No conflict of interest.

References

1. Chen KH, Lin CY, Su SB, Chen KT. Leprosy: A review of epidemiology, clinical diagnosis, and management. J Trop Med. 2022;2022: 8652062.
2. Ploemacher T, Faber WR, Menke H, Rutten V, Pieters T. Reservoirs and transmissions routes of leprosy: A systematic review. PLoS Negl Trop Dis. 2020;14(4):1-27.
3. World Health Organization. Towards zero leprosy. Global leprosy (hansen's disease) strategy 2021 – 2030. New Delhi: World Health Organization, Regional Office for South-East Asia; 2017. p.21.
4. Kumar B, Uprety S, Dogra S. Clinical diagnosis of leprosy. In: Scollard DM, Gillis TP, editors. International textbook of leprosy. Greenville: American Leprosy Missions; 2016. p.3-4.
5. DeAndrade PJS, Ferreira P, Machado A, et al. Histoid leprosy: A rare exuberant case. An Bras Dermatol. 2015;90(5):756-7.
6. Ambade GR, Asia AJ. Clinico-epidemiological profile of histoid leprosy: A prospective study. Natl J Integr Red Med. 2018;9(1):16-9.
7. Weston G, Payette M. Update on lichen planus and its clinical variants. Int J Womens Dermatol. 2015;1(3):140-9.
8. Mathur M, Jha A, Joshi R, Wagle R. Histoid leprosy: A retrospective clinicopathological study from central Nepal. Int J Dermatol. 2017;56(6):664-8.
9. Canuto MJM, Yacoub CRD, Trindade MAB, Avancini J, Pagliari C, Sotto MN. Histoid leprosy: Clinical and histopathological analysis of patients in follow-up in university clinical hospital of endemic country. Int J Dermatol. 2018;57(6):707-12.
10. Calvache N, Arias DA, Delgado VM. Histoid leprosy: An enemy not yet eradicated. Actas dermo. 2021;112(3):280-1.
11. Pandey P, Suresh MSM, Dey VM. De novo histoid leprosy. Indian J Dermatol. 2015;60(5): 525.
12. Raheja A, Kapoor A, Deora MS, Sharma YK. De-novo histoid leprosy: A series of four cases. Indian J Lepr. 2022;94:257-62.
13. Samiey F, Aljalahma J, Awadhi AA. De novo histoid leprosy with unusual histological features. Cureus. 2021;13(11):e19.