

De Novo Histoid Leprosy In A 60-Year-Old Man: Rare Case Report

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Abstract

Introduction: Leprosy is a chronic granulomatous infection caused by *Mycobacterium leprae*. Histoid leprosy is a rare form of leprosy with clinical manifestations of papules and nodules on the skin or subcutaneous tissue. Histoid leprosy can be found de novo in patients without a history of previous leprosy treatment.

Case: A 60-year-old man presented with multiple papules and nodules on the superior and inferior extremities 3 months before admission. The patient had a history of traveling to leprosy endemic areas 2 years previously. There was no history of previous leprosy treatment. Skin-slit smear examination showed a bacterial index of +4 and a morphological index of 40%. Fite Faraco staining showed acid-fast bacilli (+). Histological examination showed spindle-shaped histiocytes on the dermal layer. The patient was diagnosed with histoid leprosy with the management given 2 years of multibacillary Multi-Drug Therapy (MDT). In this case evaluation in the first and third month of treatment showed clinical and bacteriological improvement.

Conclusion: Histoid leprosy has unique and distinctive characteristics. Histoid leprosy mostly occurs with a history of previous leprosy treatment but can also occur de novo. Early diagnosis of histoid leprosy is important to reduce the transmission rate of this disease in the general population. Histoid leprosy treatment, apart from administering antimicrobial drugs, must also include education for patients and their families regarding the course of the disease, possible reactions, monitoring and management of nerve damage, disability care, social support, and rehabilitation.

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I. Introduction

Leprosy is a chronic granulomatous infection caused by *Mycobacterium leprae*. *Mycobacterium leprae* is an obligate intracellular pathogen that affects the skin and peripheral nerves causing various types of skin lesions with anesthesia, peripheral neuropathy, and muscle weakness.¹ Indonesia ranks third in the highest incidence of leprosy in 2015.² Histoid leprosy is a clinical entity of leprosy that occurs in lepromatous leprosy (LL)-type or is sometimes found in borderline lepromatous (BL)-type leprosy with clinical manifestations in papules and nodules on the skin or subcutis on normal-looking skin. Histoid leprosy is usually found in patients treated with long-term diaminodiphenyl sulfone (DDS), but histoid leprosy currently, is often found *de novo* in patients who have never received any antileprosy treatment.³

The diagnosis of histoid leprosy is made based on the typical clinical picture and supported by microscopic examination (bacilloscopy) and histopathology. Histopathology in histoid leprosy shows epidermal atrophy with Grenz zone and spindle-shaped histiocytes arranged in a storiform pattern.⁴ Histoid leprosy management consists of administering a multibacillary MDT regimen for 2 years and is better until the skin smear is found to be negative. Bartos et al 2020 in the United States reported that giving multibacillary MDT for 24 months in histoid leprosy increased clinical response (97%) and decreased bacterial index (81%) compared to clinical response (82%) and decreased bacterial index (29%) when administered only for 12 months.⁵

Early diagnosis of histoid leprosy is important to reduce the rate of transmission of this disease in the general population.⁶ This case report discusses a case of de novo histoid leprosy in a 60-year-old man. The purpose of writing this case report is so that clinicians can gain insight into the diagnosis and management of histoid leprosy so that early diagnosis and therapy can be given to reduce the morbidity of the disease.

II. Case

A 60-year-old man presented with multiple papules and nodules on both hands and knees since 3 months ago. The lesions are not itchy, not painful, not accompanied by redness but feel thick and numb. Complaints are not accompanied by fever, weakness, or joint pain. History of similar illnesses and medication

taken before the illness was denied. The patient has a history of traveling and working in leprosy-endemic areas for 3 years as a construction worker. A history of similar complaints in family members was denied.

Dermatological examination of the bilateral auricular region showed infiltrates, the anterior and posterior trunk, and the bilateral superior and inferior extremities showed multiple scattered skin-colored papules with shiny surfaces(**Figure 1**). Examination of the sensory nerves revealed stockings and gloves anesthesia.



Figure 1. (A-B) Bilateral auricular region showing infiltrates. (C-H) The anterior and posterior trunk, and superior and inferior extremities showed multiple discrete skin-colored papules and nodules.

Skin-slit smear examination with Ziehl-Nielsen (Zn) staining and histopathological examination were performed in this case to confirm the diagnosis. Skin-slit smear examination on bilateral ear lobes showed positive acid-fast bacilli (AFB) with a bacteriological index of +4 and a morphological index of 40%. Histopathological examination with Hematoxylin Eosin (HE) staining showed epidermal atrophy with flattening of the rete ridge, subepidermal Grenz zone, and spindle-shaped histiocytes on the dermal layer (**Figure 2**). Fite faraco (FF) staining were positive.

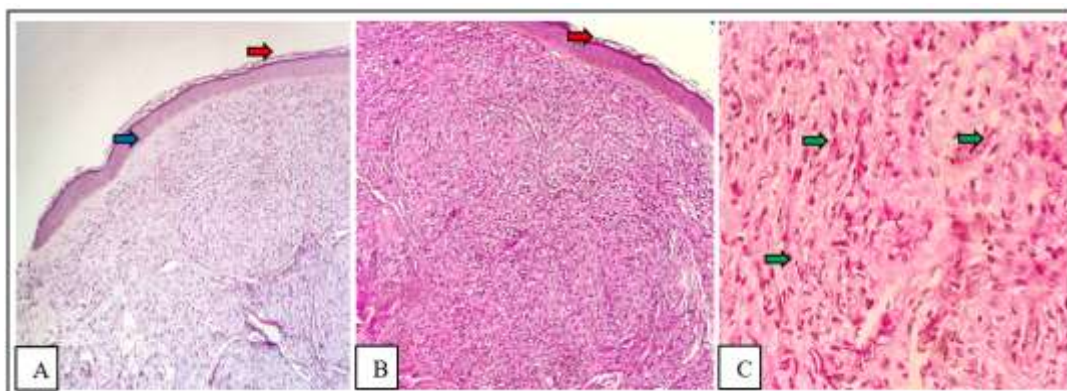


Figure 2. (A-B) Epidermal atrophy and flattening of the rete ridge (red arrow), visible subepidermal Grenz zone (blue arrow), HE, 10x. (C) The dermis shows spindle-shaped histiocytes (green arrow).

Based on the history taking, physical examination, and histopathological examination, the patient was diagnosed with histoid leprosy. The patient was given a multibacillary MDT regimen in the form of rifampicin 600 mg monthly, dapsone 100 mg daily, and clofazimine 300 mg monthly followed by 50 mg daily. Treatment was planned to be given for 2 years and until the results of the BTA re-examination were negative. In the third month of follow-up, clinical and bacterial improvement was found (**Figure 3**). No new lesions were found with

a bacterial index decrease to +2 and a morphological index of 0%.



Figure 3. Visible improvement after 3 months of therapy.

III. Discussion

Leprosy also known as Hansen disease is a chronic infectious disease caused by *Mycobacterium leprae*. *Mycobacterium leprae* is a non-motile, acid-resistant, rod-shaped bacterium with 4-7 μm long. Leprosy affects all ages and ethnicities, but the highest incidence is found in individuals aged 10-15 years and 30-60 years. Transmission of leprosy is caused by three important factors, namely individuals infected with leprosy, susceptible individuals, and close contact between the two.⁷

Leprosy remains a health burden worldwide. Epidemiological data from the World Health Organization (WHO) reported that there were a total of 214,783 new cases of leprosy from 143 countries in 2016 with a global incidence rate of 2.9 cases per 100,000 population. Indonesia is still one of the three countries with the most cases of leprosy along with Brazil and India with >10,000 new cases reported during 2016.⁸ Prakoeswa et al in 2022 reported that the incidence of leprosy in Indonesia was more commonly found in men (66.8%) with the most common type being multibacillary (86.2%).⁹ The clinical presentation of leprosy varies which is related to the spectrum of the host immunity to the disease. One of the rare manifestations of leprosy is histoid leprosy.¹⁰

Histoid leprosy was first reported by Wade in 1963 and is also known as "Wade Leprosy".¹¹ Histoid leprosy is a rare form of LL-type leprosy with an incidence of 1.2% of all leprosy patients. The incidence of histoid leprosy is found to be more common in men than women with a ratio of 5.7:1.¹² The most common age for histoid leprosy is in young adults under 40 years but can also affect those aged 50-60 years.⁴ The high incidence of histoid leprosy in men is thought to be due to greater exposure especially those who work outside the home. Mendirrata et al in 2011 reported in India that 70% of 11 histoid leprosy patients were men who worked as casual laborers.¹³ In this case the patient was a 60-year-old man who worked as a construction worker in a leprosy endemic area.

Histoid leprosy has unique and distinctive clinical and histopathological characteristics. Clinical manifestations of histoid leprosy include soft, painless, and shiny well-defined papules and nodules of varying sizes on normal-looking skin. In rarer cases, infiltrative plaque can be found. Ear infiltrates were found in 18.8% of histoid leprosy patients. The lesions are more often found in areas of bony prominences such as elbows and knees with a spontaneous and progressive onset.^{14,15}

Histoid leprosy is more common in lepromatous patients who experience a recurrence after receiving dapsone monotherapy or irregular MDT treatment.¹⁶ *De novo* histoid leprosy is the occurrence of histoid leprosy without a history of leprosy or previous history of leprosy treatment. *De novo* histoid leprosy is only found in 12.5% of histoid leprosy cases. Pathania et al in 2013 in India reported one case of *de novo* histoid leprosy with a predilection for all four extremities and trunk with an onset of 2 months.¹² Malhotra et al 2019 in India reported 6 cases of *de novo* histoid leprosy without a history of leprosy or previous anti-leprosy treatment.¹⁰ In this case, complaints appeared progressively within three months without any history of leprosy or previous leprosy treatment.

Bacteriological examination with a skin-slit smear showed the presence of AFB which was clustered and dense. The bacterial index ranges from +4 - +6 with varying morphological index.¹⁷ Typical histopathological findings in histoid leprosy are very specific in the form of epidermal atrophy, subepidermal Grenz zone, spindle-shaped histiocytes arranged in a circular or wavy pattern, and a positive result of staining for acid-fast bacilli.^{1,11} In this patient, the results of histopathological examination revealed thinning of the epidermis accompanied by a subepidermal Grenz zone, and in the dermis spindle-shaped histiocytes were found arranged

in a wavy pattern. The results of FF and Zn staining were found positive.

Until now there are no specific recommendations regarding the management of histoid leprosy.¹⁸ Gupta et al in 2015 in India reported the initial administration of rifampin 600 mg, ofloxacin 400 mg, and minocycline 200 mg followed by an MDT regimen.¹⁹ Another recommendation is only to use the multibacillary MDT regimen as a treatment for histoid leprosy for 2 years.³ Kantaria in 2014 in India reported one case of histoid leprosy which showed a good therapeutic response within one month of administering multibacillary MDT therapy.¹⁴ The patient in this case was treated with MDT regimen consisting of 600 mg rifampicin monthly, 100 mg dapsona daily, and 300 mg clofazimine monthly followed by 50 mg daily. Clinical evaluation in the first and third months of treatment showed improvement with no new lesions. Bacteriological re-evaluation with a skin-slit smear in the third month of treatment showed a decrease in the bacteriological index to +2 with a morphological index of 0%.

Patients with histoid leprosy during the disease may develop a type 2 leprosy reaction, namely erythema nodosum leprosum (ENL). Mulianto et al in 2022 reported a high bacterial index, especially in LL-type leprosy including histoid leprosy with $IB \geq 4$, which is one of the risk factors for ENL.²⁰ In this case patients were given education regarding possible reactions that could occur both during and after treatment.

IV. Conclusion

A 60-year-old man presented with multiple skin-colored papules with a shiny surface scattered discretely on the anterior and posterior trunk regions and bilateral superior and inferior extremities. Anamnesis revealed a history of traveling to a leprosy endemic area 2 years before the complaint appeared. Previous history of leprosy treatment was denied. Sensory nerve examination shows stockings and gloves anesthesia. Skin-slit smear examination revealed positive BTA with IB +4 and IM 20%. Histopathological examination showed epidermal atrophy with flattening of the rete ridge, and subepidermal Grenz zone and in the dermis, spindle-shaped histiocytes were seen. FF and Zn staining showed positive BTA. The patient was diagnosed with histoid leprosy and was given MB MDT therapy for 2 years. Clinical evaluation in the first and third months of treatment showed improvement. Bacteriological showed a decrease in bacteriological index to +2 with a morphological index of 0%.

V. References

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