Lucio's Leprosy: A Different Presentation of Hansen's Disease

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ABSTRACT

Lucio's leprosy is a distinct form of lepromatous leprosy characterised by non-nodular diffuse cutaneous involvement. Despite being endemic in underdeveloped countries, this particular form of leprosy is rarely reported outside the Mexico and Costa Rica. Owing to the absence of specific leprosy lesions and it is rarity, Lucio's leprosy causes great difficulty in diagnosis, which leads to leprosy reactions and functional impairment. We report a case of a 13-year female with diffuse skin involvement, loss of body hair, and ulcers. She was successfully treated with multidrug therapy (MDT). Timely diagnosis and prompt commencement of MDT can save patients from morbidity and mortality. This case is also noteworthy for the occurrence of Lucio's leprosy in childhood.

Key Words: *Lucio's leprosy, Childhood, Sclerodermoid skin.*

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INTRODUCTION

Leprosy is an ancient disease known to man with the highest prevalence in developing countries.[1] Mycobacterium leprae and lepromatosis are the culprit organisms causing this chronic granulomatous disease.[2] These gram-positive, acid-fast bacilli preferentially affect the skin and peripheral nerves.[3] Leprosy is classified into five types: Tuberculoid, borderline tuberculoid, mid borderline, borderline lepromatous, and lepromatous leprosy (LL) on the basis of immunity, clinical presentation, and histopathological findings.[2] Lucio's leprosy is a form of LL in which there is only a diffuse infiltration of skin without nodules.[4]

Herein, we report a case of a 13-year female with diffuse skin involvement, loss of body hair, and ulcers, who was successfully treated with multidrug therapy (MDT).

CASE REPORT

A 13-year female presented with loss of body hair for one year, which started from her legs and then involved her trunk, arms, and face with sparing of the scalp, pubic, and axillary hair. There was an ulcer formation on the right foot for one week, with puffiness of hands and feet for a few months, and blood-stained nasal discharge.

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On examination, there was thinning and loss of hair bilaterally over the eyebrows (Figure 1). Hand examination showed a shiny and taut appearance with mild skin thickening (Figure 2, 3). Similar findings were also appreciated on the legs along with non-pitting pedal oedema. There were two superficial ulcers present on the right foot (Figure 4). The erythematous rash was present in a livedoid pattern over the limbs. Examination of the face, including eyes, ear lobes, nose, and oral cavity, were all unremarkable. Sensations were impaired in gloves and stocking patterns. Ulnar, radial, and superficial peroneal nerves were bilaterally thickened.

Considering the skin findings, our differentials were Lucio's leprosy, scleroderma, myxoedema, and cutaneous vasculitis.

Routine investigations, including complete blood count, renal, liver function, coagulation profile, hepatitis B and C serology, thyroid profile, and anti-nuclear antibody (ANA), were negative. Skin biopsy showed granulomas in the mid-deeper dermis surrounding the blood vessels and adnexal structures. The Wade-fite stain was non-contributory. Slit skin smears were taken from the forehead, both ear lobes, chin and elbows, and the bacillary index was 1.5.

She was managed as a case of Lucio's leprosy. MDT was started in childhood dose. Rifampicin 450 mg and Clofazimine 150 mg orally were given monthly along with a daily dose of Dapsone 50 mg and Clofazimine 50 mg on alternate days. The patient showed remarkable improvement after a few weeks of therapy. Contact screening was negative.

DISCUSSION

The first case of Lucio's leprosy was reported in 1844. Later on, Lucio and Alvarado submitted their work in 1852.[5] Once considered as a disease of the Mexico and Costa Rica, Lucio's leprosy has now also been reported in India, Malaysia, Brazil, and Indonesia.[5-7]

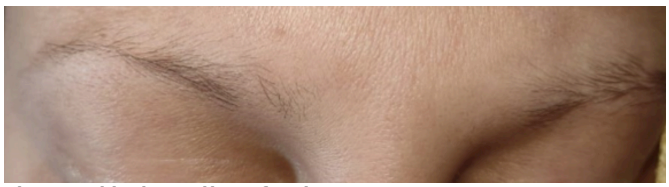

Figure 1: Thinning and loss of eyebrows.

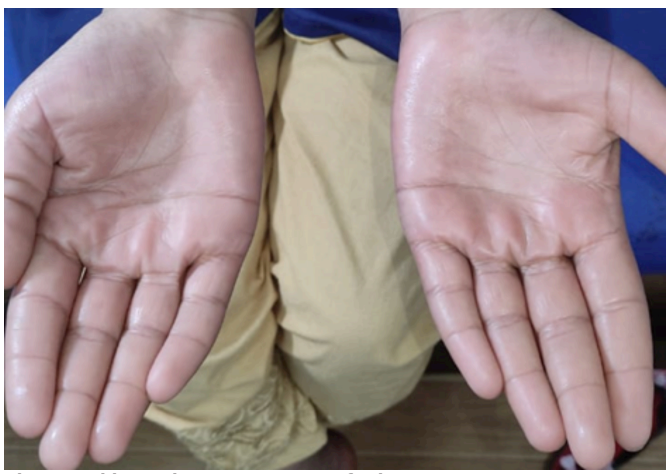

Figure 2: Shiny and taut appearance of palms.

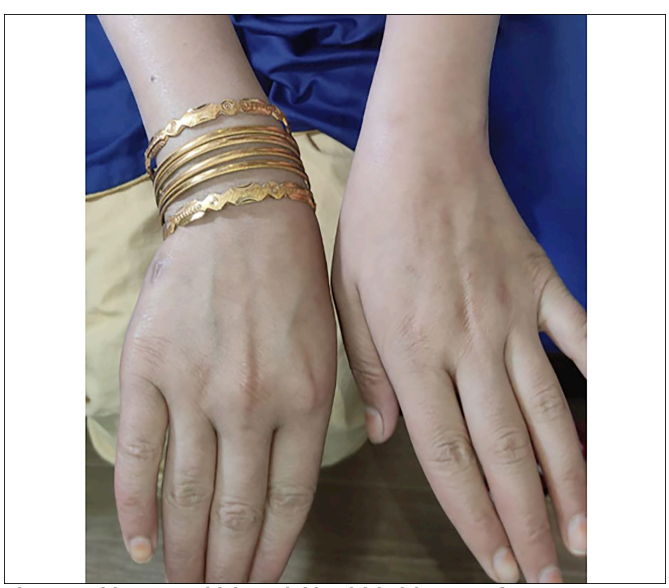

Figure 3: Shiny, taut, thickened skin with hairloss over forearms.

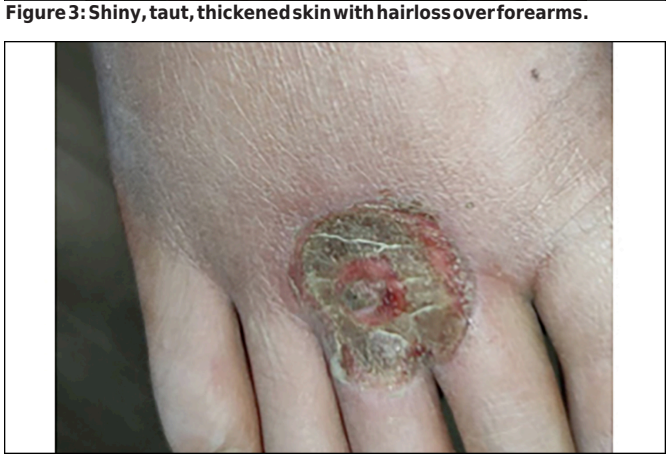

Figure 4: Ulcer on the right foot.

Lucio's leprosy is a rare type of LL, which presents as myxoedematous skin and loss of body hair. Due to the shiny appearance of the skin, this disease is also known as Lepra Bonita or pretty leprosy.[4] The noticeable clinical features are sclerodermoid skin, peripheral neuropathy, loss of eyelashes, loss of body hair, and rhinitis.[7] These characteristic findings mentioned in the literature are consistent with the clinical presentation of this patient. Other findings may include madarosis and thickened earlobes. Scalp hair are rarely involved due to the higher body temperature at this site.[6] In the present patient, there was sparing of scalp, axillary, and pubic hair. If left untreated, Lucio's leprosy can undergo a distinctive type of reaction called Lucio's phenomenon presenting as necrotising skin ulcers and stellate scars. The term "Lucio phenomenon" should be reserved for the patient who meets three criteria according to the international literature: Skin ulceration, vascular thrombosis, and invasion of blood vessels by bacilli on histopathology.[6,8] Systemic features of Lucio's phenomenon may include hepatosplenomegaly, fever, arthritis, and nephritis. Death can be due to disseminated intravascular coagulation (DIC) or sepsis.[5]

Owing to the absence of nodules and other cutaneous features of leprosy, Lucio's leprosy often goes unnoticed and imposes a great diagnostic challenge. It can easily be confused with scleroderma, myxoedema or cutaneous vasculitis.[7]

Diagnosis is based on a slit skin smear and skin biopsy. Lepromin skin test is usually negative. On histopathology, there is a diffuse infiltration of macrophages filled with acid-fast bacilli.[5] Wade-fite stain shows the presence of acid-fast bacilli. In this patient, the skin biopsy showed granulomas in the mid-deeper dermis surrounding the blood vessels and adnexal structures, and the Wade-Fite stain was negative. Studies suggest that clinical findings may not always align with histopathological findings, and the Wade-Fite stain can be non-contributory in certain circumstances. A definitive diagnosis requires the integration of both clinical and histopathological findings.[9]

The recommended treatment for Lucio's leprosy is MDT, which comprises Rifampicin, Clofazimine, and Dapsone for one year.[4] This patient received a standard paediatric dose of MDT and showed improvement. Studies are going on for newer combination regimens including monthly RMM (rifampicin, minocycline, and moxifloxacin)[10] and ROM therapy (Rifampicin, ofloxacin, minocycline)[11] to minimise the risk of side effects and improve compliance.

In conclusion, Lucio's leprosy is an extremely rare disease. To the best of our knowledge, this is the first case reported from Pakistan. The index of suspicion should be kept high as it closely resembles other skin conditions. This case is also notable for the occurrence of Lucio's leprosy in childhood. Healthcare workers and researchers should enhance their efforts to recognise and treat leprosy cases, provide vaccination programmes and educate the community to eradicate leprosy worldwide.

PATIENT'S CONSENT:
Informed consent was taken from the patient.

COMPETING INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
MS: Conception of work, data collection, literature review, and drafting of the manuscript.
AZ: Analysis of data, writing of the manuscript, literature research, and critical revision.
SAA: Conception of work, and revising the manuscript critically for the important intellectual content.
EA: Interpretation of data and critical revision.
SN: Data collection and critical revision.
All authors approved the final version of the manuscript to be published and agreed to be accountable for all aspects of the work.

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