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Original Article

Atypical Manifestations of Leprosy – A Case Series

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Atypical manifestations are not rare in leprosy and they may present diagnostic challenges. We report a series of five cases having atypical presentations. First case presented with an asymptomatic buccal lesion and later developed skin lesions. Second patient had secondary Anti-phosphospholipidsyndrome (APS) due to leprosy. We also report another interesting case of annular vesiculobullous lesions and erythema nodosumleprosum (ENL) following intake of antibiotics for pneumonia. Other two had exacerbation reactions (ER). Two of these patients were had irregular/incompleteanti-leprosy treatment earlier. These profiles will be of interest to clinicians who may encounter cases with such manifestations.

Key words : Secondary APS, Annular vesiculobullous ENL, Leprous exacerbation, atypical, leprosy

Introduction

Leprosy presents with varied manifestations and in post elimination era we should have an open eye for early diagnosis. Its emphasized in many studies that late diagnosis due to quiescent nature of multibacillary cases, lack of active case detection, increase in migrant population, social, cultural and economic inequalities are the main reasons for the rising trend. Here we report a series of five cases of atypical presentations of leprosy.

Case – 1

A 40 year old female presented with an erythematous swelling of right buccal mucosa of 3 years duration without any skin lesions. (Fig 1). Biopsy revealed non caseating granulomatous lesion possibly sarcoidosis. On follow up

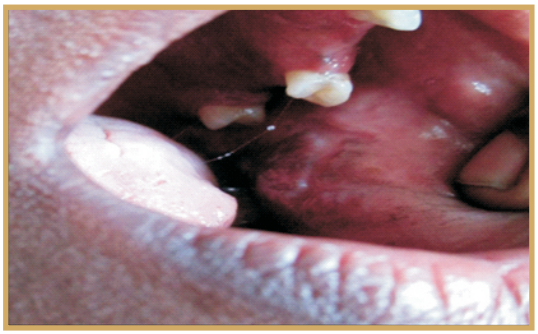

Fig 1 : Erythematous swelling of right buccal mucosa

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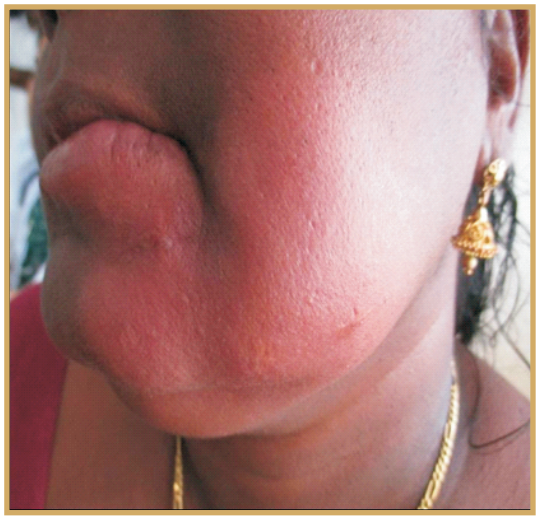

Fig 2 : Oedema of lip and a raised erythematous tender plaque

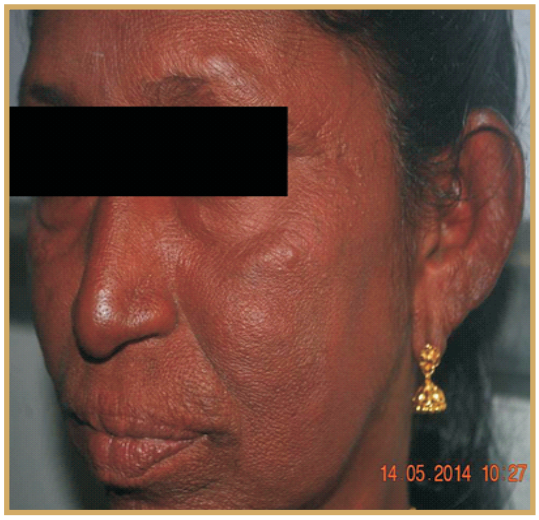

Fig 3 : Earlobe thickening

examination she showed edema of lip and a raised erythematous tender plaque of size 5x4 cms on same side of face (Fig 2). Sensory impairment was present over the lesion. ELS|SSS (Ear lobe/slit skin smears) for AFB were negative. Clinical diagnosis of HD BT with type 1 reaction was made.

Biopsy from the facial lesion was confirmatory. Patient was started on Multi-Drug Therapy (MDT) (PB) along with systemic steroids in tapering dose. She responded well to treatment.

Case - 2

A 37 year old female who was a treated case of HD LL 5 years back, presented with erythematous nodular evanescent lesions all over body of 2 months duration and right hemiparesis. She did not have any history of diabetes or hypertension. Physical examination revealed ear lobe infiltration. (Fig 3) Tender evanescent nodules over forearms and thighs, multiple nerve thickening and glove and stocking anaesthesia. Ear lobe smear/Slit skin smear showed Bacillary index BI- 4+ and Morphological index MI-0.

CT brain showed infarct left internal capsule, she was investigated for young stroke and found ANA weakly positive, Anticardiolipin antibody Ig M positive in high titre and homocysteine level was normal. Her previous obstetric history were normal. So the final diagnosis was Type 2 reaction with secondary Anti-phosholipid antibody syndrome (APS).

Skin biopsy was confirmatory. She was treated with prednisolone 40 mg tablets in tapering doses over six months, clofazimine 300 mg daily initially and gradually tapered, ecospirin 150 mg daily still continuing. She responded well and is now on follow up.

Case -3

A 30 year old male who was a on irregular treatment for HD LL 4 years back, had pneumonia 1 month back and was treated with cefotaxim, cloxacillin and linezolid presented with annular vesiculobullous lesions all over body and fever of 2 weeks duration. (Fig 4). Examination showed tender lymphnodes, infiltrated papules and plaques without anaesthesia, thickened radial

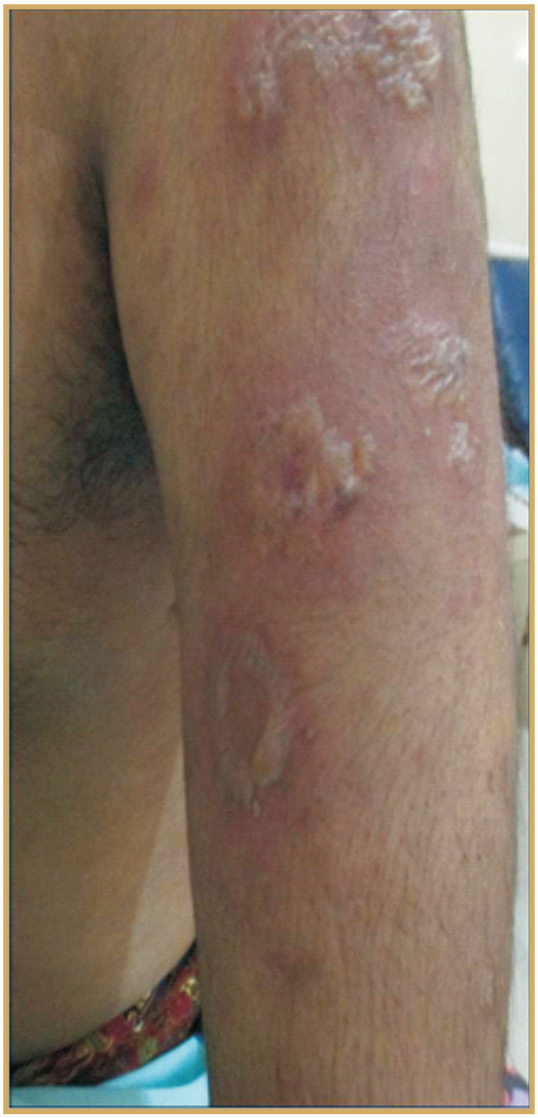

Fig 4 : Annularvesiculobullous lesions all over body

and ulnar cutaneous nerves bilaterally, and glove and stocking anaesthesia.

ELS/SSS- BI 6+, MI-20%, Tzanck smear showed few neutrophils. Skin biopsy was taken from vesiculobullous lesion and plaque which revealed HDLL with ENL.

So diagnosis was HD LL Reactivation with annular vesiculobullous ENL. Patient was started on MDT (MB) with oral prednisolone 40 mg/day which was tapered gradually over a period of six months and he improved.

Case - 4

A 55 year old male who was a diagnosed case of HD LL one and half years back took MB MDT for 5 months and stopped by himself presented with hyper-pigmented plaques with some showing ulceration over face, upper limb and lower limbs of one year duration. (Fig 5) Ear lobe nodularity, multiple nerve thickening and glove and stocking anaesthesia were also present. Sensation over the lesions was intact.

Two other family members had successfully completed treatment for leprosy.

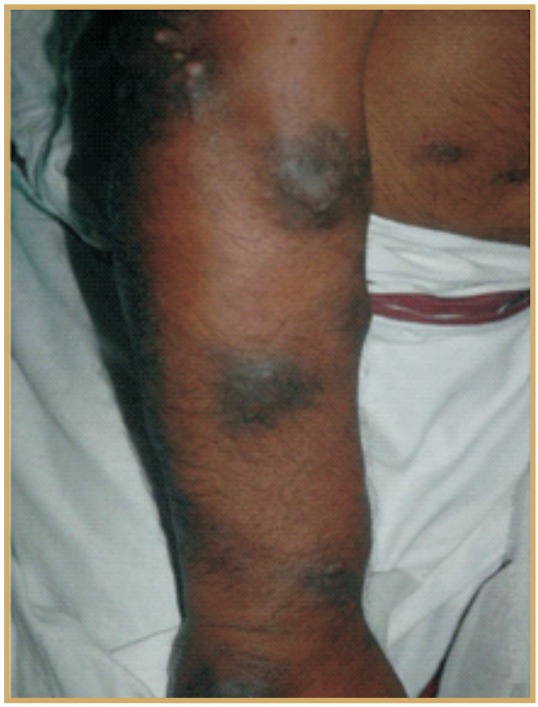

Fig 5 : Hyperpigmentedplaques, some showing ulceration

ELS/SSS-BI 5+ MI 60% skin biopsy-HD LL no features of reaction.

Diagnosed as Lepromatous leprosy Reactivation, with leprous exacerbation. Patient was started on MDT (MB) and he responded with decreasing MI and he is on follow up.

Case - 5

A 22 year old male, migrant from Darjeeling who was a case of HD LL treated one year back with MB MDT for I year, presented with raised hyper-pigmented lesions with some showing ulceration and vesiculation of 1 month duration. (Fig 6) On examination ear lobe infiltration, thickened radial and ulnar nerves and glove and stocking anaesthesia were present, sensation over the lesions was preserved.

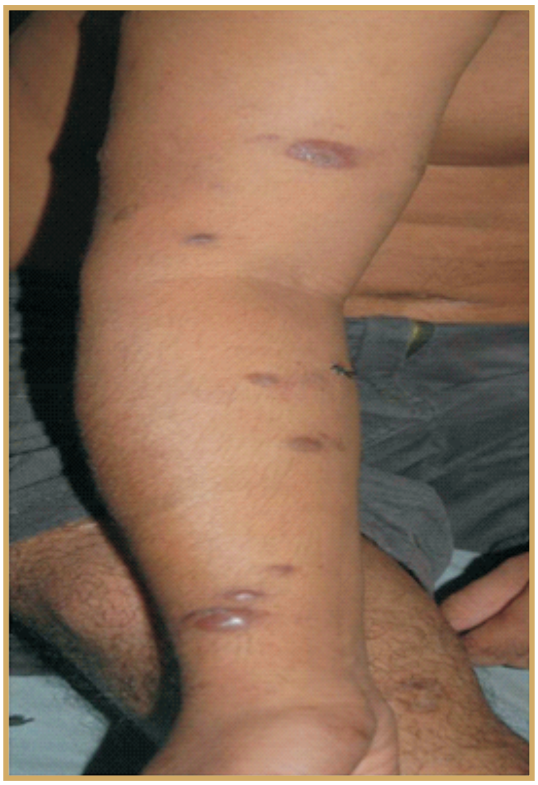

Fig 6 : Raised hyperpigmented lesions with some showing ulceration and vesiculation

ELS/SSS- BI 6+ MI- 60% Skin biopsy- HD-LL

Diagnosed as Lepromatous leprosy Relapse with leprous exacerbation.

Patient was started on MDT (MB), but was lost in follow up as he went back to his native place.

Discussion

Leprosy can present with varied manifestations that can mislead a practitioner in this post elimination era. The reported atypical manifestations include lesions mimicking angioedema, lupus vulgaris, cutaneous lymphoma (Raval 2012), granuloma annulare, erythema multiforme (Das et al 2007), single plaque LL, verrucous lesion (Yuchua-Guillenand Dofitas 2012), non healing ulcer, infiltrated linear lesions (Raiol et al 2014), nerve abscess etc.

Involvement of the oral cavity in leprosy is variable, seen in 19-60% of the patients, almost all patients in lepromatous spectrum. Scheepers (1998) found the hard palate to be the most frequent site of oral involvement in leprosy, followed by the soft palate, gingiva, tongue, buccal mucosae and lips. The buccal mucosa may appear paler than normal. In advanced cases, there may be diffuse infiltration, swellings, papules, nodules and ulceration in the buccal mucosa. Martinez et al (2011) reported a case of Borderline tuberculoid leprosy presenting ashypoastheticoral lesion without any skin lesion with nonspecific histopathology. In our patient also skin lesion was absent initially and biopsy was not suggestive. Infiltrated plaque is considered to be a leprosy specific lesion in buccalmucosa.

Loizou et al (2003) studied 112 leprosy patients & found elevated titers of APLA in 29%, anti β2 glycoprotien 1 (anti b2GPI) in 89%, & anti-Prothrombin in 21% of them.[7] Initially, it seemed that infection induced antiphopholipid antibodies (APLA) are not associated with the thrombotic manifestations of APS. This was

attributed to the fact that the binding of autoimmune APLA to phospholipid is enhanced by the cofactorβ2 GPI (i.e. β2 GPI dependent) while the binding of infection induced APLA is not enhanced by this cofactor (i.e. β2 GPI independent). Recent studies, however show that the APLA in leprosy patients are heterogeneous with respect to their β2 GPI requirement (Arvieux et al 2002). In 10 of 31 leprosysera, the APLA were β2 GPI dependent & 16 of 31 were β2GPI independent. The clinical implications of this β2GPI dependency are seen in Lucio's phenomenon in which the histopathological findings are related to micro vascular thrombosis in the absence of inflammatory infiltration of the vessel wall. It has been hypothesized that certain infections in genetically predisposed individuals may induce these APLA.

There are reports of gangrene in a case of Borderline tuberculoid leprosy associated with APLA (Akerkarand Bichile 2005) and a patient with lepromatous leprosy developing DVT with a positive APLA (Kaliyadan et al 2009). In both these cases Anticardiolipin antibody Ig M was positive, as in our patient.

There are reports of vesiculobullous lesions in type 2 reaction occurring in individuals with leprosy on ofloxacin (Kar et al 2009). In our patient it may be infection induced or antibiotic induced. Histopathology of lesions was consistent with ENL. This patient had relapse and was treated with ofloxacin based regimen.

Exacerbation reactions (ER) have been differentiated from ENL, histopathologically, bacteriologically and immunologically. Most obviously, the difference is that ER occurs in hyperactive lepromas, ENL in regressing granulomas. In common, both ER and ENL commence with polymorphs infiltrating in areas of macrophage cell death. The macrophages in ER are large and cytoplasmically active, containing increased numbers of solid organisms, while those in ENL lesions are effete, with poorly detectable degraded bacterial debris. The localized, compact, although sometimes extensive, distribution of ER contrasts with the more diffuse spread of severe ENL.

In both of our patients, features of reaction were absent both clinically and histologically even though there was an increase in size of lesions.

ER mainly seen in very advanced lepromatous patients with nodular and plaque like lesions. Histologically there are small localized areas of necrosis in the middle of a large sheet of macrophages eliciting a localized infiltration of neutrophils. Vasculitis is rarely seen. The macrophages contain a relatively largeload of acid fast bacilli which differentiate it from ENL (Karand Sharma 2010). This is due to sudden burst of bacterial multiplication which overgrows the macrophage population. These types of presentations were reported in pre MDT era, and last three patients were treated with alternative regimens.

Conclusion and Future Perspective

Atypical manifestations in this series simulate the presentations in pre MDT era especially leprous exacerbations and vesiculobullous ENL. Though it does not appear important in the present cases, the development of drug resistance should be thought of as a cause for relapse, and drug resistance testing facilities should be made more available. Irregularity of treatment were noticed in two patients which later might have led to reactivation, so close relatives should be not only examined for disease but also counseled regarding need of continuous full course of treatment for the patient. On completion of treatment patients should be reassessed for the disease status and regularity of treatment. Surveillance after completing MDT is also a need

of time for early detection of relapse. Clinical suspicion and accurate history taking is very important to diagnose such cases.

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