A Very Rare Case of De Novo Histoid Leprosy with Type 1 Reaction

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Abstract: Histoid Leprosy (HL) is a distinct and rare type of leprosy with unique clinical manifestations and specific histopathology of multibacillary leprosy (MB). Histoid leprosy generally occurs in lepromatous leprosy patients (LL) or borderline lepromatous (BL). Often it occurs after inadequate dapsone treatment despite sometimes arise de novo as well. Histoid forms can ensue accompanied by reactions of erythema nodosum leprosum (ENL) and rarely reversal reactions. Diagnosis of HL based on clinical manifestations, bacteriological examination, and histopathological examination. Clinical manifestations are typical such as coppery nodules, well-defined, might be soft or hard, shiny, with varying sizes. Histopathological findings showed a longitudinal or spindle histiocyte containing M. *leprae*. We report a rare case of histoid leprosy accompanied by reversal reactions. A 59-year-old Chinese woman, from West Borneo, complained of a painless nodule on the face and body since two months ago with swollen face and rashes. Patients never had leprosy treatment before Histopathological shows some spindle cells. Patients are diagnosed with histoid leprosy and reversal reactions. Diagnosis of histoid leprosy is made by history, physical examination, and histopathology. Patients fall into the *de novo* category due to having never been treated with dapsone before. The therapy given to patients is multibacillary multi drugs therapy (MDT-MB) and corticosteroids. The patient showed improvement in lesions after six months of MDT-MB and corticosteroids for three months. Histoid leprosy is able to occur *de novo* and accompanied by reversal reactions. Early diagnosis and complete treatment are crucial to achieve the goal of elimination of leprosy.

1 INTRODUCTION

Histoid leprosy (HL) is a rare type of leprosy with typical and histopathological clinical manifestations specific to the multibacillary type of leprosy (MB). Wade in 1960 first reported the form of leprosy, and several other cases in 1963. (Sehgal VN, 2016) Histoid leprosies generally occurs in lepromatous (LL) or borderline lepromatous (BL) type leprosy without or during or after treatment dapsone.[1,2] Some reports also show that HL could manifest in paucibacillary type (PB).(Sehgal VN, 2006; Rao AG, 2016). Additionally, this histoid can also occur in patients with severe immunosuppression conditions caused by human immunodeficiency virus (HIV) or even with erythema nodosum leprosum (ENL) reactions and reversal reactions(Sun J et al, 2017; Kolaparambath BA et al, 2014).

HL incidence in India is around 2.79-3.60% of all leprosy patients, and de novo incidents are increasing every day.(Kaur et al., 2009; Hali F et al 2011). There is still an unknown number of incidents in Indonesia. Men are dominant to women and rarely occur in children. Histoid leprosy is a variant of LL type, but the HL immune response is better both cellular and humoral immunity. .(Sehgal VN, 2016) HL diagnosis based on clinical manifestations, bacteriological examination, and histopathological examination. (Sehgal VN, 2016;Kalla G et al., 2000).

Clinical manifestations HL is very typical in the form of coppery nodules, well-defined, can be soft or hard, shiny, with varying sizes (diameter up to 3 cm) that arise in cutaneous or subcutaneous above normal-looking skin. The predilection of lesions usually on the face, arms, back, chest, abdomen, and buttocks. Bacteriological examination results show an increase of solid bacilli due to the multiplication of *M. leprae* experiencing resistance, which causes

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bacteriological index (IB) and morphological index (IM) levels high. .(Sehgal VN, 2016)

Histopathological findings HL shows histiocytes that are elongated or spindle-shaped cells containing *M. leprae*, thus forming a curved arrangement. Treatment of histoid leprosy uses multibacillary multidrug therapy (MDT-MB), consisting of rifampicin, dapsone, and clofazimine given for at least two years or up to a negative morphological index.

2 CASE

A 50 years old woman, Chinese descendants from Pemangkat West Borneo, came to the dermato-venereology clinic of Cipto Mangunkusumo National General Hospital with chief complain of painless nodules on her face and body since two months ago. Initially four months ago patient complained of red patches on both leg, and buttocks. The patches felt numb and widespread gradually. Patients were tested for acid-resistant bacilli (AFB) and blood tests in other hospitals, and the result was positive for leprosy. She was given multidrug multibacillary therapy for leprosy with a target of 1year therapy.

After the second month treatment, she has multiple erythema nodules appeared on the face, body, arms, and legs. Nodulesare painless with minimal pruritus. Nodules are expanding and multiply. The patient feels uncomfortable with a nodule on her face due to disturbing appearance. There is no fever nor joint pain. There are no lesions that become increasingly numb or weak. There are no complaints of thinning eyebrows nor dry skin.The patient also complained about swollen face and both legs with more erythema on the patches and nodules in the last one month. The patient is treated with 3 x 4 mg methylprednisolone. Swelling is reduced. But the erythema is still the same. There is no radiating nerve pain, and there are no new patches. History of taking drugs other than leprosy is denied in the past two months. There was a history of drinking herbal medicine two months ago. She never takes any leprosy drug before. No one in her family or neighbor has ever had leprosy that she knew.

Physical examination shows multiple painless erythematous papules to nodules, some of it was skin-colored, dome-shaped, on the face, chest, arms, trunk, abdomen, and legs. There also an erythematous plaque on both legs and buttocks, well defined, and some of it has some punch out-like lesion. Non-pitting edema found on the face and foot. There was enlargement without tender on both Nerve Peroneus communis and N.Tibialis posterior. Skin smear examination shows no solid bacilli but positive fragmented bacilli (+2 Bacteriological Index) on both ears.

Histopathology examination shows Grenz zone, circumscribed macrophage granuloma with the predominance of spindle-shaped cells, foreign body giant cells, and some foamy macrophages. Some of the cells look edema. Fite Faraco shows no acid-fast bacilli. All the examination leads to histoid leprosy with reversal reaction for diagnosis. The patient is treated with WHO regimen multidrug therapy multibacillary such as rifampicin 600mg, clofazimine 300mg, and dapsone 100mg once a month with dapsone 100mg and clofazimine 50mg daily for two years. The patient also got methylprednisolone 32mg daily (equivalent prednisone 0.8mg/kg) then gradually reduced weekly and eventually stopped for reversal reaction. Clinical improvements both subjective, and objective in six months of multidrug multibacillary therapy and three months of corticosteroid therapy.

3 DISCUSSION

Histoid leprosy (HL) is a rare type of leprosy with typical and histopathological clinical manifestations specific to the multibacillary type of leprosy (MB). .(Sehgal VN, 2016). It is characterized by unique clinical, histopathological, and microbiological features. Histoid leprosy generally occurs in lepromatous (LL) or borderline lepromatous (BL) type leprosy without or during or after treatment dapsone. This form of leprosy is relatively common in patients on dapsone monotherapy and irregular treatment. Sometimes, it can arise de novo as well. .(Sehgal VN, 2016;Alious VN, 2006). Some reports also show that HL could manifest in paucibacillary type (PB) .(Sehgal VN, 2006,Rao AG, 2016). Additionally, this histoid can also occur in patients with severe immunosuppression conditions caused by human immunodeficiency virus (HIV) or even with erythema nodosum leprosum (ENL) reactions and reversal reactions.(Sun J et al., 2017; Kaur, et al, 2009).

There is male preponderance, rarely in children, and the average age at diagnosis is between 21 and 40 years. Clinically, it is characterized by cutaneous or subcutaneous nodules and papules, which are painless, coppery nodules, well-defined, can be soft or hard, shiny, with varying sizes (diameter up to 3

cm) that arise in cutaneous or subcutaneous above normal-looking skin. The lesions are usually located on the posterior and lateral aspects of the arms, buttocks, thighs, dorsum of the hands, lower part of the back, and over the bony prominences, especially over elbows and knees. (Sehgal VN, 2016). Clinically, histoid leprosy may mimic lepromatous leprosy or ENL reaction.

However, specific histopathology of histoid leprosy differentiates it from LL type which shows macrophage granuloma with a variable amount of foamy changes with many bacilli and globi and ENL reaction which shows features of acute inflammation predominantly having neutrophils accompanied by edema along with granular AFB. Histoid leprosy might represent an enhanced response of the multibacillary disease in localizing the disease process. An increase in both cell-mediated and humoral immunity against *Mycobacterium leprae*, as in lepromatous leprosy, has been hypothesized.

From history, the patient's gender and age were not in accordance with the epidemiological data on the incidence of the most common histoid leprosy. However, epidemiological data obtained from India, not Indonesia.(Kaur et al., 2009). She is a Chinese descendant which are quite susceptible to leprosy.[12]Patients had never taken leprosy treatment before which is defined as "*de novo*".(Pandey P et al, 2015). The patient complained of swelling, old reddening patches, and a history of drinking herbs. This is in accordance with the reversal reaction. In some cases, a reversal reaction with histoid leprosy has been found simultaneously. (Sun J et al., 2017;Singh N et al., 2015).On physical examination, face, chest, arms bilaterally, back, abdomen, and limbs shows multiple erythematous-skin-colored nodules, dome-shaped, shiny, milliary-nummular size. There is no tenderness. Predilection and morphology of the lesions are in accordance with the appearance of histoid leprosy but can still be diagnosed in comparison with MH nodularlepromatous type.[1]Clinical differences between nodular lepromatous type and HL are in nodular lepromatous type, and nodules arise from infiltrated regions while in HL from healthy skin. The nodular lepromatous type has diffuse nodes whereas well defined in the histoid node.

The differential diagnosis, which is erythema nodosum lepromatous, can be excluded because there is no tenderness and no history of pain. Erythematous plaques on the buttocks and limbs, well defined, with punch-out like lesion accompanied with a dry white patch above them and hypoesthesia, are thought to be borderline tuberculoid lesions. Edema and warmth on plaque palpation are thought to be due to reversal reactions. Nerve enlargement is found in both N. peroneus communis and posterior tibialis. Investigation of slit skin smear was obtained, and the result is +2 in the bacterial index from right ear lesions. This indicates that the patient belongs to the multibacillary type. AFB results were only found in a few fragmented *bacilli* that were not compatible with histoid leprosy. However, in some cases, AFB is only positive in histoid lesions, not in skin lesions that look normal. This is what distinguishes lepromatous leprosy.[1] It is necessary to do a histopathological examination to determine the type of leprosy further.

Histopathological examination with hematoxylin-eosin (HE) staining was obtained, and it shows Grenz zone, macrophage granuloma with foam cells, and a lot of spindle cells. HL specific histopathological features, namely the depletion of the epidermis due to the pressure of the dermal pseudocapsule mass consisting of histiocytes consisting of spindles and intertwining. It is different from lepromatous histopathology, which is thinning of the epidermis, the rete ridge becomes flatter, the grenz zone, and the dermis is found diffuse leproma consisting of foamy macrophages and lymphocytes and plasma cells. Spindle cells are pathognomonic markers in histoid histopathology. Fite-Faraco found no AFB. From these results, it can be concluded that in accordance with histoid leprosy despite there is no AFB in Fite-Faraco staining.(Sehgal VN, 2016) [1]This could be caused by the low sensitivity of Fite-Faraco staining (40-70%), and it worsens when the bacterial index is below 3.(Cabic E, 2018;Adiga et al., 2016)

In this case, MDT-MB therapy was given, which provided clinical improvement after six months of treatment. This is in accordance with the study of Hali et al., Who evaluated treatment responses in two HL patients and received improvement after three months of providing MDT-MB.[10]Patients still need MDT-MB for at least the next 18 months.[1]Patients were also given 32 mg /day of Methylprednisolone (equivalent to Prednisone 40 mg/day) for two weeks then tapered off every two weeks for 12 weeks to treat the reversal reaction. Reversal reactions in patients have been completely gone after giving 12 weeks. The prognosis in this patient is *Bonam* because there are no complications in the patient, and there are no other systemic diseases. The function of patients is*dubia ad bonam* due to possible improvement in nerve function in the hands and feet due to leprosy, according to these patients, there is no sensory or motor impairment.

This patient needs to be examined but must wait at least once a month. Prognosis and sanactionam are *dubia ad bonam* because histoid leprosy can be cured if taking medication regularly and completing treatment for two years but can relapse again while getting treatment or after complete treatment.

4 CONCLUSION

Histoid leprosy is a rare type of leprosy and the diagnosis made by history, physical examination, and histopathology. Patients fall into the *de novo* category due to having never been treated with dapsone before. The therapy given to patients is multibacillary multi drugs therapy (MDT-MB) and corticosteroids. The patient showed improvement in lesions after six months of MDT-MB and corticosteroids for three months. Histoid leprosy is able occurred *de novo* and accompanied by reversal reactions despite its very rare. This case may act as reservoirs of the disease and lead to further spread of leprosy. Early diagnosis and complete treatment are crucial to achieve the goal of elimination of leprosy.

REFERENCES

Sehgal VN. Histoid Leprosy. In: Kumar B, Kar HK, editors. IAL Textbook of Leprosy. 2nd ed. New Delhi: Jaypee Brothers Medical Publishers (P) Ltd; 2016. p. 268–77.

Alious Z, Sbai M, Elhaouri M, Bouzidi A, Boudi O, Ghfir M, et al. Histoid leprosy with erythema nodosum leprosum. Acta Leprol. 2003; 12(3): 107-11.

Sehgal VN. Spontaneous Appearances of Papules,Nodules, and/or Plaques:A Prelude to Abacillary, Paucibacillary, or Multibacillary Histoid Leprosy. Skinmed Dermatol Clin. 2006:139-41.

Rao AG. Borderline Tuberculoid Leprosy Associated with Histoid Leprosy. Indian J Dermatol. 2016;61(5):580.

Sun J, Tu P, Yi S, Fu W. Type I Lepra Reaction as the Presenting Sign of Histoid Leprosy. Ann Dermatol. 2017;29(5):646-8.

Singh N, Kumari R, Gupta D, Thappa DM. Type 1 lepra reaction in histoid leprosy. International Journal of Dermatology. 2015;54:564–7.

Alious Z, Sbai M, Elhaouri M, Bouzidi A, Boudi O, Ghfir M, et al. Histoid leprosy with erythema nodosum leprosum. Acta Leprol. 2003;12(3):107-11.

Kolaparambath BA, Rajagopal R. A case of histoid leprosy in a HIV infected person on HAART not responding to conventional MB-MDT. Indian J Lepr. 2014;86(1):15-8.

Kaur I, Dogra S, De D, et al. Histoid leprosy: a retrospective study of 40 cases from India. Br J Dermatol. 2009;160:305–10.

Hali F, Benchikhi H, Azzouzi S, Zamiati S, Latifi A, Sbai M. Familial histoid leprosy. Ann Dermatol Venereol. 2011;138(1):42-5.

Kalla G, Purohit S, Vyas MC. Histoid, a clinical variant of multibacillary leprosy: Report from so called endemic areas. Int J Lepr Other Mycobact Disease. 2000;68:267-71.

Misch EA, Berrington WR, Vary JC, Hawn TR. Leprosy and the Human Genome. Microbiol Mol Biol Rev. 2010;74(4):589–620.

Pandey P, Suresh MSM, Dey VK. *De Novo* Histoid Leprosy. Indian J Dermatol. 2015;60(5):525.

Cabic E, et al. Alternative Method for Histopathological Detection of Mycobacterium Tuberculosis and Mycobacterium Leprae using a Modified Acid-Fast Technique. Philippine Journal of Pathology. 2018;3(1).

Adiga DSA, Hippargi SB, Rao G, Saha D, Yelikar BR. Evaluation of Fluorescent Staining for Diagnosis of Leprosy and its Impact on Grading of the Disease: Comparison with Conventional Staining. J Clin Diagn Res. 2016 Oct;10(10):EC23–EC26.