Atypical clinical manifestations of leprosy in Indonesian male adolescent: a case report

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Leprosy, also known as Hansen's disease, is a chronic infectious disease with high prevalence, but often neglected in Indonesia. Indonesia ranks the 3rd worldwide, after India and Brazil, with 17,439 new cases reported in 2019. This disease is caused by Mycobacterium leprae, an acid-fast, rod-shaped bacillus, which mainly affects the skin, peripheral nerves, upper respiratory tract mucosa, and eyes. Early diagnosis of the disease is fundamental, because delayed treatment may lead to severe deformities and disabilities. The current multidrug treatment (MDT) for leprosy is widely available in Indonesia for free.

According to the World Health Organization (WHO), leprosy is one of 20 diseases recognised as neglected tropical diseases (NTDs), a group of disease which present significant burden amongst the poorest, often unheard communities. Eradication of leprosy is challenging because of the difficulty in diagnosis, as leprosy imitates various diseases, as well as treatment delay, high transmission, and social stigma. The regions of highest leprosy prevalence in Indonesia are in Java, Sulawesi, Maluku, and Papua.

Among 17,439 new cases in 2019, 1,861 (10.1%) were children under 15 years of age. The detection of new cases in children indicates high transmission, and lack of mechanisms to control endemic infections. The aim of this report is to share our experience in diagnosing advanced stage leprosy with atypical clinical characteristics in a male adolescent.

Keywords: leprosy; Hansen's disease; erythema nodosum leprosum; adolescent

The Case

A 16-year-old male came to the Emergency Department of Dr. Cipto Mangunkusumo Hospital, Jakarta, with the chief complaints of high fever above 39°C and painful tissue edema at the anterior compartment of the right forearm, the right hand, and right ring finger. Three months before admission, the patient had difficulty moving his right arm and it became swollen, but there was no history of trauma. The patient lived in the East Palu District of Sulawesi. He visited the primary health center, received antibiotics and analgesics, and the pain subsided for a while. One month later, the right ring finger looked swollen with...
erythematous patches; these patches also appeared on the whole body, specifically the trunk and extremities. There was no change of sensation, as well as no pain or itching of the affected areas. He had mild intermittent fever.

Four days prior to admission, the patient was referred to the Orthopedic Clinic in Dr. Cipto Mangunkusumo Hospital, Jakarta, due to his deteriorating condition: high fever, right arm pain, and an abscess on the right middle finger. His symptoms worsened so he was referred to the Emergency Department. He had no previous history of chronic disease, no history of similar disease in the family, and his basic immunization was complete. There was history of unprotected sexual intercourse seven months prior to this time.

The patient had normal nutritional status. Physical examination revealed tachycardia due to fever of 39°C, normal eye examination/vision, normal intraoral condition with no oral thrush, as well as normal heart, lungs, abdomen, and genitalia. Examination of the limbs revealed right arm limited range of motion, pain, and edema, especially in the elbow, forearm, and fingers. Skin inspection showed multiple erythematous nodules, patches, and pustules, with diameters of 1 to 3 centimeters (cm), on the trunk and extremities, which were circumscribed and discrete (Figure 1). Palpation of the right ring finger revealed edema, warmth, tenderness and hypoesthesia. Nerve palpation showed enlargement of the right auricularis magnus nerve, left ulnar nerve, and left common peroneal nerve. Laboratory results are shown in Table 1, and included leukocytosis, increased erythrocyte sedimentation rate, and increased C-reactive protein. Albumin level was not examined because the edema was localized in the right arm.
differential diagnosis included cellulitis, erythema nodosum leprosum (ENL), bacterial impetigo, Scarlet fever, systemic lupus erythematosus (SLE), sexually transmitted infections (STIs) such as syphilis. The patient was initially treated with intravenous, broad-spectrum antibiotics and analgesics.

To confirm the patient’s diagnosis, several additional examinations were performed. The venereal disease research laboratory (VDRL) and Treponema pallidum hemagglutination (TPHA) results were both negative. Gram staining of pus from the pustule showed abundant leukocytes [>30 per high power field (HPF)] and Gram-positive staphylococci (4-5 per HPF). Human immunodeficiency virus screening was non-reactive, and other results were as follows: T-cell cluster designation [CD4+: 336 cell/mm$^3$ (32%), CD8+: 179 cell/mm$^3$ (17%)], immunoglobulin (IgG) 1,457 mg/dL, and anti-streptolysin titer (ASTO): 178 IU/mL. Human immunodeficiency virus screening was non-reactive, and other results were as follows: T-cell cluster designation [CD4+: 336 cell/mm$^3$ (32%), CD8+: 179 cell/mm$^3$ (17%)], immunoglobulin (IgG) 1,457 mg/dL, and anti-streptolysin titer (ASTO): 178 IU/mL. Additional results revealed no immunodeficiency, low chance of STIs, and positive ANA result. To complete our diagnosis, skin biopsy was subjected to slit-skin smear with Ziehl-Neelsen staining. The results are shown in Table 2 and Figure 2.

Our investigation revealed the patient have 2 out of 3 cardinal signs of leprosy: (1) thickened or enlarged peripheral nerve with loss of sensation/weakness of the muscles supplied by that nerve, and (2) presence of acid-fast bacilli in a slit-skin smear. The patient was diagnosed with multibacillary (MB) leprosy subcategory borderline lepromatous (BL), with ENL. The patient underwent multidrug treatment MB regimen which consisted of rifampicin, clofazimine, and dapsone, combined with methylprednisolone, in collaboration with the Dermatology and Venereology Departments. Patient was observed in the outpatient clinic for two weeks, and sent back to his hometown. Multidrug treatment was given for 12 months duration, with good response.

**Discussion**

Leprosy is a highly endemic disease in Indonesia, ranking 3rd in new cases in the world; 10% of the new cases in Indonesia are children below 15 years old. Leprosy is considered to be a neglected disease by the WHO because it present significant burden amongst the poorest, often unheard communities and leprosy elimination programs have not been entirely successful. Problems with leprosy is diagnosis difficulty, because it can mimic other diseases, also known as the “great imitator” of diseases. In Indonesia, leprosy is mostly managed by dermatologists and venereologists, but all doctors in Indonesia should understand and maintain a healthy suspicion for this disease, due to the high prevalence and high burden of disease. To simplify the diagnosis of leprosy, the WHO recommends clinical examination, with or without slit-skin smears or pathological examination of biopsies. In clinical settings, leprosy can be diagnosed based on the presence of at least one of the three cardinal signs: (1) definite loss of sensation in a pale (hypopigmented) or reddish skin patch; (2) thickened or enlarged peripheral nerve with loss of sensation and/or weakness of the muscles supplied by that nerve; or (3) presence of acid-fast bacilli in a slit-skin smear.

<table>
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<tr>
<th>Table 2. Skin biopsy acid-fast bacilli smear showed presence of acid-fast bacilli on skin lesions from various parts of the body</th>
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<tbody>
<tr>
<td>Biopsy location</td>
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<tr>
<td>Right ear</td>
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<td>Left ear</td>
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<td>Back</td>
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<td>Left upper limb</td>
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[notes: a +1 result was interpreted as 1-10 bacilli in 100 fields; +2 result was interpreted as 1-10 bacilli in 10 fields]

**Figure 2. Ziehl-Neelsen staining showing acid-fast bacilli**
Our patient showed chronic and progressive disease with signs of multiple skin lesions such as erythematous rash, nodules and edematous finger with a large pustule, right arm weakness, and enlargement of the peripheral nerves. Such clinical signs should trigger a diagnosis of leprosy that can be treated promptly to prevent further disability. Multiform skin lesions, which in our opinion causing difficulty in diagnosing leprosy in our patient can be explained by the ENL reaction, an immunological complication affecting approximately 10% of patients with borderline lepromatous (BL) leprosy. We want to emphasize that a positive ANA result is common in people with leprosy, which may confuse the leprosy diagnosis, so it needs to be interpreted carefully.

The final diagnosis for our patient was multibacillary leprosy, borderline leprosy, with erythema nodosum leprosum.

Treatment of leprosy included a three-drug regimen of rifampicin, dapsone, and clofazimine, for durations of 6 months for paucibacillary (PB) leprosy and 12 months for MB leprosy. Corticosteroids must be given to patients with ENL, to suppress the inflammation process. Patients must be closely monitored after treatment, as another skin biopsy may be needed to confirm disease activity. Family and relatives must also be screened for leprosy; single dose rifampicin is recommended as prophylaxis for at-risk contacts.

In conclusion, leprosy is a highly endemic disease in Indonesia. Left untreated, leprosy can cause severe disabilities. Leprosy is diagnosed by hypopigmented skin lesions with loss of sensation, enlargement of peripheral nerves and muscle weakness, or slit-skin smear showing acid-fast bacilli. Positive ANA examination is common in leprosy. Treatment of leprosy consists of MDT, combined with corticosteroid for ENL cases.

**Conflict of Interest**

None declared.

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**References**