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Unusual Presentations of Lepromatous Leprosy - A Case Series

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Abstract: ***Leprosy is a great imitator which can have various clinical presentations mimicking many other diseases in the field of medicine. Being at the poor immunity spectrum of the disease, lepromatous leprosy (LL) can cause physical morbidity and disability leading to social stigma. Physicians must have knowledge regarding these unusual presentations so that early identification and prompt treatment of diseases can be done in order to prevent the complications and also its impact on the patient's life.***

Keywords: Lepromatous Leprosy, Unusual Presentations, Case Series, Diagnosis, and Treatment

1. Introduction

Leprosy is a chronic granulomatous disease caused by mycobacterium leprae which primarily affects the skin and peripheral nerves. Lepromatous leprosy (LL) usually presents with symmetrical hypopigmented macules, papules, nodules, madarosis and diffuse infiltration of skin and ear lobe with glove and stocking anaesthesia. It can also have other various perplexing cutaneous presentations. Most common atypical presentations include histoidhansens, lazarine leprosy, lucio leprosy and pure neuritic type. This case series is a retrospective analysis of various unusual presentations of lepromatous leprosy.

Case 1:
20 year old male presented with multiple asymptomatic skin lesions over the left shoulder (fig.1a) for the past 9 months. Dermatological examination revealed multiple erythematous papules and plaques with scaling arranged in a linear pattern. Auspitz sign was negative. Rest of the skin, hair, nails and mucosa was normal. Provisional diagnosis of Linear psoriasis/linear lichen planus was made.

To our surprise, biopsy (fig.1b) was suggestive of lepromatous leprosy with positive fitefaraco.

Peripheral nerve, sensory and motor examination was found to be normal. Following which slit skin smear from the lesion showed a positivity of 6+ with globi formation (fig.1c).

Case 2:
35 Year old male presented with multiple asymptomatic nonprogressive skin lesions over the back for more than 4 years. On examination, multiple skin coloured papules coalescing to form plaque which was soft in consistency and nontender with few areas showing erythema (fig.2a). Provisional diagnosis of connective tissue nevus was made and biopsy was done which opened a new window. fig.2b). Slit skin Smear showed a positivity of 6+ (fig.2c).

Case 3:
22 Years old female, a software engineer by profession, presented with a progressively increasing swelling over the left ankle for the past 10 months. O/E single large irregular plaque of size 5x6 cm, studded with multiple nodules of varying sizes was present over the medial malleolus of left ankle. It was indurated, non tender with no discharge and not associated with regional lymphadenopathy (fig 3a). Our provisional diagnosis was chromoblastomycosis/mycetoma.

Biopsy was done and tissue smear for KOH was negative for fungus. Histopathological examination showed features suggestive of lepromatous leprosy (fig3b). No fungal elements seen. FiteFaraco stain was positive.
Tissue culture was found to be negative for fungus. . Slit Skin Smear showed 6+ (fig 3c).

Case 4:
A 40 Year old male presented with a single asymptomatic nonprogressive painless ulcer over the right medial malleolus. D/E showed single well defined punched out ulcer of size 4 *4cm over the right medial malleolus. Floor was covered with slough and on palpation there was no tenderness (fig.4a). Provisional diagnosis was non healing ulcer for evaluation.

Pus culture and sensitivity was done and appropriate antibiotics given. Ulcer did not show any improvement. ANA profile, cANCA, pANCA were done which turned out to be negative. Finally touch smear threw a light into the diagnosis of lepromatous leprosy by showing the presence of lepra bacilli and biopsy from the edge of the lesion proved the same (fig.4b). SSS was also confirmatory.

Case 5:
65 Year old Male presented with asymptomatic multiple skin coloured lesions over the chest and back for 10 months. On examination, multiple skin coloured atrophic wrinkled plaques admitting the finger with rim of normal skin present over the lower back and chest (fig 5a). Peripheral nerves, sensory and motor examination was normal. Provisional diagnosis –Anetoderma for evaluation.
Other causes of secondary anetoderma was ruled out.

Biopsy helped us to arrive at a diagnosis of lepromatous leprosy (fig 5b). Fitefaraco stain was positive (fig 5c). Van gieson staining - Elastolysis+ (fig 5d).

Case 6:
60 year old male came with multiple skin lesions for last 1 year. On examination, multiple atrophic plaques which was yielding on pressure was present involving chest and abdomen (fig.6a). Peripheral nerves, sensory and motor examination was normal. Provisional diagnosis of Anetoderma for evaluation was made.

Histopathological examination showed thinned out epidermis and dermis contains foamy macrophages and loss of elastic fibers. Bacteriological index of slit skin smear was 5+ (fig.6b)

Case 7:
25 Year old male presented with complaints of pain and difficulty in using right upper limb for 7months without any skin lesions. Grade 1 thickening of right ulnar nerve was present. Other peripheral nerves were normal. No sensory or motor deficit was present. Provisional diagnosis was neuritis for evaluation. Nerve conduction study showed both sensory and motor axonal neuropathy of right ulnar nerve. Nerve biopsy showed reduction of myelinated fibers, presence of foamy macrophages (fig 7) presence of acid fast lepra bacilli.

Case 8:
22 year old male presented with complaints of gradually progressive skin lesion over face without photosensitivity for 4 months. On examination, single arcuate skin coloured edematous plaque was present over left side of face near nasolabial fold (fig.8). Diascopy did not reveal apple jelly nodules. Peripheral nerves, sensory and motor examination was normal. Provisional diagnosis of lupus vulgaris/lupus pernio was made.

ANA was negative, Mantoux showed positive results with induration of 20 mm. Pulmonary tuberculosis was ruled out. Finally biopsy done showed features suggestive of lepromatous leprosy (fig.8a). FiteFaracco was positive. Slit skin smear was positive (fig.8b).

Summary of all the 8 cases discussed above.

S. No.	Age	Sex	Provisional Clinical Diagnosis
1	20	Male	LINEAR PSORIASIS / LINEAR LICHEN PLANUS
2	35	Male	NAEVI
3	22	Male	CHROMOBLASTOMYCOSES/ MYCETOMA
4	40	Male	NON HEALING ULCER
5	65	Male	ANETODERMA (A)
6	60	Male	ANETODERMA (B)
7	25	Male	NEURITIS FOR EVALUATION
8	22	Male	LUPUS VULGARIS/LUPUS PERNIO

2. Discussion

Lepromatous leprosy usually presents as symmetrically distributed infiltrated papules, nodules, plaques and also diffuse infiltration along with many symptoms and signs such as madarosis, gynaecomastia, pedal edema, nasal stuffiness, epistaxis, hoarseness of voice, glove and stocking anaesthesia. Most of the times, patients in the lepromatous spectrum rarely present to us in the early stages as there are no symptoms of nerve involvement earlier and early skin lesions are not likely to be noticed by the patient. This is doubly unfortunate, for not only is the patient infectious and disease can progress to form early deformities.

After the advent of MBMDT in 1981, the progression of the disease to lepromatous pole has been decreased, disabilities are prevented and overall the curability rate of Leprosy is also increased. Now being in the elimination era of the disease, we face many atypical presentation of the same which makes diagnosis and treatment difficult.

A typical presentations of lepromatous leprosy reported so far includes erythema gyratumrepens like 1,, molluscumcontagiosum like 2 lesions, single plaque3, zosteriform lesion, anetoderma like, Iongstanding ulceration, erythema multiforme like lesion, granuloma annulare like lesion, adenomasebaceum like, asymptomatic buccal lesions 4, secondary antiphospholipid syndrome 4, sweet's syndrome like5and verrucous lesions.

All these presentations had intact sensation at the time of presentation on the contrary which was the unique finding noted. Histopathology came for the rescue and played a major role in concrete diagnosis in our cases. Thus all suspected cases, especially in endemic regions, should be subjected to histopathologicexamination. There is a need to keep this infective condition as an alternate diagnosis to all unusual cutaneous lesions as there are continued reports of atypical presentations in the post elimination era. Our thorough literature search did not reveal any explanation for this atypical presentation. Probably localisation of high bacillary load with poor cellular immunity may be the convincing pathology. The propensity of leprosy for unusual presentations is likely to lead to an undue delay in the correct diagnosis.

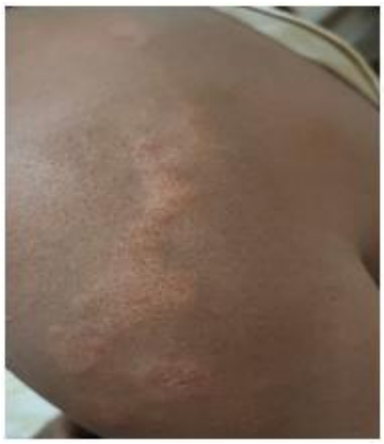

Figure 1 (a): Shows multiple erythematosus papules & plaques with scaling in linear distribution

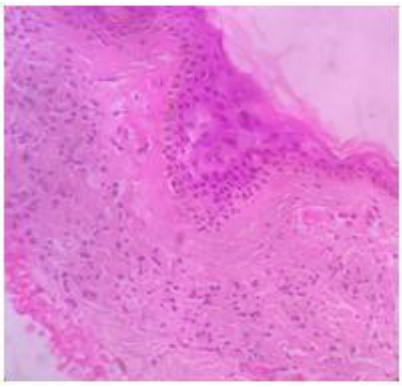

Figure 1 (b): HPE image

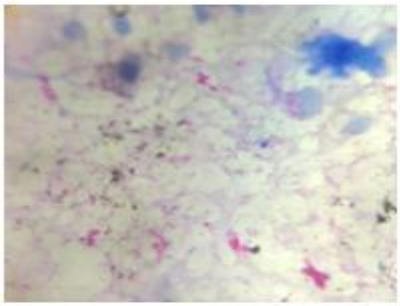

Figure 1 (c): Slit skin smear image

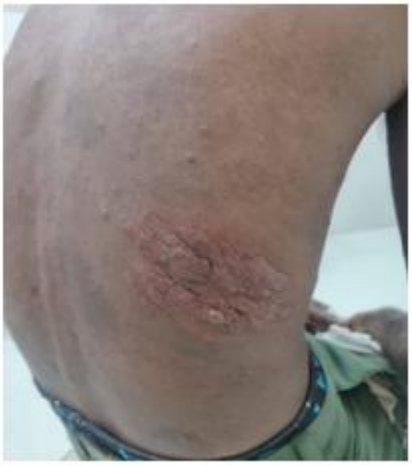

Figure 2 (a): Shows multiple skin coloured to erythematosus soft nontender papules coalescing to form plaques

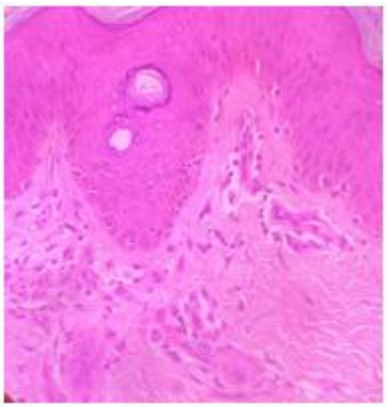

Figure 2 (b): HPE image

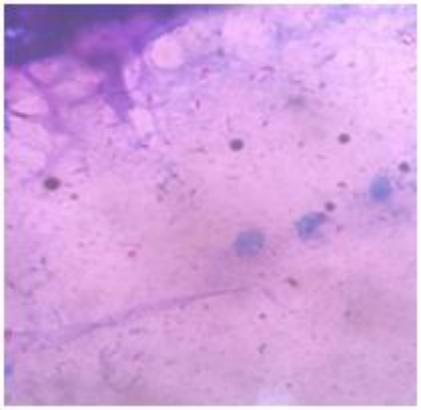

Figure 2 (c): Slit skin smear image with leprabacilli

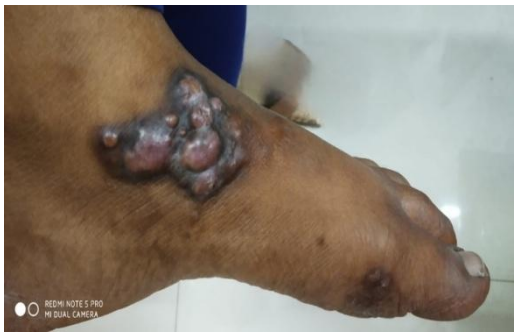

Figure 3 (a): Shows single large plaque studded with multiple nodules

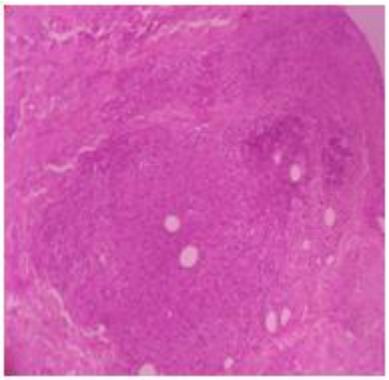

Figure 3 (b): Histopathological examination showing diffuse infiltration of dermis with foamy macrophages

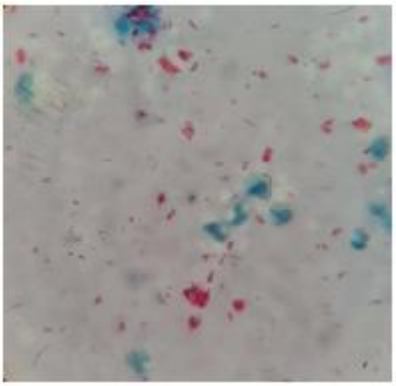

Figure 3 (c): Shows slit skin smear with globi formation

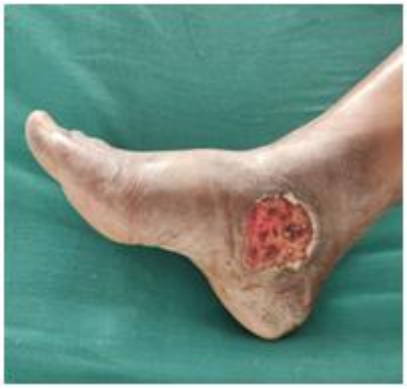

Figure 4 (a): Shows single well defined punched out ulcer with floor covered with slough

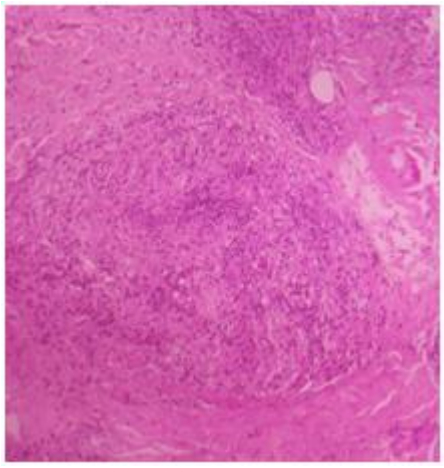

Figure 4 (b): HPE image

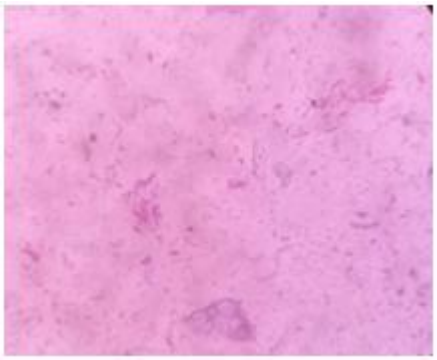

Figure 4 (c): Slit skin smear with globi formation

Figure 5 (a): Shows multiple skin coloured atrophic wrinkled plaques

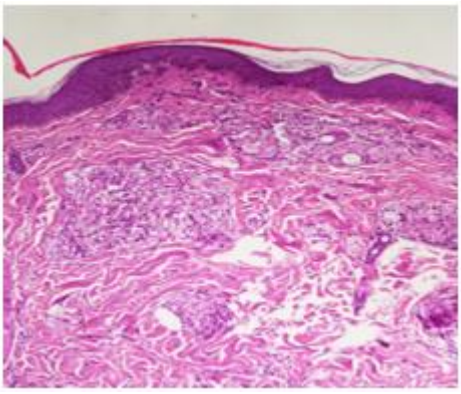

Figure 5 (b): Bhistopathological examination of hematoxylin and eosin stained smear showing infiltration with foamy macrophages

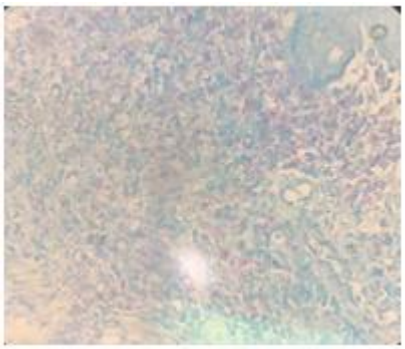

Figure 5 (c): Fitefaracco staining showing bacilli

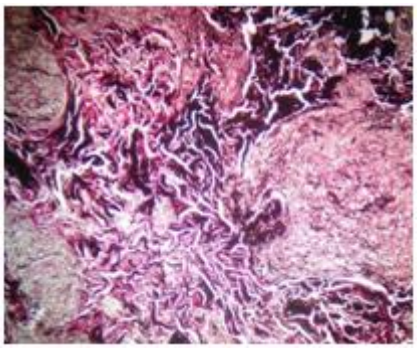

Figure 5 (d): Shows tissue section showing positive staining for elastin in vangieson staining suggestive of elastolysis

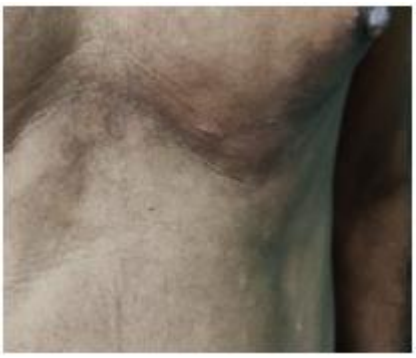

Figure 6 (a): Shows multiple atrophic plaques on chest and abdomen

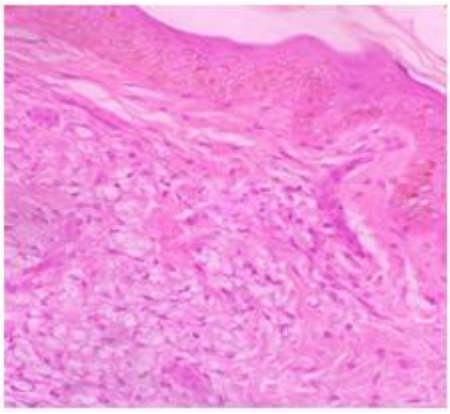

Figure 8 (b): HPE image with infiltration of foamy macrophages

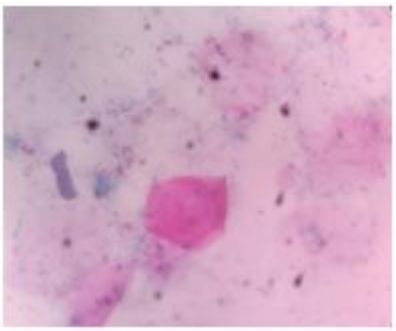

Figure 6 (c): Slit skin smear showing leprabacilli

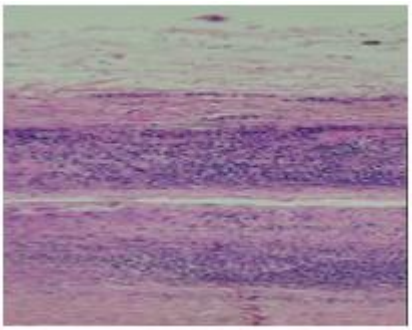

Figure 7 (a): histopathological examination of the nerve tissue showing infiltration with foamy macrophages

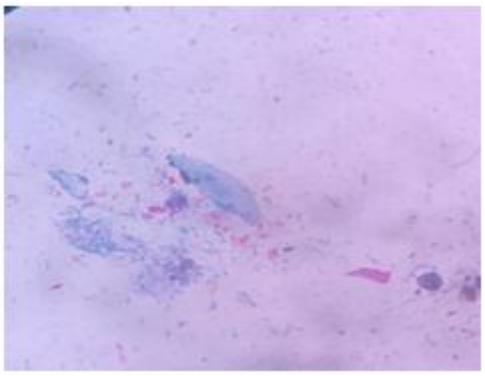

Figure 7 (b): Slit skin smear with lepra bacilli

Figure 8 (a): Shows single arcuate skin coloured edematous plaque near the ala of left side of nose

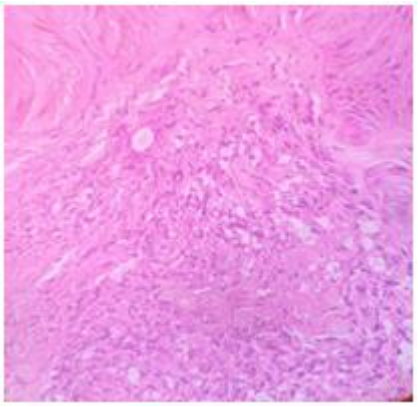

Figure 8 (b): HPE image

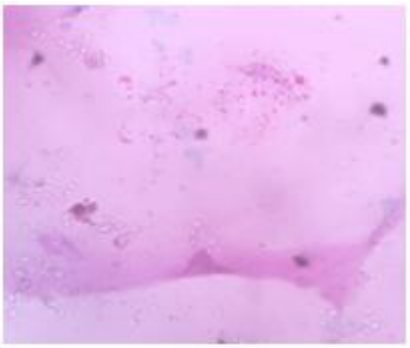

Figure 8 (c): Slit skin smear with bacilli

3. Conclusion

Leprosy is a great mimicker and can have variety of clinical presentations. Without timely diagnosis and treatment, it can lead to disfigurement, paralysed extremities and physical disabilities. Leprosy should be strongly ruled out in all suspected cases so as to bring about early induction of treatment & prevention of deformities and disabilities in affected patients.

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Organized State conference - CUTICON TN - 2019
National SOP for Newborn skin approved by IAP

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