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Case Report

Hansen's Disease Presenting as Erythroderma in a Tribal Woman of South Gujarat

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The term erythroderma is defined as generalized erythema with scaling, affecting 90% of the skin surface and is considered as a dermatologic emergency. It can precipitate secondary to skin diseases such as psoriasis, atopic or seborrheic eczema, drug-induced, congenital conditions, due to T-cell lymphoma of the skin or idiopathic and rarely infectious causes. We report a case of a 40-year-old female, who presented to us with complaints of generalized erythema and scaling involving the trunk, face, and extremities along with tightness of skin and generalized swelling. There was also the presence of oral ulceration, and low-grade fever, along with generalized weakness and lethargy which was aggravated over the last few days. Loss of lateral one-third of eyebrows was also noted suggesting atopic cause. With a differential diagnosis of erythroderma due to atopic eczema or other infection, a biopsy was done. Histopathological examination was suggestive of borderline lepromatous Leprosy with type 1 lepra reaction, Fite Faraco stain was also positive showing solid staining bacilli. To confirm neurological examination was done, and it was observed that she had bilaterally symmetrical nerve thickening of all the major peripheral nerves. The slit skin smear from the ear lobe stained positive for *M. leprae*. Further, on clinicopathological correlation, the diagnosis of borderline lepromatous leprosy with type I reaction (Reversal reaction) was confirmed. In rare sporadic cases, an infectious disease can also precipitate into erythroderma. Thus, in India, especially in endemic zones a high index of suspicion along with proper clinical history, examination, and routine tests like slit-skin smears and biopsy are essential to reach the precise diagnosis of an underlying disorder and therefore crucial to institute the appropriate therapy. Awareness of leprosy with reactions or undergoing therapy with drugs like dapsone as a cause of erythroderma is necessary. The case presented in this report shows simple tests like slit-skin smears for acid-fast bacilli (AFB) alone or in conjunction with histopathology will help establish the diagnosis and also classify the spectrum of the disease.

Keywords: Erythroderma, Hansen's Disease, Type 1 Lepra Reaction.

Introduction

Leprosy is a slowly progressing, mildly contagious disease caused by *Mycobacterium leprae* and *M. lepormatosis*, primarily affecting the skin, peripheral nerves, and various organs (Global Leprosy Update 2019). Clinical manifestations ranging from macules, papules, plaques, and nodules affecting peripheral body surface area, along with edema of lower extremities, and hypoesthesia of peripheral extremities (White

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& Franco-Paredes 2015). Ichthyosis of limbs and various deformities of the face, hands, and feet can develop in advanced stages of patients with lepromatous pole (Alencar et al 2012).

Leprosy is still endemic in several countries, including India and there are locations in India still having high endemicity. Government data from districts like Tapi in Gujarat with more than 80% tribal population show that incidence of leprosy had gone up from 9.37 per 10,000 population in 2010 to 17.16 per 10,000 population in 2014 (Sharma 2019). More recently, the Tapi district of Gujarat has achieved the elimination goal in March 2020 (National Health Mission, Govt of Gujarat, https://nhm.gujarat.gov.in/nlep1.html). Awareness of leprosy, especially about its atypical forms assumes greater importance in low endemic situation.

Lepra reactions can cause severe skin alterations, neural disability, and, consequently, social and functional stigmas. Skin lesions are polymorphic, ranging from a single hypochromic hypo aesthetic macule to diffuse skin infiltration (Pandhi & Chhabra 2013). This polymorphism in the presentation can be a cause for delayed diagnosis, leading to the progression of disabilities and increased risk of transmission.

Erythroderma is an inflammatory skin disorder presenting as generalized erythema & scaling (Sehgal et al 2004) affecting more than 90% of the body surface area and is considered a dermatologic emergency. Several diseases can precipitate or can present as an exacerbation of preexisting dermatoses (i.e., psoriasis, atopic dermatitis, endogenous eczema), drug reaction, cutaneous T-cell lymphoma, rarely caused by infections (i.e., scabies, dermatophytosis) (Ingram 2106). Baldissera et al (2019) also reported a case of borderline lepromatous leprosy with erythroderma. Here we came across a case that displayed erythroderma possibly secondary to infectious cause (leprosy) which though rare but does occur (Patki & Mehta 1989).

Case Report

A 40-year-old female patient belonging to the local tribe from Tapi district, Gujarat presented to us with chief complaints of generalized erythema,

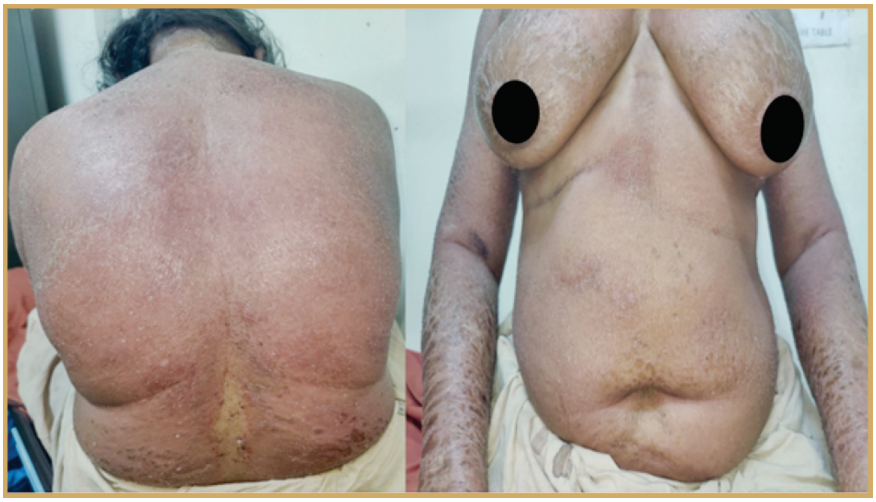

Fig. 1 : Erythema with scaling and induration of trunk. There is the presence of notable ichthyosis over the forearm in the right-side image.

scaling present on the face, trunk, and extremities (Fig. 1) for 1 year. She also complained of feeling tightening of skin and swelling over her hands, feet, and face for 1 week, along with low-grade fever, weakness, and oral ulceration for the last 4 to 5 days. The total duration of appearance of symptoms which were relatively non-debilitating was 1 year. Initially, the patient developed asymptomatic erythematous skin lesions on the face for which she visited a general practitioner and was prescribed certain oral and topical drugs for which no documents were obtained. No other significant history was obtained.

On examination, diffuse erythema with scaling and diffuse plaques were present over the face, trunk, and both upper and lower limbs. There was the presence of anasarca (Figs. 2 and 3). Bilateral supraciliary madarosis was also noted (Fig. 4). The oral examination revealed a few erosions on the soft palate. There was generalized ichthyosis with sparing of inframammary folds, popliteal, and cubital fossa. Examination of the lymph

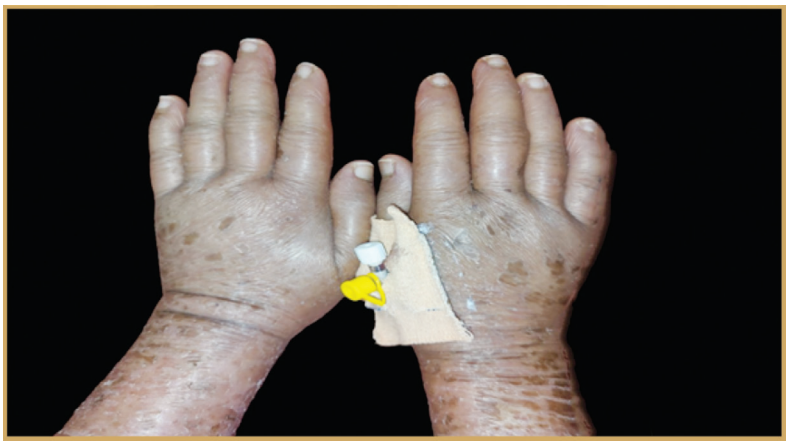

Fig. 2 : Oedema of both hands with ichthyosis.

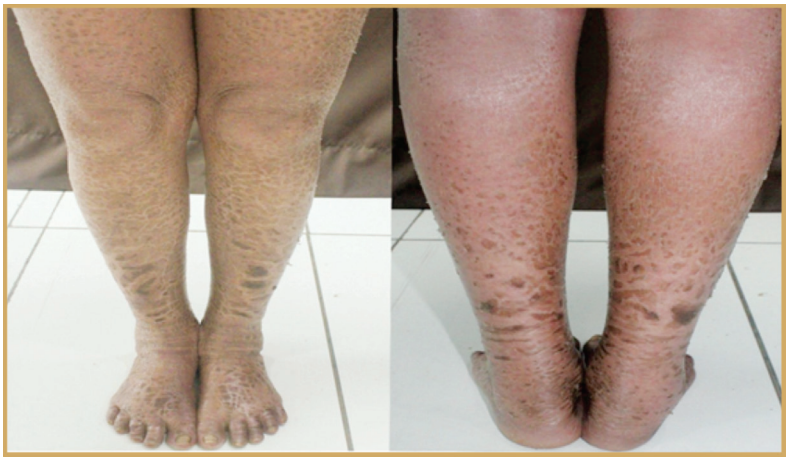

Fig. 3 : Erythema, edema, and ichthyosis of lower limbs.

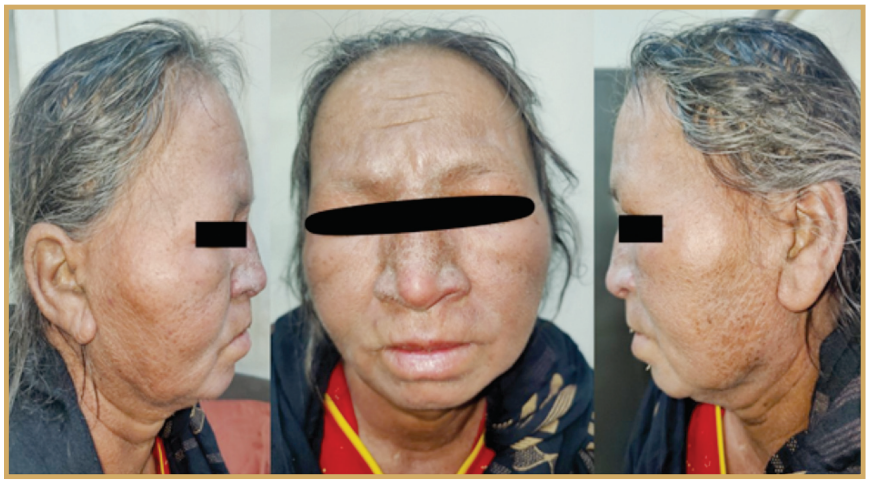

Fig. 4: Facial edema, erythema, and bilateral ear infiltration with madarosis over face and ears.

nodes showed 3 enlarged palpable tender nodes which were elliptical, and firm in consistency, with normal overlying skin surface present in both inguinal regions. A complete blood count revealed hemoglobin of 11.1g/dl, a total count of 13000/cmm, a platelet counts of 160000/cmm, lymphocyte count of 23% lymphocytes, and an erythrocyte sedimentation rate of 26 mm per hr. Liver function test, and renal function test, were within normal limits, and urine routine microscopy examination was within normal limits. The patient was seronegative for HIV and HBsAg. CRP was non-reactive. Thus, clinically with a differential diagnosis of erythroderma due to psoriasis, drugs or atopic eczema were made. Skin biopsy was done from two sites; 1) plaque on the lower posterior trunk and 2) the extensor aspect of the right forearm. Skin biopsy specimen showed effacement of the rete ridges and prominent papillary dermal edema with the presence of multiple large, ill-demarcated elongated peri-neural and peri-appendageal diffuse macrophage granulomas (Figs. 5a, 5b) with foamy histiocytes, abundant lymphocytes, and plasma cells which were predominantly located in the papillary and mid dermis. The deep dermal macrophage granulomas in the peri neural region displayed an onion peel appearance with the presence of a few globi and lymphocytosis (Patnaik et al 2014). This suggested the case as borderline lepromatous leprosy with (Reversal reaction) type 1 reaction. The histopathologist asked for detailed clinical history regarding pruritus and anesthetic patches, sensory examination of extremities, drug history, and clinical images and advised a slit skin smear till he obtained Fite-Faraco stained slides.

Again, a detailed retrospective history was taken when she revealed that a few months ago she noticed a few patches on her body, which gradually progressed to her trunk and extremities and were associated with episodes of high-grade fever with chills followed by the development of generalized erythematous tender nodules for which she visited some private dermatologist and was prescribed oral steroids, multivitamins, and capsule clofazimine (100mg) once daily nearly 8 months ago without making a proper diagnosis, however, the diagnosis was not mentioned in the prescription. The patient had a couple of such

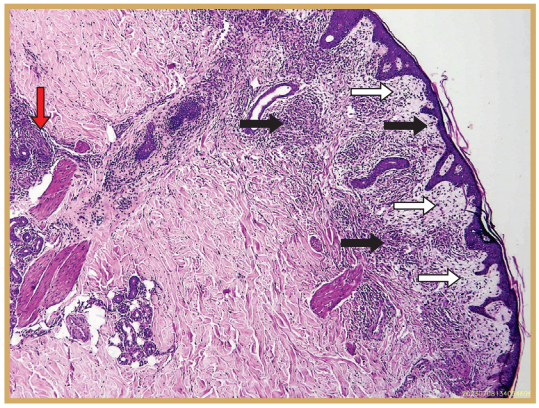

Fig. 5a : Histopathology using H and E stain at 4X displays focal effacement of the rete ridges (blue arrow), and extensive papillary dermal edema (white arrows) with the presence of multiple diffuse large (black arrows), elongated peri-appendageal diffuse macrophage granulomas (Orange arrow) in the mid dermis.

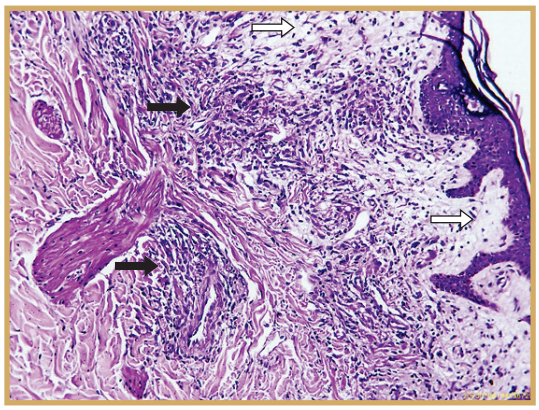

Fig. 5b : Histopathology using H and E stain at 10X displays irregular rete ridges pattern with focal effacement of the rete ridges, and prominent papillary dermal edema (white arrows) with the presence of multiple diffuse macrophage granulomas (Black arrows) in the upper dermis.

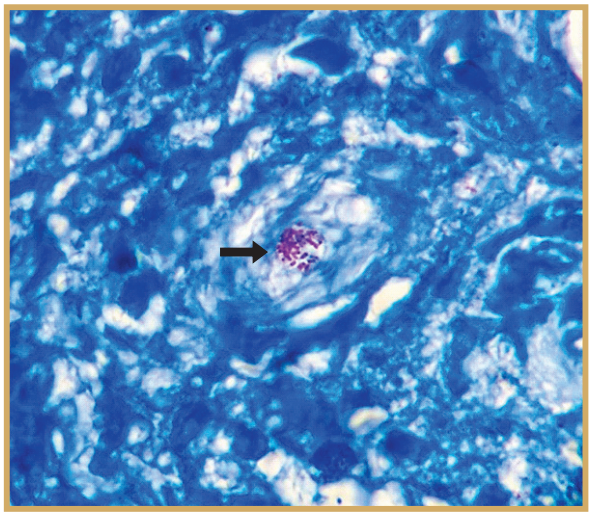

Fig. 5c : Fite Faraco staining showing the presence of acid-fast bacilli (Black arrow) in the tissue.

episodes in the last few months. Thus, a suspicion of dapsone syndrome in a case of leprosy was made and further the patient was probed, but a detailed history and older prescriptions did not reveal any history of taking WHO MDT, rifampicin, or dapsone separately.

On performing examination ear infiltration was observed and on sensory examination, we found

loss of temperature sensation and altered touch sensation over the extremities distal to the elbow and knees. Neurological examination revealed thickened, enlarged, tender bilateral ulnar nerves and common peroneal nerves with grade 1 neuritis. Bilateral posterior tibial, superficial radial cutaneous, and great auricular nerves were prominently thickened, and non-tender. Slit skin smear from the ear lobe was positive for *M. leprae* with a bacillary index of 3+, and from the skin lesion (left side of lower posterior trunk) was 1+. The Fite-Faraco stain was positive for *M. leprae* with a bacillary index of 3+ (Fig. 5c) and a morphological index of greater than 90% solid bacilli.

Thus, on clinicopathological correlation, the diagnosis of type I reaction (Reversal) in a case of borderline lepromatous leprosy was made. After confirmation of diagnosis, the patient was administered tab. prednisolone (40mg) in tapering doses, tab. paracetamol (500mg) BD, WHO Adult MB MDT Pack. After 15 days of initiation of prednisolone, complaints of fever gradually subsided and the erythema, edema, and scaling, reduced over extremities, and the patient continues to take MB MDT with regular follow-up at the hospital.

Discussion

It was rightly quoted by Baldissera et al. that some cases of erythroderma always remain idiopathic until appropriately investigated (Baldissera et al 2019). The specific aspects or diagnostic features of the original disease are typically masked by the characteristic signs of erythroderma, thus a detailed and precise clinical history, appropriate examination, especially past history, treatment history, and appropriate investigations are necessary. It is the responsibility of the clinician to probe in detail the points which a patient sometimes fails to reveal but are essential to a clinician in making a suitable clinical diagnosis and ordering laboratory tests.

Erythroderma due to dapsone syndrome was another obvious differential diagnosis considering the symptoms but as observed in history the patient was prescribed clofazimine but not directed to start the WHO multi-bacillary multi-drug therapy nor prescribed treatment as per the guidelines. Thus, the patient had not consumed dapsone before presentation thus ruling out dapsone syndrome. Dapsone syndrome is usually characterized by high-grade fever, maculopapular rashes, hepatitis, lymphadenopathy, and lymphocytosis, and it generally appears within the first 6 weeks of treatment. From the private clinician's perspective, it is a missed opportunity to diagnose leprosy and report the authorities to provide appropriate to the patient which is available free of cost in the form of WHO MD-MDT and under proper supervision by the health care workers thus assuring adherence to therapy.

The precise immunological mechanisms underlying erythroderma and type 1 lepra reaction are still unclear, especially in this case where the person has been suffering from Hansen's for nearly one year and has taken clofazimine yet developed a reversal reaction. It is well documented that type 1 lepra reaction would constitute episodes of aggravated Th-1 responses, which are precipitated by the release of antigens from killed *M. leprae* either by mycobactericidal drugs or by spontaneous flare-ups of the anti-mycobacterial immunoreactivity in patients with unstable immunity in the borderline group (Walker & Lockwood 2008). This case report brings some unusual immunological findings where features of type I Lepra reaction are observed to present over two to three months in the form of erythema, scaling with anasarca that probably reflect the complex interplay between reversal reactions in leprosy and the development of erythroderma post clofazimine. (Fonseca et al 2017, Miyashiro et al 2017).

Our patient presented with generalized erythema and scaling along with ear infiltration, ichthyosis,

supraciliary madarosis, edema of hands, and feet, low-grade fever, tenderness of nerves. A positive slit skin smear for acid-fast solid staining bacilli and a biopsy displaying papillary dermal oedema, diffuse macrophage granulomas with oedema, lymphocytosis, and foamy macrophages and both being positive for *M. leprae* on Fite- Faraco stain, led to diagnosis of borderline lepromatous leprosy with type I reaction. Skin lesions were masked by the generalized swelling, erythema and scaling.

In routine field settings, the clinical diagnosis of leprosy is the usual approach rather than histopathological confirmation which is not normally available and also not required for common presentations. However, for a rare case like presented in this study this misled the clinician(s) into considering it as a case of erythroderma due to other causes and thus even during biopsy leprosy was not even a differential diagnosis suggested to the histopathologist. The situation would have been different if there had been awareness about this possibility and slit-skin smears were done earlier.

In type 1 lepra reaction, history is short and there is a sudden change in skin lesions alone, nerve lesions alone, or both. Just edema in the dermis will not go in favor of type 1 lepra reaction. In ENL or type 2 lepra reaction, erythroderma is not known to occur. It needs to be emphasized that in this case the things that favor Type I reaction are histopathological findings along with clues in the clinical history, i.e. The patient complained of oedema over hands, feet and face for 1 week, along with low grade fever and weakness for 4-5 days. That goes adequately with the signs of Type I reaction. In earlier reports erythroderma has also been associated with borderline leprosy (Miyashiro et al 2017, Baldissera et al 2019) and also borderline cases with reversal reaction (Miyashiro et al 2017). These reports thus corroborate our findings. Moreover, our case responded well to MB-MDT with oral steroids and thus it also confirms our interpretation.

India is a nation that contributed nearly 62% of the global Leprosy cases to the world and a 37.7% increase in new cases detection in 2022 (WHO 2023), we always need to remember, that leprosy is the great imitator (Kundacki & Erdem 2019). Unusual presentations like BL leprosy with type 1 reaction presenting with erythroderma in our case though rare has been reported from India (Patki & Mehta 1989), Brazil (Miyashiro et al 2017) and Mexico (Romero et al 2020). Like our case, Romero et al (2020) didn't consider leprosy in the initial diagnosis, thought of psoriasis and lymphoproliferative disorders when asking for biopsy which on histopathology was confirmed as lepromatous leprosy and the patient responded well to multi-drug treatment. Patki & Mehta (1989) identified dapsone hypersensitivity as the cause for erythroderma. Miyashiro et al (2017) established erythroderma in their case due to borderline leprosy and reversal reactions which was the diagnosis in our case as well.

In conclusion, this case report highlights the importance of a thorough history taking and examination of erythrodermic patients, especially in a leprosy-endemic nation like ours which sometimes needs a high index of suspicion and ask for routine tests like slit -skin smear for AFB. Biopsy will help classify the spectrum but would not be needed to diagnose leprosy and initiate appropriate treatment to manage such rare cases.

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