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Case Report

A Case of Borderline Lepromatous Leprosy in Type 1 Lepra Reaction Presenting as Impending Erythroderma

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Erythroderma is a dermatological emergency presenting with generalized scaling and erythema involving more than 90% of body surface area. Many pre-existing dermatoses, underlying systemic conditions, drugs even idiopathic conditions may present as erythroderma. Leprosy is known to have an unpredictable course and varied clinical presentations depending upon on the bacillary load and individual immunity. Erythroderma in leprosy as initial presentation is rare, only few such case reports exist in literature. We report a case of an elderly male who presented with rapidly progressing generalized erythema and scaling with impending erythroderma and was diagnosed to have borderline lepromatous disease with type1 cutaneous lepra reaction. This case highlights the importance of high index of suspicion of leprosy when encountering such case in routine clinical practice to prevent delay in the diagnosis, management and to avoid the likely complications.

Keywords : Erythroderma, Leprosy, Type 1 Lepra reaction

Introduction

Erythroderma or exfoliative dermatitis is an inflammatory skin disorder comprising of erythema and scaling covering more than 90 % body surface area. Pre-existing dermatoses such as psoriasis, atopic dermatitis, contact dermatitis and systemic skin conditions including malignancy and drug intake can all manifest with erythroderma (Manjunath et al 2014). Erythroderma can be fatal due to numerous metabolic and systemic complications including acute skin failure, which is of importance to the dermatologist (Okoduwa et al 2009). An early diagnosis of a patient presenting with erythroderma is very essential to initiate definitive therapy at the earliest. Leprosy is

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disease of peripheral nerves and skin, classically presents with hypoaesthetic, hypopigmented macules to widespread, numerous, erythematous numb or normoaesthetic patches depending on the pole of the disease. Borderline lepromatous cases have multiple lesions which are generally tending to symmetry. Type 1 lepra reaction generally occurs after starting MDT in borderline pole but patients may present in type 1 reaction. During type 1 lepra reaction, the pre-existing lesions get more erythematous, indurated and occasionally ulcerated (White & Fraco-Paredes 2015). So far there are only three case reports where erythroderma was the initial presentation in leprosy.

We report a case where an elderly man presented with rapidly progressive erythema and scaling where high index of suspicion, promptly conducted slit skin smear and histopathological examination clinched the diagnosis of borderline lepromatous leprosy.

Case Report

A 62 years old male, a resident of Haryana (India) with no known co morbidities presented with a 3 weeks history of insidious onset gradually progressive red raised scaly lesions, initially over face and ears and gradually progressed to involve his entire trunk and extremities. The lesions were not associated with any itching, burning sensation, pain or loss of skin sensations. The individual did apply hair dye about a week prior to his symptoms. There was no history suggestive of type 2 lepra reaction, signs / characteristics of lepromatous pole, neuritis, muscle weakness and any feature suggestive of acute or chronic skin disease or skin failure. There was no history of significant weight loss, malaise, fever, photosensitivity, joint pain, oral ulceration or intake of drugs or even ayurvedic/alternative medicines. General and systemic examinations were essentially normal. Dermatological examination revealed numerous, polycyclic, erythematous, indurated, coalescing, normoaestheticscaly plaques involving his face, earlobes, trunk and extremities covering about most of body surface area (Fig. 1a, b, c, d). The lesions on the trunk and extremities were tending to symmetry. There was sparing of the hair bearing areas of the body i.e scalp, axilla and groin, mucosa, genitalia, palms and sole.

A differential diagnosis of disseminated eczema, cutaneous T cell lymphoma, parapsoriasis, drug rash, sarcoidosis, sub-acute cutaneous lupus erythematosus and leprosy was considered. Leprosy was considered due to two elevated 'inverted saucer' shaped erythematous plaque over chest (Fig. 1c) and ear lobe infiltration, however it was the most improbable differential diagnosis considering the three weeks onset of symptoms and absence of cardinal features of leprosy, i.e. no loss of sensation or thickened peripheral nerves. His slit skin smear was positive for Acid Fast Bacilli (3+) and his histopathological examination of the skin biopsy showed mononuclear inflammatory infiltrate containing lymphocytes and numerous histiocytes, with vascular, adnexal and neural involvement along with mild dermal edema and dermal lymphocytic pkynosis opined as borderline lepromatous leprosy (Hematoxylin & eosin, x40X and x 100). (Fig. 2).

Considering the complications related to impending erythroderma, patient was admitted. He was given supportive care in the form of maintaining ambient room temperature, adequate nutrition, barrier nursing. He was started on Multi drug therapy multibacillary regimen (MDT MB) for leprosy and on oral prednisolone (1mg/kg/day) for Type 1 Lepra reactions as he had erythematous plaques and multiple indurated plaques on his face. In the next 2weeks, patient started responding to treatment

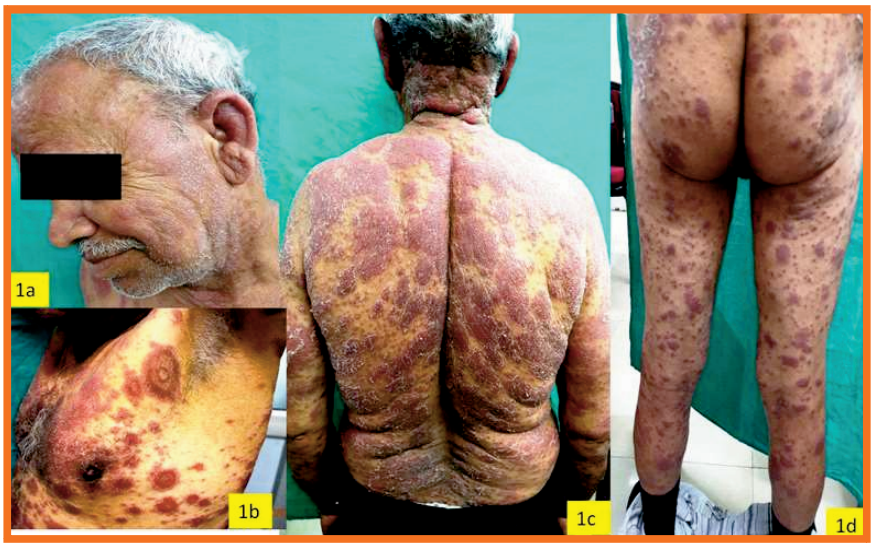

Fig. 1a : Erythematous indurated scaly plaques over face with ear lobe infiltration,
Fig. 1b : Inverted saucer shaped lesions on the left upper chest,
Fig. 1c : Generalized involvement of erythematous scaly polycyclic coalescing plaques over trunk &
Fig. 1d : Generalized involvement of erythematous scaly polycyclic coalescing plaques over back

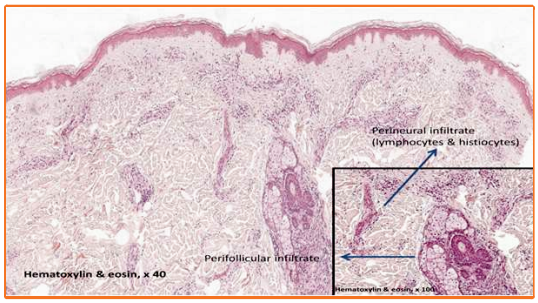

Fig. 2 : Mononuclear inflammatory infiltrate containing lymphocytes and numerous histiocytes, with vascular, adnexal and neural involvement (Hematoxylin & eosin, x40, x 100 inset)

with decrease in erythema, induration and scaling over the plaques. Presently the patient is on MDT MB and on tapering doses of oral prednisolone and has shown a good response. Erythema and scaling has totally regressed leaving behind post inflammatory hyperpigmented macules (Fig. 3).

Discussion

Leprosy is one of the oldest diseases known to mankind. Classically it has distinct spectrum of clinical, histopathological features based upon immunity of affected individual. Leprosy causing erythroderma is rare and the immunopathogenesis is still debatable. Lepra reactions are known to occur in 30-50% of individual during the course of the disease with type 1 lepra reaction representing an exacerbated Th1 response (Fonseca et al 2017). Immune dysregulation is seen in erythroderma with reports that benign forms of erythroderma presents with slight predominance of IFN-γ over IL-4 expression, while in malignant forms IL-4 is predominant (Sigurdsson et al 2000). Although

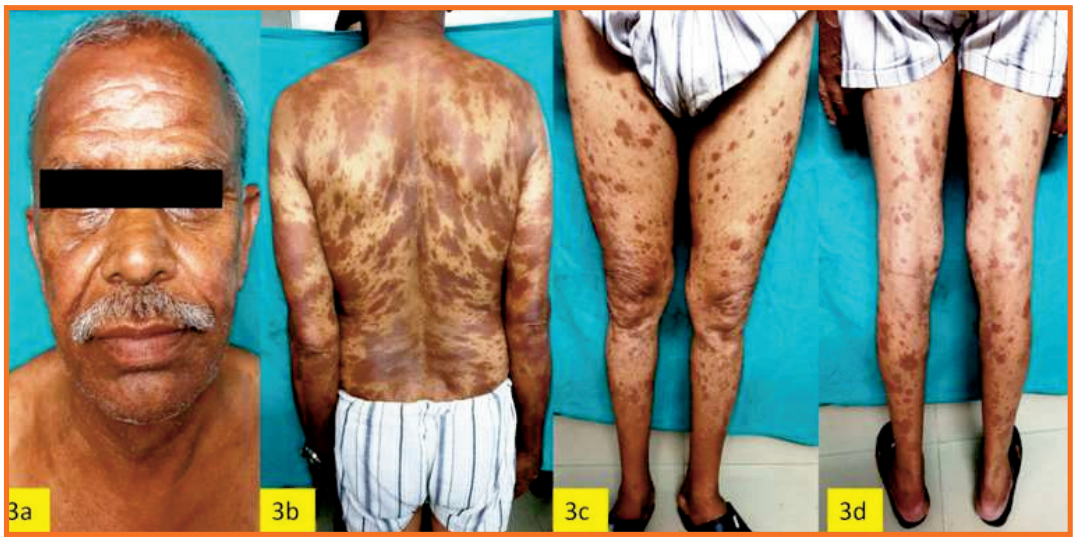

Fig. 3: Resolved skin lesions over face (a), chest (b), legs (c & d).

very little data is available, but some correlation in the immunopathogenesis can be made in patients of borderline leprosy who have type1 reaction and presents with erythroderma. The diagnosis of leprosy is primarily based on clinical histopathological and slit skin smear reports but a high index of suspicion is mandatory in endemic regions which can have varied presentations. There are case reports of generalized body involvement finally being diagnosed as Leprosy. Baldissera et al (2019) have reported a 71 years old male patient with a 1-year old history of erythroderma finally diagnosed as a case of leprosy and Shenoy et al (2015) had reported of a 55 years old male patient with a positive deck chair sign. Prashar et al (2013) had first reported a case of lepromatous leprosy with similar features and a positive deck chair sign. Sparing of the abdominal creases and axilla, a sign described as "deck-chair sign". The sign has been classically described in papuloerythroderma of Ofuji, but may not be a specific sign since it is described in certain other conditions such as generalized acanthosis nigricans, Waldenstrom's macroglobulinemia, large plaque parapsoriasis, angioimmunoblastic T-cell lymphoma, and erythroderma due to various causes.

Our case was different as leprosy induced erythroderma in our patient was of acute onset and there was a paucity of cardinal signs of leprosy in our case. Thus, an early diagnosis could only be made by demonstratingacid fast bacilli in slit skin smear and a histopathology which was consistent with borderline lepromatous disease.

A delay in diagnosis in cases of leprosy and lepra reactions can lead to an irreversible neural damage and deformities. Further erythroderma in such patients can lead to increase morbidity and mortality due to its complications. Also, if such cases are left untreated for long may become potential reservoir of infection in the society so defeating whole purpose of leprosy control programme in the country. Hence, the importance of this case is to highlight the requirement of high index of suspicion of leprosy in cases of even erythroderma as well. This case report thus aims at making the readers aware

about such atypical presentations of leprosy which would become more important in future low endemic scenario.

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