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Case Report

Childhood borderline lepromatous leprosy

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Leprosy (Morbus Hansen) is a chronic, severe infectious disease caused by the Mycobacterium leprae that mostly affects the skin, mucosa, eyes, and nerves. Despite the availability of effective treatment, leprosy has become a major public health problem in many developing countries. **[Paediatr Indones. 2024;64:356-62; DOI: https://doi.org/10.14238/pi64.4.2024.356-62**].

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For twenty-two years, leprosy has been declared "eliminated as a public health hazard", yet new cases continue to emerge in endemic areas. There were 127,558 new leprosy cases detected globally in 2020, according to official figures from 139 countries in the six WHO regions. These data includes 8,629 children below 15 years of age. The new case detection rate among the child population was recorded at 4.4 per million child population. Among the new cases, 7,198 new cases were detected with grade 2 disabilities (G2D), and the new G2D rate was recorded at 0.9 per million population.[1] Because of their underdeveloped or neonatal immunity and exposure to intrafamilial contacts, children tend to be the most population to *Mycobacterium leprae* infection. Leprosy is a master imitator, presenting as subtle hypopigmented patches on the face, arms, and the cold parts before spreading extensively across the skin and causing neuromuscular symptoms such as sensory loss and muscle weakness. As a result, in locations where leprosy is still prevalent, it should be considered a differential diagnosis even in nonendemic areas, not just by dermatologists but also by doctors, neurologists, and pediatricians who care for children and adolescents.[2-4]

The case

We present a case of leprosy in a 9-year-old Indonesian male patient who came to the Haur Gading Health Center, South Kalimantan, with the chief complaint of three stiff fingers on his left hand four years ago. The patient's parents also complained of hypopigmented patches, which were neither itchy nor painful, on the patient's back and buttocks since five years ago. The patient also complained of a bent finger three years ago. The patient lives with his parents and often meets his mother's sister, who passed away five years ago. His mother's sister was diagnosed with leprosy but does not take medication regularly. Meanwhile, family members who lived with the patient did not have complaints before. The patient had received complete basic immunizations, including the *Bacillus Calmette-Guerin* (BCG) vaccine. The patient's mother also denied history of persistent cough and other chronic diseases.

On general physical examination, no abnormalities were found. The patient was found to have compos mentis consciousness and good general condition, weighing 17 kg. Eyebrows were within normal limits, and eyelids could open and close

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perfectly. The examination of the nose, ears, and throat found no abnormalities. There were no infiltrates in the right and left ear lobes. The extremities were warm, and there was no edema. Regional lymph node enlargement was not found. Dermatological status of vertebral location, right and left lumbar, and sacral area showed hypopigmented plaque efflorescence, multiple, well-defined, geographic shape, and size varied from 0.5 × 1 cm to 1 × 1.5 cm. **Figure 1** and **2** show the multiple hypopigmented patches on the left and right buttocks and back. Sensibility examination in leprosy lesions found a decrease in the sensation of pain, touch, and temperature in the lesion. On the tips of the fourth and fifth fingers of the left hand, there were reddish nodules and thickened nails (**Figure 3**).

Nerve examination revealed thickening and enlarged of the ulnar nerve and median nerve on the left hand. The left hand's third, fourth, and fifth fingers look stiff when moved. The voluntary muscle test (VMT) showed muscle weakness in the thenar and hypothenar muscles and numbness in the anterior fingers of the left hand (**Table 1**). From the history and physical examination of the patient, the patient was diagnosed to be suspected of borderline lepromatous (BL) leprosy. The patient was planned for a follow-up examination in the form of a slit-skin smear and laboratory examination. In the follow-up, the skin slit smear showed the presence of acid-fast bacilli (AFB) scrapings on the skin of the right and left ear lobes with bacterial index +3 (**Figure 4**). **Table 2** shows the laboratory examination results.

From the history, physical examination, and skin slit smear examination, the working diagnosis was confirmed as a borderline lepromatous type of leprosy. The management given is multidrug therapy (MDT) multibacillary for children (rifampicin 300 mg per month, dapsone 25 mg daily, and clofazimine 100 mg per month, followed by 50 mg every 2 days) for 12 months. The patient's family was given education about the disease, the examination results, the treatment, and a one-month control was recommended. The patient was referred to physiotherapy center regularly. Periodical observations after the patient received treatment did not find new spots on the patient's skin; some previous hypopigmented patches seemed to fade, especially on the back. The fingers of the left hand are still stiff, but the nodules are slightly faded (**Figure 5**). The patient denied any history of fever. Regular physiotherapy for the patient is still ongoing.

Discussion

Leprosy is a disease that can infect people of all ages. The prevalence of leprosy in children can serve as an indicator of the disease's prevalence in the general

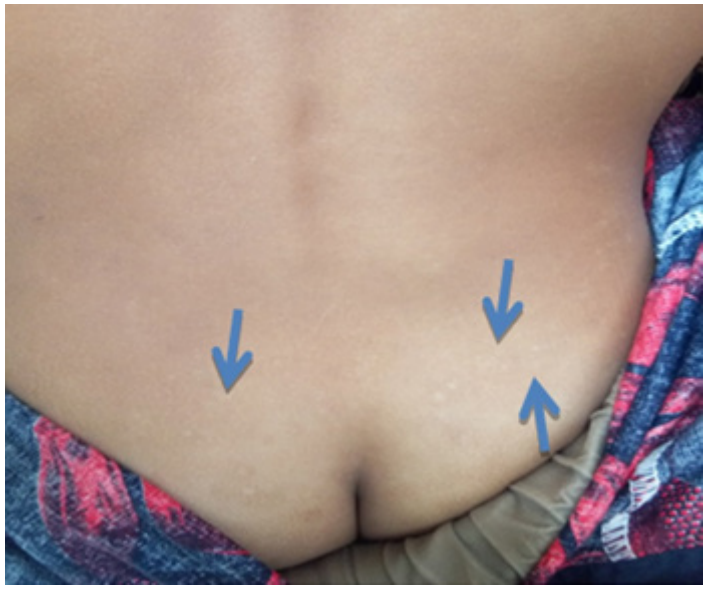

Figure 1. Multiple hypopigmented patches on the left and right buttocks, which were not itchy and pain

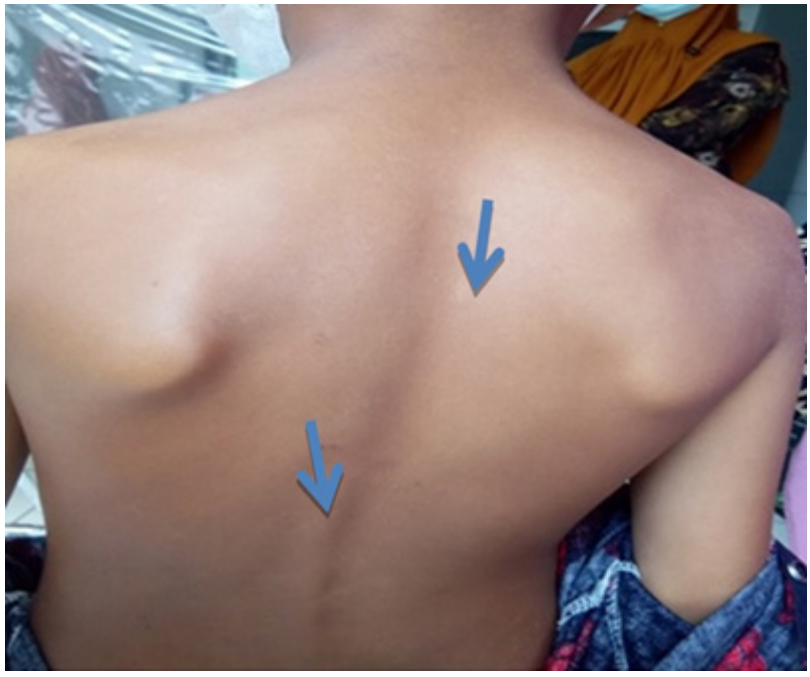

Figure 2. Multiple hypopigmented patches on the back, which were not itchy and pain

Figure 3. Red nodule lesion on left hand before treatment

population, and a tool for determining how the disease is transmitted.[1,4,5] The incubation period for leprosy ranges from 2 to 4 years, although 3 months to 40 years has been reported.[2] Children are more susceptible to leprosy because their immune system is not yet fully developed. The onset of this disease in children is between 5 and 14 years, with the same prevalence in boys and girls.[4-7] Incidence in children under the age of one year has been documented as two cases of a 6-month-old child with leprosy that was confirmed

Table 1. Sensoric and motoric tests

Vertebral segments	Sensory Test		Motoric Test	
	Right	Left	Right	Left
C5,6,7	Normal	Normal	Normal	Normal
C5,6	Normal	Normal	Normal	Normal
C7,8	Normal	Abnormal	Normal	Abnormal
C8 T1	Normal	Abnormal	Normal	Abnormal
T4	Normal	Normal	Normal	Normal
T10	Normal	Normal	Normal	Normal
T12	Abnormal	Abnormal	Normal	Normal
L1,2,3,4	Abnormal	Abnormal	Normal	Normal
L4,5	Abnormal	Abnormal	Normal	Normal
S 1,2	Normal	Normal	Normal	Normal
S 2,3,4	Normal	Normal	Normal	Normal

C=cervical; T=thoracal; L=lumbar; S=sacral

Table 2. Laboratory examination results

Hematology	Result	Reference value
Hemoglobin, g/dL	10.1	13.50-17.50
Hematocrit, %	31	41.00-53.00
MCV, fL	80.6	80.00-96.00
MCH, pg	26.3	28.00-33.00
Erythrocyte, $x10^6/mm^3$	3.84	4.50-5.90
Leucocyte, $x10^3/mm^3$	6.78	4.1-11.0
Neutrophil, $x10^3/mm^3$	2.52	2.5-7.5
Lymphocyte, $x10^3/mm^3$	3.19	1.0-4.0
Monocyte, $x10^3/mm^3$	1.07	0.1-1.2
Thrombocyte, $x10^3/mm^3$	325	150-440
Random blood glucose, g/dL	102	<200

by histopathological examination.[8]

M. leprae bacteria are likely to enter the host through the skin and the upper respiratory tract.[9] Close contact with people with leprosy poses a significantly greater risk than those who do not live at home. The possibility of contracting the leprosy disease increases 4 times if there is contact with leprosy sufferers in the surrounding environment; the risk becomes 9 times greater in household contact and increases if the contact is a multibacillary type of leprosy patient.[10] In children, the source of leprosy infection is obtained from the sufferers with the untreated multibacillary type of leprosy in the family or community.[10,11] In a retrospective study conducted

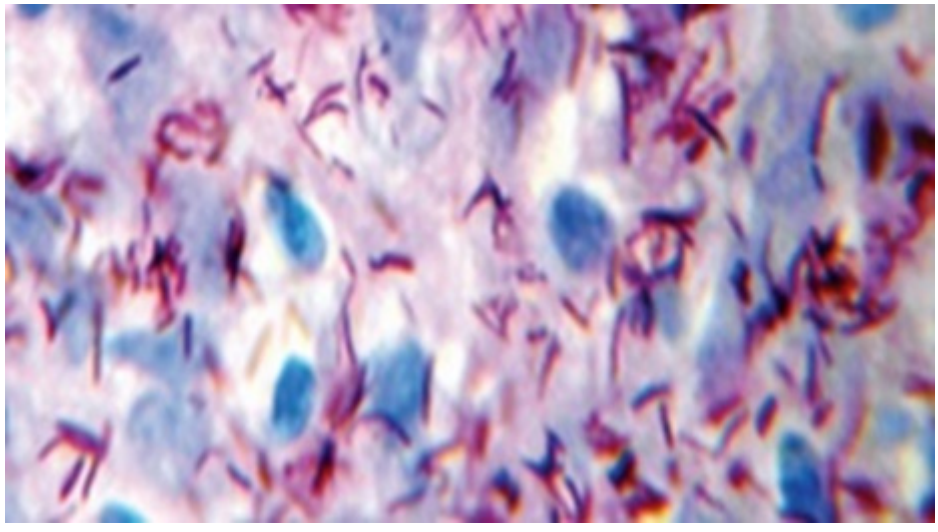

Figure 4. Slit-skin smear result showing staining, slender bacilli

Figure 5. Fading nodular lesion on left hand after 1 month of treatment, though the fingers of the left hand are still stiff

in India, more than one-third of leprosy cases in children (35%) had household contact with leprosy sufferers.[1,11]

The administration of the *Bacillus Calmette-Guerin* (BCG) vaccine as protection against *M. leprae* infection showed varying results. The effectiveness of the vaccine as protection against leprosy reached an average of 26%. A study in Brazil with a large sample size showed that the protective effect of this vaccine was 56% significant in the incidence of contact leprosy, with protection against multi-bacillary leprosy at 89% for the age under 5 years, while the protective effect was not found in older children.[12] This result indicates a protective effect of the BCG vaccine against the incidence of multibacillary type leprosy.[13] However, several factors also play a role in the incidence of leprosy and the type of leprosy. These include genetic, nutritional, and environmental factors (living in endemic areas).[14,15] Focusing on environmental factors, based on a study in Flores, 4,774 populations lived in the study area, of which 4140 were detected with leprosy, where the figure reached 87%. While 39% were multibacillary leprosy and 61% were single lesion paucibacillary or 2-5 lesions.[14] In our case, the patient had been immunized with the BCG vaccine as an infant. Several factors that cause the patient to get a multibacillary type of leprosy infection are the patient living in a leprosy endemic area, genetic factors, and the host's susceptibility to germs. The patient also contacted his mother's sister intensely, who suffered from multibacillary leprosy.

The diagnosis of leprosy was established based on the cardinal signs of leprosy through clinical examination, supported by AFB examination on slit-skin smear.[4,8] Since 1996, WHO has recommended diagnosis of leprosy based on at least one of three cardinal signs: (i) hypopigmented skin patch with loss or reduced sensation; (ii) enlarged nerve; (iii) slit-skin smear-positive for leprosy bacilli. However, several studies on leprosy diagnostics, including blood/serum samples, have been conducted. Presently, confirmatory tests for leprosy (microscopy on slit-skin smears and biopsy) are usually carried out only in referral centers.[8] In this case, the patient had a hypopigmented skin patch with loss sensation, enlarged median nerve, and skin smear showed the presence of acid-fast bacilli (AFB). In 1962, Ridley and Jopling classified leprosy based on clinical features, which include typical tuberculoid (TT), borderline tuberculoid (BT), borderline borderline (BB), borderline lepromatous (BL), and lepromatous leprosy (LL).[7,13] Based on history taking, physical examination, and follow-up examination, the patient, in this case, was diagnosed with type borderline lepromatous (BL) leprosy.

Leprosy in children is usually in the form of hypoesthetic or asymptomatic lesions, while the patient rarely complains of neural manifestations. The appearance of leprosy in children is clinically different from that in adults. Lesions are usually fewer and less defined than those in adults, and they predominate in

exposed body areas.[1,16,17] Clinical features of leprosy type BL (borderline lepromatous), classically, are that the lesion begins with the macula. Initially, it is in small amounts and quickly spreads throughout the body. The macula is clearer and more variable in shape. Although small, the papules and nodes are more defined, with an almost symmetrical distribution of the lesions. Normal skin areas were found between the lesions. Lesions are one and another have different sizes and shapes. Infiltrates may appear to form a plaque appearance, especially in the face and ears. Signs of nerve damage such as loss of sensation, hypopigmentation, reduced sweating, and loss of hair appear more rapidly than in type LL. Nerve thickening may be palpated at the site of predilection. The AFB examination showed that many M. leprae bacteria were found on the BL spectrum.[8,13] Histopathological examination of the type of BL leprosy showed a collection of macrophage cells. These macrophages have a foamy cytoplasm, similar to the lepromatous leprosy (LL) type. In addition, the presence of the Grenz zone can also be seen, and it is easy to find bacilli.[11,13,16]

The *World Health Organization* (WHO) divides leprosy patients into 2 groups based on clinical criteria by using the number of skin lesions and nerves involved, as well as the examination of skin smears in determining the treatment of leprosy. This division includes paucibacillary type leprosy (1-5 skin lesions) and multibacillary type leprosy (more than 5 skin lesions). In addition, patients with smear-positive are also classified as a multibacillary type of leprosy, regardless of the clinical picture.[13] In this case, the patient had more than five skin lesions, and the skin smear examination revealed a positive smear. Therefore, the patient was given multibacillary treatment.

Treatment of leprosy based on WHO criteria called multidrug therapy (MDT) consists of several antibiotics. Multibacillary (MB) leprosy was given a combination of rifampin, dapsone, and clofazimine.[7] In children aged 10-14 years, there is a special package treatment regimen distinguished from adults, with a duration of administration of 12 months. This regimen includes rifampin 450 mg monthly, dapsone 50 mg daily, and clofazimine 150 mg monthly, followed by 50 mg every 2 days.[13,18,19] The incidence of disability in children is quite low compared to adults because the duration of the disease is shorter, and the form of the disease is milder. However, the incidence of deformity increases with age and with long-standing disease.[15,19]

In conclusion, deformity and disability result from the delay in diagnosis, having a substantial influence on the physical, emotional, and financial aspects of the child and his family. In this case, leprosy in the patient was suspected of being possibly transmitted from his mother's sister, who had contact with the patient intensely. The results of bacteriological examination with Ziehl-Neelsen staining of tissue scrapings showed acid-fast bacilli. The treatment given was multibacillary multidrug therapy for children for 12 months. In the absence of an effective vaccine, early diagnosis and treatment are critical in preventing disability and deformity and reducing the physical, psychosocial, and economic burden of disease.

Conflict of interest

None declared.

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