

An unusual case of multibacillary leprosy mimicking prurigo nodularis

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Abstract

Leprosy is a chronic infectious disease with a wide range of clinical manifestations. The early diagnosis of leprosy is a worldwide challenge. We present a case of leprosy with unusual severe pruritus and generalized excoriated papules and nodules.

KEYWORDS

leprosy, multibacillary, prurigo

1 | INTRODUCTION

Leprosy, a chronic granulomatous infection, is caused by *Mycobacterium leprae*. It commonly affects the skin and nerves. The clinical manifestation of leprosy is highly variable, and it is a disease known as the “great imitators.” Therefore, it might be mistaken for other skin diseases. Since multidrug therapy (MDT), the prevalence of leprosy has decreased to <1 in 10 000. However, there are still newly diagnosed and relapsed cases.^{1,2} Herein, we present an unusual case of leprosy that appears worthy of record since it presented with features that could lead to a misdiagnosis such as severe pruritus, excoriations, papules, and nodules.

2 | CASE REPORT

A 27-year old woman presented to Razi dermatology hospital in Tehran complaining of xerosis and generalized pruritus for 8 months. It started in the third trimester of her pregnancy. Excoriated papular lesions appeared gradually on her back and extremities. She visited several physicians without any improvement in her symptoms. She gave a family

history of leprosy with a generalized skin eruption in her mother two years ago that was successfully treated and cured.

On physical examination, the patient was well-nourished with generally good health. Numerous excoriated papules and nodules were noted on her back, buttocks, and extremities. Papules were discrete and excoriated. Some of the nodules showed central depression or umbilication with elevated indurated borders (Figure 1). The skin of the upper extremities was more severely affected than the lower limbs. No pustules were noted. The intervening skin between eruptions appeared normal. No neurologic impairment was detected at the time of the presentation. The general physical examination was otherwise regular.

A punch skin biopsy from the trunk lesion was performed. The provisional differential diagnostics were prurigo nodularis, bullous pemphigoid, perforating disorders, and leprosy.

Histologic examination with Hematoxylin-Eosin stain showed a few linear granulomas, composed of lymphocytes, macrophages and plasma cells surrounding the smooth muscles, blood vessels, and eccrine glands. Also some Virchow cells in the dermis was obvious (Figure 2A, B). Bacilli of *Mycobacterium leprosy* were found in the skin biopsy stained with Zeil-Nelson stain (Figure 2C). The nasal smear also

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showed multiple bacilli with features of *Mycobacterium leprae*. A final diagnosis of multibacillary leprosy was made. MDT for multibacillary leprosy composed of Rifampin 600 mg and Clofazimine 150 mg once a month followed by Clofazimine 50 mg daily with Dapsone 100 mg was started and continued for 12 months. An improvement in skin lesions and pruritus was observed.

Her infant was 6-month-old, born full-term via vaginal delivery. He was well-nourished, well-developed and breastfeeding. Physical examination of the baby revealed a single hypopigmented patch on the back. He had no neurologic deficit. Cervical, axillary, and inguinal lymph nodes were not palpable. A nasal acid-fast smear from the infant showed numerous acid-fast positive bacilli. Therefore, the infant was diagnosed with multibacillary leprosy, although he presented with a single skin lesion. The infant was 6 kg, consequently Rifampicin 60 mg with Clofazimine 35 mg monthly and then Clofazimine 5 mg daily and Dapsone 12 mg daily were started for him for 1 year.

3 | DISCUSSION

Clinical manifestations of leprosy are significantly variable worldwide. It is variable even among patients in the same country.³ The majority of cases are found in developing countries. The rate of new case detection of leprosy is still high globally, with about 250 000 new cases being registered each year.³ However, in Mansouri and coworkers' study, the annual prevalence and new case detection rates of leprosy significantly decreased in Iran between 2005 and 2015.⁴ Therefore, leprosy is not a severe problem in our country. The current prevalence of leprosy in Iran is only 0.12 cases per 10 000 general population.⁵ Considering that Iran is attempting to eradicate this disease, careful attention to diverse leprosy presentation and complications is necessary.⁴

According to the patient's immunological response, leprosy classified into five distinct groups; tuberculoid leprosy (TT), borderline group (borderline tuberculoid (BT),

borderline-borderline (BB), borderline lepromatous (BL)), and lepromatous leprosy (LL). Borderline cases exhibit an intermediate T cell immune response. In 1997, the World Health Organization created a division to facilitate classification and treatment of leprosy in endemic areas. Leprosy is divided into two major categories. Paucibacillary is termed less than five skin lesions, while multibacillary leprosy is defined when more than five skin lesions present. The size and histological features of the lesions are not considered in this division. Multibacillary (MB) leprosy includes BB, BL, and LL.¹

As slow-growing mycobacteria cause leprosy, the symptoms can appear within a period of 3-5 years after exposure.⁶ *M. leprae* exhibits exceptionally high genetic homogeneity within different strains across the world. Thus, it may not account for such a diverse range of clinical manifestations. The complexity in the presentation of leprosy is related to the immunological status of the host.⁷ Hormonal changes in pregnancy and lactation could cause variation in the host immune response. Leprosy reactions, an immunologically mediated complications of leprosy, may occur before, during, or after treatment with MDT or may even be the presenting feature of the disease. Type 1/reversal reaction (T1R) and type 2/erythema nodosum leprosum (ENL) reaction are two major types of reactions that are recognized. T1R predominantly observed in up to 30% of patients with borderline disease. T1R is a type IV hypersensitivity reaction. Clinically, T1R presents as an inflammation of the existing lesions or onset of new skin lesions.⁸ Pregnancy is a trigger of leprosy reactions. Type 1 reaction occurs during puerperium when cellular immunity returns to normal, while the peak of the type 2 reaction is during the third trimester of pregnancy and breastfeeding. Both types of reactions persist throughout the lactation period. The risk to the mother and fetus is related to the bacterial load. During pregnancy, there is a high risk of severe leprosy symptoms. Neuritis affects around half of the mothers with leprosy, which may present as silent neuritis.⁶

Children of mothers with leprosy have an increased risk of premature delivery, low birth weight, and small placenta.

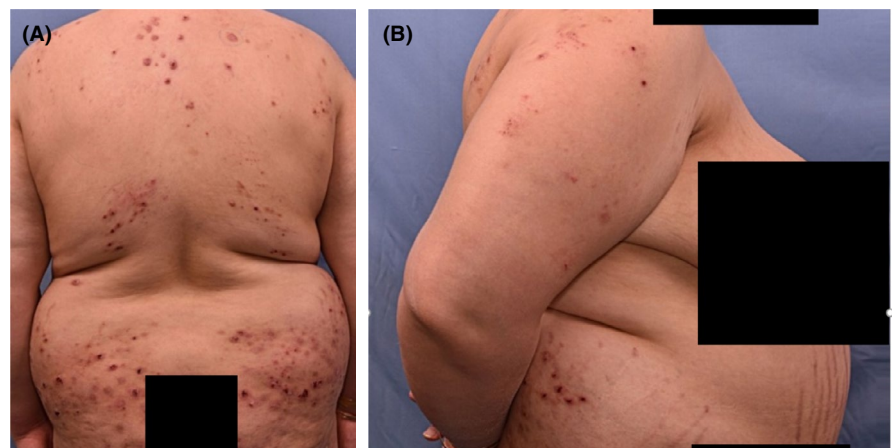


FIGURE 1 A, Multiple erythematous excoriated papules and nodules seen on the trunk and buttock. Some of them exhibit central umbilication. B, Right lateral view, multiple discrete papules, and nodules

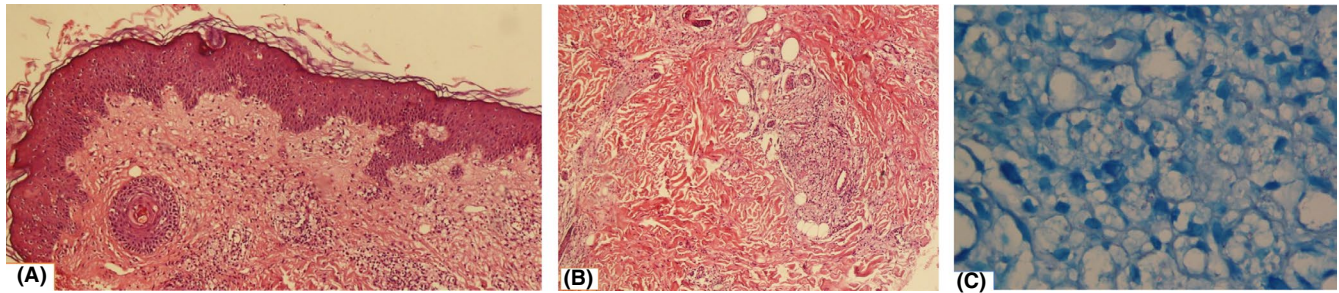


FIGURE 2 A, Few linear granulomas, composed of lymphocytes, plasma cells, and macrophages are seen in the dermis. B, Multiple granulomas mainly consist of lymphocytes, Virchow cells surrounding the eccrine ducts, blood vessels, and smooth muscles (C) Zeil-Nelson staining of the skin biopsy was positive for *M. Leprae*.

Approximately, 20% of children born from mother suffering from leprosy, develop leprosy anytime till puberty.^{6,9} Caesarian section is not necessary for mothers with leprosy. Although *M. leprae* might be found in breast milk, there is no evidence that orally ingested bacilli can cause leprosy. The main transmission route from mother to baby is airborne secretions of the mother in close contact with her baby.¹⁰

WHO recommends treating both paucibacillary and multibacillary disease with multidrug therapy, Rifampicin, Clofazimine, and Dapsone. Adult paucibacillary leprosy is treated by Rifampicin 600 mg and Clofazimine 150 mg on the first day of the month, followed by Dapsone 100 mg daily and Clofazimine 50 mg daily for 6 months. The recommendation for adult multibacillary leprosy is the same regimen for 12 months.¹¹ In most patients, these regimens effectively eradicate *M. leprae*. The infectiousness becomes unlikely after starting multidrug therapy. WHO regimen for leprosy in childhood; 10-14 years old, is Rifampicin 450 mg in addition to Clofazimine 150 mg once a month, then Dapsone 50 mg daily with Clofazimine 50 mg daily. This regimen is approved for multibacillary and paucibacillary leprosy for 6 and 12 months, respectively. WHO adjusted the dose for children <10 years old or <40 kg to Rifampicin 10 mg/kg monthly, clofazimine 6 mg/kg monthly, and then Clofazimine 1 mg/kg daily with Dapsone 2 mg/kg daily for the same period as adults.¹¹

Multidrug treatment for leprosy is effective and safe for both the mother and the baby during pregnancy and lactation. Although Rifampicin could cross the placenta and may be excreted in small amounts (5% of the therapeutic dose) in breast milk, no adverse effects on the fetus or baby has been reported till now.¹² Approximately 15% of the maternal weight-related Dapsone dosage may transfer to breast milk. WHO categorized Dapsone as "not safe" during breastfeeding; however, the American Academy of Pediatrics considers it as compatible with lactation. As for Clofazimine, it can cross the placenta, and about 22% of the maternal weight dosage enters breast milk; it may cause skin discoloration in the fetus or infant. Therefore, multidrug leprosy should be prescribed unchanged during pregnancy and lactation.¹²

Our patient presented with sudden onset of erythematous nodules -On the trunk during the third trimester of her pregnancy. The skin lesions and symptoms exacerbated after delivery, which may be due to T1R before initiation of therapy. Her 6-month-old baby was full-term with normal birth weight. The infant presented with only one hypopigmented skin lesion. Many acid-fast bacilli were found in his nasal smear; therefore, he received medication as a case of multibacillary leprosy.

CONFLICT OF INTEREST

None.


AUTHOR CONTRIBUTIONS

ZR: wrote the initial draft of the manuscript. PN: supported preparation and writing the manuscript. KKH, ND, JTF, and ZN: assisted in the preparation of the manuscript. MD: supported preparation and writing the manuscript. All authors: reviewed and approved the final manuscript.

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