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Diffuse multibacillary leprosy with Lucio's phenomenon in east Lombok, west Nusa Tenggara, Indonesia

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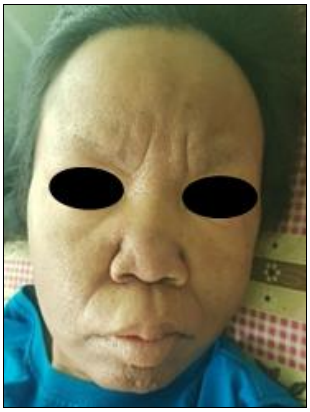
Abstract
Introduction: Lucio's phenomenon (LP) is a rare leprosy reaction characterized by severe necrosis of the skin. LP is a variant of type II leprosy reaction by the involvement of immune complexes. LP is usually found in Mexico and Central America. This reaction is especially found in diffuse non-nodular lepromatous leprosy who have not received or completed leprosy treatment.
Case: A 36 years woman with newly diagnosed Lucio's phenomenon after initially being misdiagnosed with fungal infection for several months.
Discussion: In clinically, LP may mimic other diseases such as mycosis and allergic reaction. Until now there is no consensus regarding LP treatment and combination MDT-MB and systemic corticosteroids are the options in this case.
Conclusion: A proper history, physical examination, and histopathology are important diagnostic approaches to avoid misdiagnosis or underdiagnosis in LP cases especially in leprosy endemic regions. The combination of MDT-MB and systemic corticosteroids in FL gives a good response.

Keywords: Lucio's phenomenon, leprosy, ulcer, Indonesia

Introduction
Leprosy or Morbus Hansen is a chronic granulomatous infection caused by the obligate intracellular acid-fast bacillus (AFB) *M. leprae* [1, 2]. Lucio's phenomenon (LP) is a rare leprosy reaction characterized by severe necrosis of skin [2, 3]. It was first reported by Lucio and Alvarado in 1852 in Mexico as diffuse necrotic skin lesions in patients with non-nodular lepromatous leprosy. In 1948, Latapi and Zamora named Lucio's phenomenon for this reaction after finding histopathologic features vasculitis and skin necrosis in the same type of leprosy [2-5]. LP are commonly found in Mexico and Central America, but several cases have also been reported in United States, Peru, Hawaii, Brazil, India, Malaysia, and Indonesia [2-9]. Although Indonesia is a leprosy endemic region and the 3rd highest leprosy cases in the world with 17.439 cases in 2019, LP still rarely reported.[10] LP is defined as a type 2 variant of leprosy reaction [2, 5, 7, 9]. LP often occurrence patients with diffuse non-nodular leprosy or Lepromatous Leprosy (LL) and Borderline Leprosy (BL) who do not receive or complete a leprosy treatment [2-6]. We reported the first case of LP in East Lombok Regency, West Nusa Tenggara in a patient who had never been diagnosed with leprosy.

Case
A 36 years woman from Sasak tribe has been referred by Public Health Centre to emergency room of Regional Hospital R. Soedjono Selong with a painful blackish-red ulcer on both legs that already appeared for one weeks. Based on the patient's history the first lesion appeared approximately 2 years ago as brownish white patches that feel dry and slightly itchy on both legs and arms. Three months ago, she was diagnosed with a fungal infection but did not show any improvement after receive several treatments. Six days before hospitalization the skin patches on the legs suddenly became bigger and spreading over the dorsum of the foot followed by a black colour changing. The next day, on the surface of the skin lesion, some vesicle started to appear, which quickly eroded causing painful ulcer. Based on all those symptoms she was suspected to have a drug allergic reaction and was referred to the hospital. The patient also admitted that one year ago the both of her feet sometimes feel numbness.

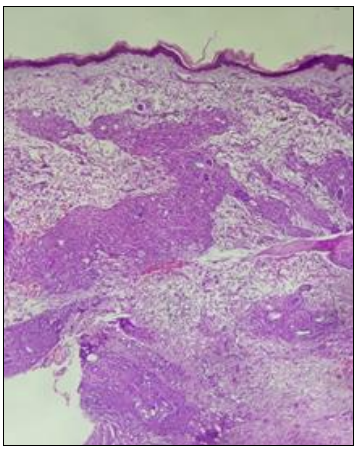
On physical examination we found subfebrile fever (37.8 ℃). The patient has leonine facies, madarosis at superciliary and eyebrows, saddle nose and also infiltrates in her eyebrows, nose, and chin. On both arms, there were hypo-hyperpigmented anaesthesia patch lesions covered with fine scales. There were extensive deep blackish-red necrotizing ulcers with irregular shaped and overlying brown sludge predominantly over the legs and dorsum of the foot. Some vesicles in the lesion have been eroded-excoriated and also overlying serous crust. No nerve enlargement was found. The patient has never been diagnosed with leprosy or received leprosy treatment.



Picture 1: Leonine facies with superciliary madarosis, saddle nose, and infiltrate in eyebrows, nose, and chin

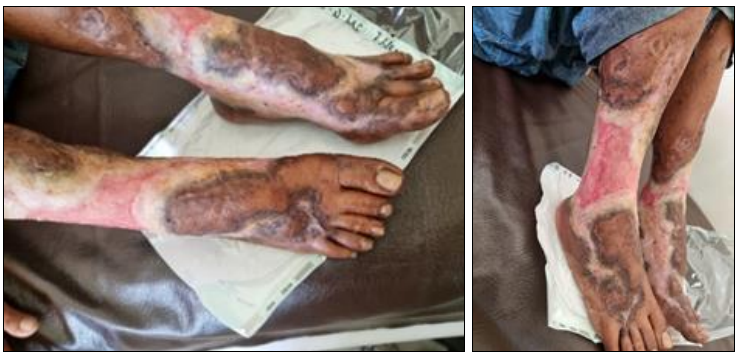
Picture 2: Blackish-red necrotizing ulcer with irregular shape and sludge

Based on skin slit smear examination from earlobes revealed bacterial index (IB) of +3. Histopathological from skin biopsy with Hematoxylin-Eosin (HE) staining showed macrophage granulomas, Virchow cells, vasculitis, obliteration with thrombosis of superficial and mid-dermis and extravasation of red blood cells. Complete blood count revealed hemoglobin 8.6 g/dL, white blood cell, coagulation profile, liver function test, kidney function test, blood sugar and chest X-ray were normal.



Picture 3: Granuloma macrophages, Virchow cell, dan thrombosis vasculitis in dermis

The patient was diagnosed with lepromatous leprosy (Ridley-Jopling classification) with Lucio's phenomenon. Patients received World Health Organisation (WHO) Multi Drug Treatment-Multibacillary (MDT-MB) therapy with rifampicin (600 mg/month), dapsone (100 mg/day), and clofazimine (300 mg/month) and 50 (mg/day) for 12 months in combination with systemic corticosteroid (prednisone 1 mg/kg/day) for 2 weeks and then tapered off periodically. Intravenous antibiotic Ceftriaxone was given to prevent secondary infection. Wound debridement followed daily dressing with sodium chloride 0, 9% solution and fusidic acid 2% cream. Follow-up once a week and one month after treatment, the skin lesion showed improvement with hypochromic atrophic stellate scar (central achromic scar) surrounding by hyperpigmented borders.



Picture 4, 5: After one month treatment showing healing of the ulcer with central achromic scar and hyperpigmented border

Discussion

Lucio's phenomenon is a rare variant of type II leprosy reaction where immune complexes are involved (hypersensitivity reaction type III). LP often is seen in non-nodular diffuse lepromatous leprosy or LL and BL who have not been treated or completed for leprosy treatment. Pathogenesis of LP is unclear, several hypothesis revealed invasion of a large number of AFB in blood vessel causes the formation of an immune complex, then triggers vasculitis and endothelial proliferation, resulting in

obliteration and thrombosis that causes tissue necrosis. Sharma *et al*. [2] reported deposits immunoglobulin and complement in vascular and perivascular have been founded in the lesion of LP. Generally onset between 1 and 3 years after manifestation of disease, especially for multibacillary leprosy. In other hands, a case study by CheYa *et al*. [3] showed that the onset of LP may occur more than 5 years in the course of the disease.

The manifestation of skin lesions initially is purpura or plaques or red macular infiltrates that are commonly found on the extremities with vesicles or bullae. Then the lesion has erosion and becomes an ulcerated irregular shape with central necrosis surrounded by hyperpigmentation area which may be accompanied by pain and secondary infection. Lesions are usually found in extremities and rarely affecting the face and trunk [3-6]. The skin necrosis without vesicles or ulcers is unusual, only 3 out of 12 patients with LP.[4] Fever may occur if they experienced secondary infection or anemia [5, 7].

Histopathological examination was obtained from a skin biopsy and will show are a granulomatous inflammation (foamy macrophage cells or Virchow cells), epidermal and dermal necrosis, endothelial proliferation, vasculitis, obliteration, occlusion of the lumen and thrombosis of medium sized blood vessels in the superficial and mid dermis. In AFB and Fite-Farco staining, *M. leprae* colonization was found on endothelial, interstitial, and foam cells [2-6, 11-12]. A study conducted in Malaysia reported that up to 44% of LP cases seen initially in primary care were misdiagnosed [3]. Pradana *et al*. [6] also reported that the patient in their case report initially were diagnosed with drug allergic reactions. Our patient was initially diagnosed with fungal infection and then being suspected with a drug allergic reaction in Public Health Centre. Other differential diagnoses of LP include vasculonecrotic erythema nodosum leprosum (ENL), cutaneous vasculitis, pyoderma gangrenosum, connective tissue disease, cryoglobulinemia, cutaneous tuberculosis, and dermal mycoses [2, 5, 11]. ENL is most commonly confused with LP but the clinical and histopathological features may aid in differentiating the two, which ENL is frequently associated with constitutional symptoms, neuritis, and visceral involvement (iridocyclitis, hepatitis, and orchitis). Then histopathologic show vasculitis more often shows in small-medium blood vessels of the dermis with predominantly neutrophil infiltrate in the dermis and hypodermis [2].

There are three criteria for defined LP by skin ulcers, vascular thrombosis, and invasion of M. leprae into endotel [7]. The study by Velarde-Felix *et al*. [8] also characterized FL with existing immune complexes, necrotizing vasculitis on superficial and medium-sized blood vessels, diffuse skin infiltration, necrosis dermal, and sometimes with systemic reactions. In our case, the patient presented almost all characteristics of LP.

There is no consensus regarding LP treatment where currently treatment is using empirical study, based on existing case reports. Many studies were reported MDT-MB WHO regimen was showing effectiveness for treating LP. Using corticosteroids as LP therapy is still controversial, but several studies have shown good results in accelerating the healing process. The dosage used is 0.5-1 mg/kg/day. Systemic antibiotics are allowed if a secondary infection is suspected [2-5, 11]. Sharma *et al*. [2] reported short-term administration of systemic corticosteroids can control the immune reaction in the early phase of the disease, especially in severe cases. In addition, prognosis and mortality are variable due to not many of the cases that have been reported [3, 5]. Gao *et al*. [11] result combination MDT-MB with prednisolone 1 mg/kg/day caused partial resolution of skin lesion. Some literature reported thalidomide and pentoxifylline are accepted for the FL cases as well [7, 12]. Skin lesions will heal within 2-8 weeks after therapy and leaving a stellate atrophic scar or central achromic scar [2-4, 11]. We give a combination of MDT-MB and short-term systemic corticosteroids for the patient then after 1 month of treatment showed improvement. Other studies also reported that administration of the MDT-MB regimen at early diagnosis improve the prognosis, but other conditions such as extensive skin lesions, anemia, and sepsis can be aggravating the disease [5, 11].

Conclusion

LP is a variant of type 2 leprosy reaction which is an uncommon case that being reported in Indonesia. On top of that, some LP cases are misdiagnosed or underdiagnosed. It is important to receive a proper medical history and physical examination to get the right diagnostic, especially in endemic regions. Histopathological features also play an important role in establishing the diagnosis of LP and ruling out the differential diagnosis. A combination of FL therapy with MDT-MB and systemic corticosteroids gives a good result.

References

1. Salgado CD, Brito AC, Salgado UI, Spencer JS. Leprosy. In: Kang S, Amagai M, Bruckner AL, Enk AH, Morgolis DJ, McMichael AJ *et al*., editors. Fitzpatrick's Dermatology 9th Ed: McGraw-Hill Education 2019, 2892-919.
2. Sharma P, Kumar A, Tuknayat A, Thami GP, Kundu R. Lucio phenomenon: a rare presentation of hansen's disease. J Clin Aesthet Dermatol [Internet] 2019;12(12):35-38. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7002045/
3. CheYa SN, Muhamad R, Zakaria R, Ishak A, Abdullah WN. Lucio phenomenon: sequelae of neglected leprosy. Korean J Fam Med [Internet] 2021;42(3):245-249. Available form: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC8164933/
4. Mukhija A, Kumawat K, Namdeo C, Jain O. Lucio phenomenon and diffuse non-nodular lepromatous leprosy – an uncommon occurrence in India. International Journal of Scientific Research [Internet] 2021;10(6):55-56. Available from: https://www.worldwidejournals.com/international-journal-of-scientific-research-(IJSR)/article/lucio-phenomenon-and-diffuse-non-nodular-lepromatous-leprosy-an-uncommon-occurance-in-india/MzU1NjI=/?is=1&b1=5&k=2
5. Marrisa M, Rihatmadja R, Surya D, Lim HW, Menaldi SL. Lucio's phenomenon: a report on six patients in tertiary referral hospital in Indonesia. J Gen Proced Dermatol Venereol Indones [Internet] 2020:5(1);40-47. Available form: http://www.jgenprodvi.ui.ac.id/index.php/jdvi/article/view/225/pdf

6. Pradana P, Rosandi R, Daili ES, Sirait SP, Rihatmadja R *et al*. Gambaran klinis dan histopatologi fenomena lucio. MDVI [Internet] 2016:43(4);147-152. Available form: http://www.perdoski.or.id/doc/mdvi/fulltext/45/303/LAPORAN_KASUS_2.pdf
7. Rocha RH, Diniz LM, Cabral AN, Emerich PS, Oliveira MB, Amaral AC. Lucio's phenomenon: exuberant case report and review Brazilian cases. An Bras Dermatol [Internet] 2016;91(5, 1):60-63. Available form: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5324995/
8. Velarde-Felix JS, Villa GA, Cabrera LV. Lucio's phenomenon associated with Mycobacterium lepromatosis. Am J Trop Med Hyg [Internet] 2016;94(3):483-484. Available form: http://europepmc.org/article/PMC/4775876
9. Ramal C, Casapia M, Marin J, Celis JC, Baldeon J, Vilcarromeo *et al.*; Central for Disease Control and Prevention. Diffuse multibacillary leprosy of lucio and latapi with lucio's phenomenon, Peru. Emerging Infectious Diseases [Internet] 2017;23(11):1929-1930. Available form: https://wwwnc.cdc.gov/eid/article/23/11/17-1228_article
10. World Health Organization. Global leprosy (hansen disease) update, 2019: time to step up prevention initiatives. Wkly Epidemiol Rec 2020.
11. Gao W, Chen Z, Jiang H, Shi Y, Zhang W, Wang H. Diffuse multibacillary leprosy patient with lucio's phenomenon and positive anticardiolipin antibody misdiagnosed as lupus erythematosus panniculitis in the people's Republic of China. Infect Drug Resist 2019;12:741-744. Available form: https://pubmed.ncbi.nlm.nih.gov/31114260/
12. Barrios GA, Pulgarin JA, Canizales CE, Zambrano DM, Montealegre FE, Cardona AM *et al*. Lucio's phenomenon: a report of two cases and review of the literature. Case Report in Internal Medicine [Internet] 2019;6(2):15-19. Available form: https://www.researchgate.net/publication/332614694_Lucio%27s_phenomenon_A_report_of_two_cases_and_review_of_the_literature