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Case Report

Childhood borderline lepromatous leprosy: a case report

Grace Dumamawarni Hutahaean[1], Martin Susanto[1], Tiopan Napitupulu[1], Andre Marolop Pangihutan Siahaan[1], Beny Atmadja Wirjomartani[2]

[1]Faculty of Medicine, University of Sumatera Utara, Medan, Indonesia; [2]Faculty of Medicine, University of Padjadjaran, Bandung, Indonesia.

ABSTRACT

Background. Leprosy in children is a strong indicator of the recent failure of leprosy control and disease transmission programs. For twenty-two years, leprosy has been declared 'eliminated as a public health hazard,' yet new cases continue to emerge in endemic areas. The new case detection rate among the child population was recorded at 4.4 per million children. Because of their underdeveloped or neonatal immunity and exposure to intrafamilial contacts, children tend to be the most vulnerable population.

Case. We present a case of the borderline lepromatous type of leprosy in a 9-year-old Indonesian male patient with the chief complaint of three stiff fingers on his left hand that began four years ago and hypopigmented patches on the back and buttocks that began five years ago. In this case, there was a history of leprosy in his mother's sister, who had died. Leprosy in the patient was suspected of possibly being transmitted from his mother's sister who had intense contact with the patient. The results of bacteriological examination with Ziehl-Neelsen staining of tissue scrapings found acid-fast bacilli. He was treated with a multibacillary multidrug regimen for 12 months. Periodical observations after the patient received the treatment revealed no new spots on the patient's skin, some of the previous hypopigmented patches seemed to fade, especially those on the back.

Conclusions. 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 the disease.

Key words: childhood, borderline lepromatous, Morbus Hansen, leprosy.

Leprosy (Morbus Hansen) is a chronic severe infectious disease caused by *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. 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 World Health Organisation (WHO) regions. This includes 8,629 children under 15 years. The new case detection rate among the child population was recorded at 4.4 per million children. 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 vulnerable population to *Mycobacterium leprae* infection. Leprosy is a master imitator, presenting as subtle hypopigmented patches on the face, arms, and cold parts of the skin 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]

✉ Andre Marolop Pangihutan Siahaan
andremarolop@usu.ac.id

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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 which had started 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 that began five years ago. The patient also complained of a bent finger that occurred three years ago. The patient lived with his parents and often met his mother's sister, who had passed away five years ago. His mother's sister was diagnosed with leprosy but had not taken medication regularly. Meanwhile, family members who live with the patient did not have any complaints. The patient had received complete basic immunizations including the Bacillus Calmette-Guerin (BCG) vaccine. A history of persistent coughing and other chronic diseases was denied by the patient's mother. The patient had not sought doctor's advice or received any medication before.

On general physical examination, no abnormalities were found. The patient was found to have compos mentis consciousness and a good general condition, weighting 17 kg. Eyebrows were normal and eyelids could open and close 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 their sizes varied from 0.5 × 1 cm to 1 × 1.5 cm. Fig. 1 and Fig. 2 show the multiple hypopigmented patches on the left and right buttocks and back. A sensory examination of the leprosy lesions found a decrease in the sensations of pain, touch, and temperature on the lesions. On the tips of the fourth and fifth fingers of the left hand, there were reddish nodules and thickened nails (Fig. 3). A nerve examination revealed thickening and enlargement of the ulnar nerve and median nerve of the left hand. The third, fourth, and fifth fingers of the left hand look stiff and were 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 I). From the history and physical examination the patient was suspected of having borderline lepromatous (BL) leprosy. The patient was scheduled 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-

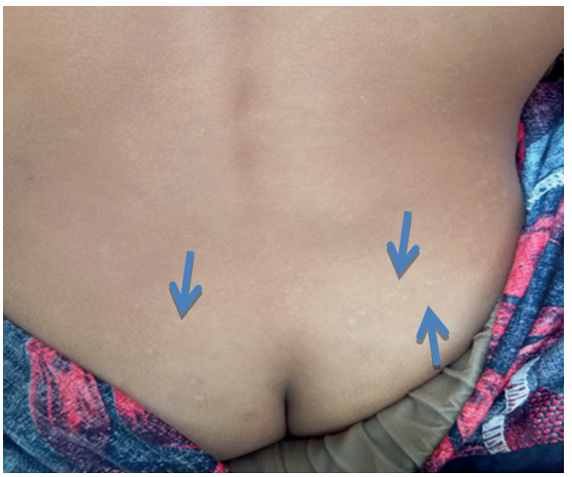

Fig. 1. Multiple hypopigmented patches on the left and right buttocks, which were not itchy and pain.

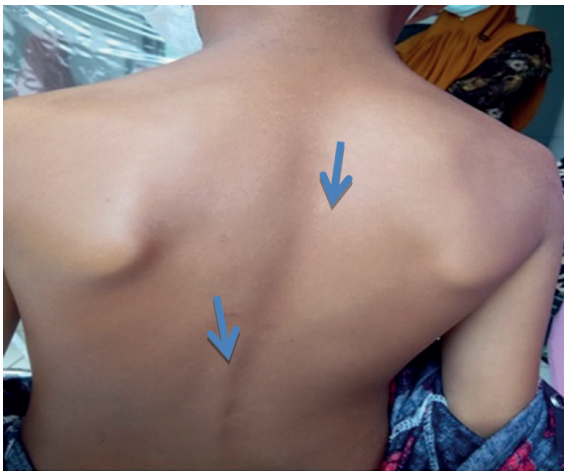

Fig. 2. Multiple hypopigmented patches on the back, which were not itchy and pain.

Fig. 3. Red nodule lesion on left hand before treatment.

fast bacilli (AFB) scrapings on the skin of the right and left ear lobes with a bacterial index of +3 (Fig. 4). The laboratory examination results revealed that the hemoglobin value decreased to 10.1 g/dl, while the white blood cell (WBC) and platelet counts were normal ($6.78\times10^3/mm^3$ and $325\times10^3/mm^3$). No abnormality was found in the blood glucose test (102 g/dl).

Based on the skin slit smear examination strengthened by the previous history and physical examination, the working diagnosis was confirmed as a borderline lepromatous type of leprosy. The prognosis for leprosy is good, as long as the patient has an early diagnosis and treatment. The management was given

Table I. Sensory and motor tests.

	Sensory Tests		Motor Tests	
	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
S1,2	Normal	Normal	Normal	Normal
S2,3,4	Normal	Normal	Normal	Normal

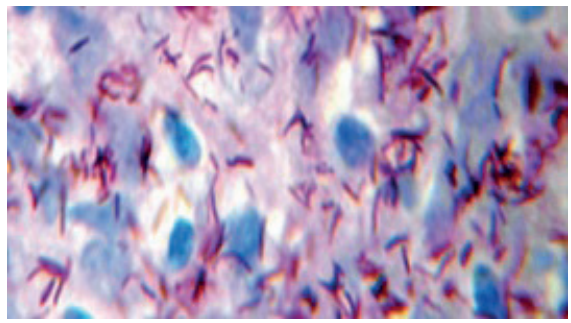

Fig. 4. Slit-skin smear result showing staining, slender bacilli.

as multidrug therapy (MDT) multibacillary (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 results of the examination, the treatment given, and a one-month regular control was advised. The patient was referred for regular physiotherapy. Periodical observations after the patient received treatment did not find new spots on the patient's skin, some of the previous hypopigmented patches seemed to fade, especially on the back. The fingers of the left hand were still stiff but the nodules had slightly faded (Fig. 5). The patient's general condition was better, and he acknowledged that all signs and symptoms had improved. The patient denied any history of fever or any adverse effects of the treatment. Regular physiotherapy for the patient is still ongoing.

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

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 population, as well as a tool for determining how the disease is transmitted.[1,4,5] The incubation period for leprosy ranges from 2 to 4 years, although an incubation period of 3 months to 40 years has been reported.[2] Children are more susceptible to leprosy because their immune systems are not yet fully developed. The age of 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 by Brubaker, Meyers, and Bourland, who published two cases of a 6-month-old child with leprosy that were confirmed by histopathological examination.[8]

M. leprae bacteria are likely to enter the host through two routes: 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 in India, more than one-third of leprosy cases in children (35%) had household contact with leprosy sufferers.[1,11]

The administration of the 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 children under 5 years, while the protective effect was not found in older children.[12] This 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 and type of leprosy. These factors include genetic, nutritional, and environmental factors (living in endemic areas).[14,15] In terms of environmental factors, according to Bakker et al's research in Flores, 4,774 people lived in the study region, of which 4,140 had leprosy, a figure that reached 87%. Where 39% were found to be multibacillary leprosy and 61% were single lesion paucibacillary or 2-5 lesions.[14] In this case, the patient was immunized with the BCG vaccine as an infant, and several factors contributed to the patient becoming infected with a multibacillary type of leprosy, including the patient living in a leprosy endemic area, genetic factors, and susceptibility to germs. The patient also made frequent contact with his mother's sister, who had multibacillary leprosy.

The diagnosis of leprosy was established based on the cardinal signs of leprosy through clinical examination, supported by AFB examination on a 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 on blood/serum samples have been carried out. 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 a loss of sensation, enlarged median nerve, and skin smear showed the presence of 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 the patients' history, physical examination, and follow-up examination he was diagnosed with BL type 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 aspects of leprosy type BL include the fact that the lesion typically begins with the macula. Small amounts at first, then rapidly spread throughout the body. The macula is more distinct and varies in shape. Although small, the papules and nodes are more defined, with the lesions distributed fairly symmetrically. Normal skin can be discovered between the lesions. Lesions differ in size and shape from one another. Infiltrates may show as plaques, particularly in the cheeks and ears. Nerve damage symptoms such as loss of feeling, hypopigmentation, decreased sweating, and hair loss emerge faster 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 as in the LL type. In addition, the presence of the grenz zone can also be seen and it is easy to find bacilli.[11,13,16]

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 leprosy 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. The diagnosis was confirmed by skin smears taken from the ear lobes which showed the presence of acid-fast bacilli. However, no biopsy was taken from the skin lesions. Hypopigmented macules and patches as seen in this boy were very nonspecific, especially in dark-skinned individuals. Many skin diseases can be listed in the differential diagnosis of such hypopigmented lesions in children, including postinflammatory hypopigmentation. Therefore, in the absence of histopathological confirmation, these lesions can not be defined as skin lesions of leprosy with certainty.

Treatment of leprosy based on WHO criteria which is called MDT, that consists of several antibiotics. Multibacillary leprosy is given a combination of rifampin, dapsone, and clofazimine.[7] In children aged 10-14 years, there is a special package treatment regimen that is 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] 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.[15,18]

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 intense contact with the patient. The results of bacteriological examination with Ziehl-Neelsen staining of tissue scrapings found 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.

Ethical approval

The patient agreed and signed informed consent regarding publishing the case in an academic journal without exposing his identity.

Author contribution

The authors confirm contribution to the paper as follows: study conception and design: GDH, MS, RDTN, AMPS, BAW; data collection: GDH, MS, RDTN, AMPS, BAW; analysis and interpretation: GDH, MS; draft manuscript preparation: GDH, MS, RDTN, AMPS, BAW. All authors reviewed and approved the final version of the manuscript.

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Conflict of interest

The authors declare that there is no conflict of interest.

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