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Case Report

Two atypical presentations of lepra reactions: Case report

Tony Kuncheria[1],*, V Siva Subramanian[1]

[1] *Dept. of Dermatology, Vinayaka Missions Medical College and Hospitals, Karaikal, Puducherry, India*

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ABSTRACT

Hansen's disease is a chronic infectious granulomatous disease with varied presentation, especially in the setting of lepra reactions. So here we report two such atypical presentations each of Type 1 and Type 2 Lepra reactions. The first was of an middle aged male presenting with painful raised lesion over both thighs with high grade fever with no multidrug therapy taken and no skin lesions and was diagnosed as type 2 lepra reaction while the second was that of a elderly male presented with raised margin over the pre-existing lesions with neuritic features with no multidrug therapy taken as was diagnosed as type 1 lepra reaction.

1. Introduction

Leprosy is a chronic disease, which remains active for a long time. During the long course of activity of the disease, in several patients, there are acute bouts of exacerbation characterized by sudden increase in the activity of the disease. Reactions can be defined as sudden tissue responses, resulting from the liberation of the bacilli or their products, into the tissues. We report two such cases with atypical presentation of lepra reactions. The first was of an middle aged male presenting with painful raised lesion over both thighs with high grade fever with no multidrug therapy taken and no skin lesions and was diagnosed as type 2 lepra reaction while the second was that of an elderly male presented with raised margin over the pre-existing lesions with neuritic features with no multidrug therapy taken as was diagnosed as type 1 lepra reaction.

2. Case Report 1

The first was of an middle aged male presenting with painful raised lesion over both thighs since 4 days(Figure 1) associated with Fever since 4 days. Patient was apparently normal 4 days back then he developed painful red coloured raised lesion over both thighs(Figure 2) which was insidious in onset, gradually progressive associated with fever which was more at night. There was no history of cough, cold, stuffiness of nose, bleeding from nose , redness of eye, joint pain, deviation of angle of mouth , hoarseness of voice, no history of loss of lateral one third of eyebrows. History of drug intake prior to the onset of lesions was not present. There was no history of taking Multidrug therapy in the past. No associated comorbidities were present.

2.1. Clinical Examination

General physical and systemic examinations were normal.

Cutaneous examination revealed presence of crops of tender , erythematous nodules over anterior aspect of bilateral thighs.(Figure 2). There was no hypopigmented or hyperpigmented large macule in the body. Oral cavity was normal. Ocular examination was normal with no features of iritis and iridocyclitis. No abnormality was detected on genital examination. Ulnar nerve was thickened and tender bilaterally. Card test was positive bilaterally. Systemic examination was normal. The diagnosis of leprosy in Type 2 lepra reaction was made on history and clinical examination. Patient was explained about the diagnosis

* *Corresponding author*.
E-mail address: tonykakka@gmail.com (T. Kuncheria).

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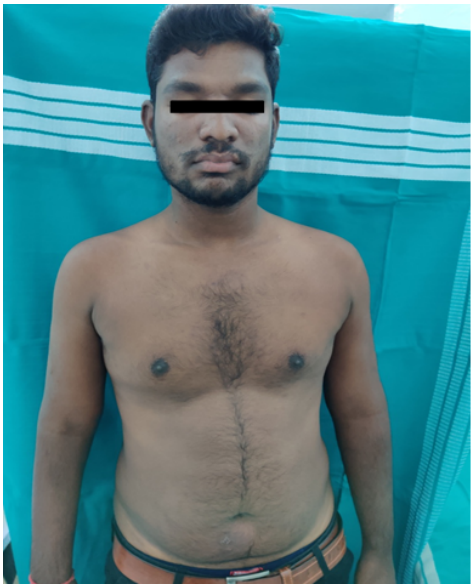

Fig. 1:

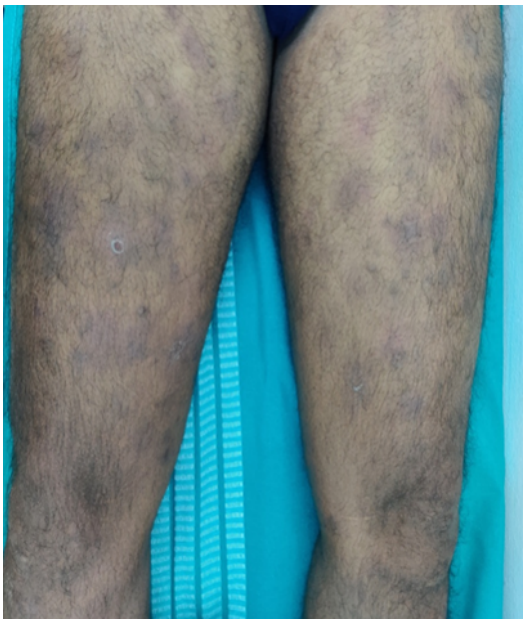

Fig. 2: Multiple red colour raised lesions over both thighs

along with treatment approach and informed consent was taken for further investigations and treatment protocol.

2.2. *Investigations*

WBC count and ESR was elevated. Slit-skin smear was negative. 6mm punch biopsy was taken from the left thigh area over the nodule under all aseptic precautions . Histological examination showed dermis and subcutaneous tissue shows multiple nodules composed of sheets of macrophages.(Figures 3 and 4). Many capillaries show perivascular lymphocytic infiltration and polymorphs.(Figure 5).

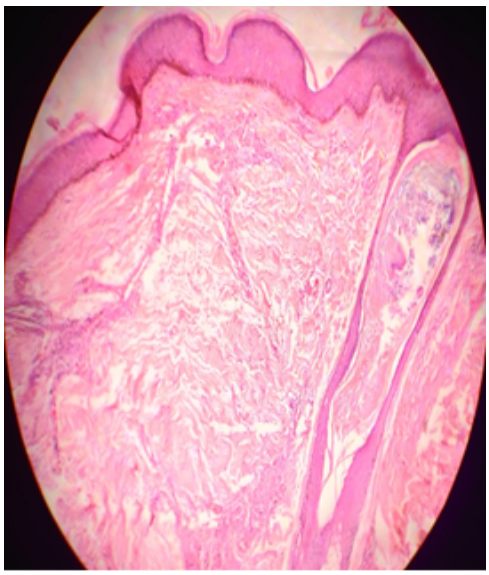

Fig. 3: Photomicrograph showing dermis and subcutaneous tissue shows multiple nodules composed of sheets of macrophages.(H&E stain,x10)

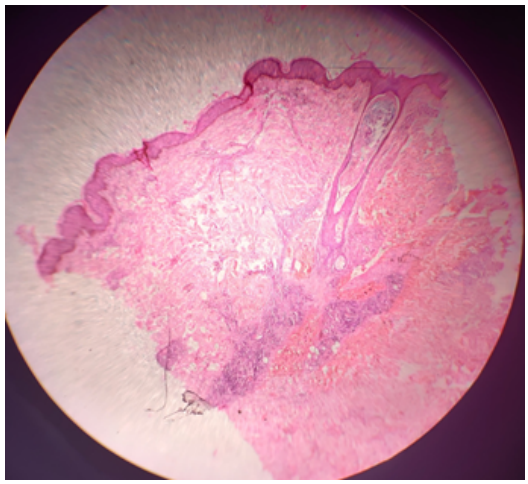

Fig. 4: Photomicrograph showing dermis and subcutaneous tissue shows multiple nodules composed of sheets of macrophages.(H&E stain,x10)

Based on clinical features, morphology of lesions and histopathology studies, a final diagnosis of Hansen's disease in Type 2 reaction was made. Hence the treatment with oral prednisolone (40mg/day) along with Multivitamins was given. Lesions started resolving without the emergence of new lesion. Therefore, the dose of prednisolone was tapered gradually (30mg/day) and then to (20mg/day). Then we contacted DD leprosy centre and he was referred to kottucherry government hospital and was started on

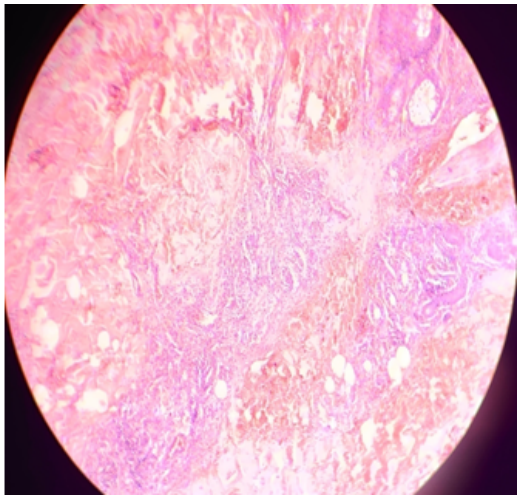

Fig. 5: Photomicrograph showing many capillaries show perivascular lymphocytic infiltration and polymorphs.(H&E stain,x40)

antileprosy treatment (multidrugtherapy).

3. Case Report 2

The second was that of an elderly male presented with light coloured flat lesions present over bilateral upper limb, front and back of trunk, bilateral thighs, bilateral buttocks and over left chest since 30 years and aggravated with raised margin since 1 month. Patient was apparently normal 30 years ago then he developed itchy flat light coloured lesion which started over right upper limb which was insidious in onset gradually progressive associated with itching. History of decreased sweating over the lesions was present, decreased hair over the lesion was present , decreased sensation over the large lesions and intact sensations on smaller lesions was present and history of numbness over affected area was present since 1 month. Loss of lateral one-third of eyebrows was present. No history of bleeding from nose, swelling over bilateral ankle and feet. There was no history of taking Multidrug therapy in the past. No associated comorbidities were present.

3.1. Clinical examination

General physical and systemic examination were normal.

Cutaneous examination revealed presence of multiple ,well defined, hypopigmented macules and large macules with raised margin and flattened centre, asymmetrical in distribution over face, bilateral upper limb, front and back of trunk, bilateral thigh.(Fig.6,7,8). Total Number of lesions were nine. Left supra orbital nerve was thickened, Bilateral greater auricular nerve was thickened , Bilateral ulnar nerve thickened and tender, Right posterior tibial nerve was thickened and tender. Card test was positive bilaterally. Oral cavity and ocular examination was normal. No abnormality

Fig. 6: Multiplehypopigmented large macules over chest.

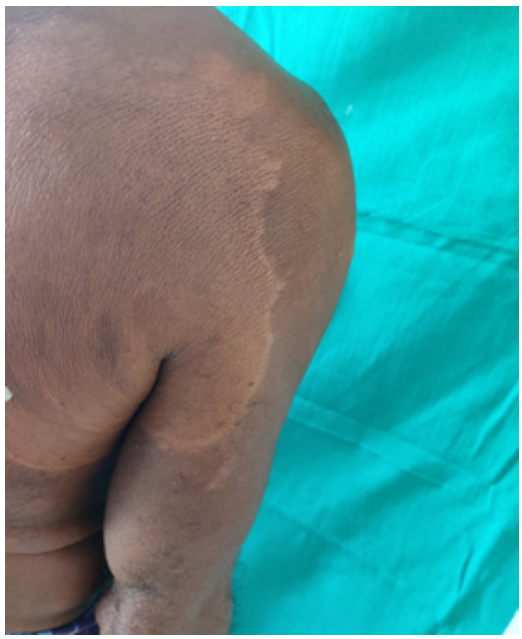

Fig. 7: A large macule with raised margin and flattened centre present on right scapular region.

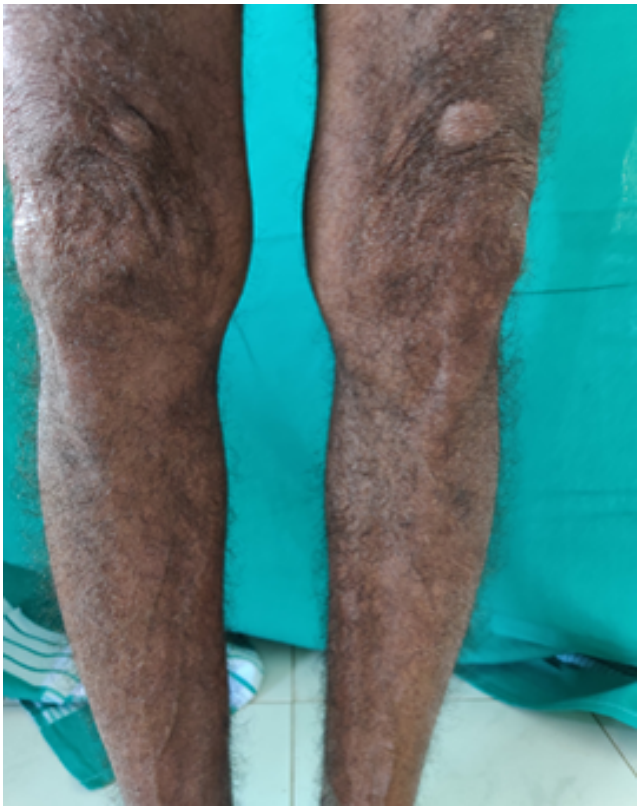

Fig. 8: Multiple large macules with raised margin present over bilateral thighs(anterior aspect).

was detected on genital examination. Systemic examination was normal. The diagnosis of leprosy in Type 1 lepra reaction was made on history and clinical examination. Patient was explained about the diagnosis along with treatment approach and informed consent was taken for further investigations and treatment protocol.

3.2. Investigations

Complete hemogram and other relevant biochemical tests, such as complete urine analysis, serum creatine , blood urea , uric acid , lipid profile, liver function tests were within physiological limits. Slit-skin smear was negative. 6mm punch biopsy was taken from the right scapular area over the large macule under all aseptic precaution. Histological examination showed epidermal atrophy(Figures 9 and 10) and Perineural lymphocytic infiltration present.(Figure 11).

Based on clinical features, morphology of lesions and histopathology studies, a final diagnosis of Borderline tuberculoid leprosy with Type 1 reaction was made. Hence the treatment with oral prednisolone (20mg/day) along with Multivitamins was given. Lesions started resolving without the emergence of new lesion. Therefore, the dose of prednisolone was tapered gradually (15mg/day) .Then we contacted DD leprosy centre and he was directed

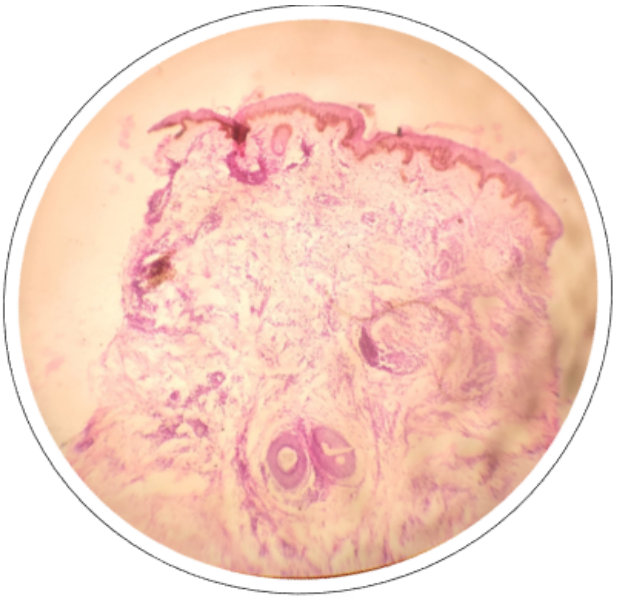

Fig. 9: Photomicrograph showing epidermal atrophy(H&E stain, x10).

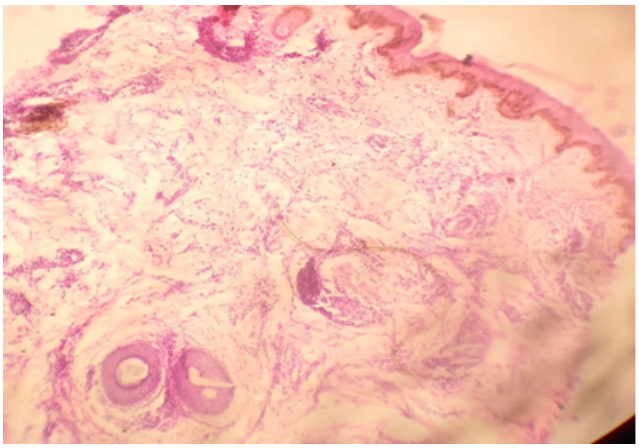

Fig. 10: Photomicrograph showing epidermal atrophy(H&E stain, x10).

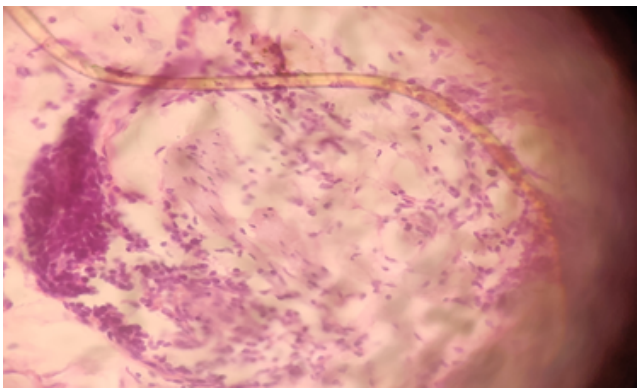

Fig. 11: Photomicrograph showing perineural lymphocytic infiltration present(H&E stain, x40).

to kottucherry government hospital and was started on antileprosy treatment (multidrugtherapy).

4. Discussion

Leprosy is a chronic disease, which remains active for a long time. During the long course of activity of the disease, in several patients, there are acute bouts of exacerbation characterized by sudden increase in the activity of the disease. The word reactions in leprosy are used to describe an episode in a major disease. They can occur in tuberculoid spectra and lepromatous spectrum. Reactions can be defined as sudden tissue responses, resulting from the liberation of the bacilli or their products, into the tissues. Manifestation can be local or systemic. Leprosy is a quiescent disease unless, it is complicated by reactions. Reactions make the patient to seek medical attention. Reactions are responsible for the most permanent nerve damage, deformity and disability. Borderline group of patients are the most vulnerable group with the highest risk of developing Type 1 reaction.[1] Borderline lepromatous and borderline borderline patients have a higher risk than borderline tuberculoid patients.[2] However, reactions occur earlier in Borderline tuberculoid patients.[3] In this study the patient was in borderline tuberculoid spectrum and he presented with reaction thirty years after the appearance of initial lesions. Patients with multiple and disseminated patches involving larger body areas and multiple nerve involvement are at increased risk of developing type 1 reaction.[4–6]Nerve enlargement , tenderness and paraesthesia on palpation are associated with an increased risk of a reaction.[7] Reaction may be present at the time of presentation or develop during treatment and even after release from treatment.[8] At a fully monitored field control unit at Koraput leprosy eradication project, the data showed that type 1 reaction occurred in 3.9 percent of borderline cases. Of the borderline cases , borderline borderline(BB) type showed maximum rate of reactions. The borderline lepromatous (BL) type can present with both type 1 and type 2 reactions with total incidence of 12.8 percent. While borderline tuberculoid(BT) type constituted 74 percent of the total cases, Type 1 reaction occurred in only 3.1 percent cases. Reaction also occurred in 0.8 percent of release from treatment cases.[9] Type 1 reaction occurs as a result of increased activity of the body's immune system, particularly cell mediated immune response fighting the leprosy bacillus or remnants of dead bacilli. Clinical features include reaction may be the first presenting sign of the disease and usually last for few weeks to few months. General condition of the patient is satisfactory. Usually there is no fever and patient does not feel ill. There will be increase in inflammation in skin lesions or nerves or both. Skin lesions becomes erythematous and/or edematous, and may ulcerate. Edema of hands, feet and face can also be a feature of reaction. Systemic symptoms are rare. Neuritis is present if the patient has nerve pain, paraesthesia or tenderness, which precedes nerve function impairment. This is seen in our case. If left unnoticed it may lead to silent neuropathy. Immunology of Type 1 reaction is associated with upregulation of interferon-gamma production leading to granuloma formation, enhanced macrophage microbicidal activity and inflammation. This reaction is associated with enhanced bacterial clearance.

Erythema nodosum leprosum or type 2 reaction is an acute inflammatory condition involving a TNF alpha and immune complex mediated immune response. Type 2 reaction occurs mostly during the course of antileprosy treatment. A few cases present for the first time with features of reaction before leprosy is diagnosed and treatment started. Here also the patient presented with features of reaction before leprosy is diagnosed and before the treatment has started. Most frequently seen in lepromatous leprosy patients. TNF-alpha is known to be a pyrogen which may be responsible for the rise of temperature and further tissue damage during type 2 reaction.[10] Precipitated by physical or psychological stress, pregnancy, puberty, lactation or alcohol intake. Erythema nodosum leprosum precedes fever, joint pain, malaise, and headache. Subnormal temperature in morning and high in evening. The fresh crops of erythema nodosum leprosum lesions usually appear in the evening when endogenous cortisol production is at its lowest.[11] Skin lesions are brightly erythematous, slightly raised nodules or plaques, variable in size. Nodules measuring 0.5 to 3cm on the face, thigh, legs. Nodules disappear in few days and successive crops appear. Upon resolution, leave purplish stain and desquamate. Neuritis is less compared to type 1 reaction

5. Conclusion

Hansen's disease is a chronic infectious granulomatous disease with varied presentation, especially in the setting of lepra reactions. So here we report two such atypical presentations each of Type 1 and Type 2 Lepra reactions.

6. Source of Interest

None.

7. Conflict of Interest

None.

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Author biography

Tony Kuncheria Post Graduate

V Siva Subramanian Professor and HOD

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