Various faces of Hansen's disease

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Leprosy is a chronic granulomatous disease caused by *Mycobacterium leprae*. Leprosy once considered a taboo is still misdiagnosed and underdiagnosed. In many cases leprosy is treated as common disorders like psoriasis, pyoderma, angioedema, pre vitiligo. Leprosy can present in many diverse ways which can be confused with many treatable and non treatable, infectious and non infectious forms. Leprosy is considered on the verge of elimination. But Leprosy cases are being newly diagnosed day by day. Here we are presenting 4 atypical cases of leprosy which did not seem to have classical presentation but were diagnosed as leprosy when investigated.

Keywords: Leprosy, Angiodema, lepromatous leprosy

Introduction

Leprosy is a chronic infectious disease caused by *Mycobacterium leprae*, an acid-alcohol resistant bacillus with affinity for skin and peripheral nerve cells. This disease can present a wide range of manifestations, including secondary to systemic involvement. Here we present 4 cases of atypical manifestations of leprosy manifesting as angiodema, lesions mimicking lupus vulgaris, mimicking cutaneous T cell lymphoma, lepromatous leprosy with single plaque with gynaecomastia and non healing ulcer over right finger.

Case report: 1

Tuberculoid Leprosy Mimicking Angioedema

A 21 year old male presented with complaint of edema over right side of forehead involving both

upper and lower right eyelids (Figure 1) since 15 days. No history of fever, malaise, numbness over the lesion was present. Patient gave no history of trauma or intake of any drugs. Patient was earlier treated as angioedema with no improvement in the clinical condition. On general examination, vital parameters were normal. On examination, non tender erythematous swelling involving the right upper and lower eyelids extending towards the right side of the forehead was present. On reevaluation touch and temperature sensations were altered over the lesion though there were no palpable or tender nerves. Routine investigations were in normal range. A punch biopsy from the lesion was taken. On histopathology (Figure 2) epitheloid granuloma was seen in the dermis with giant cells and moderate lymphoplasmacytic infiltration suggestive of

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Fig 1: Erythematous swelling involving the right upper and lower eyelids extending towards the right side of the forehead.

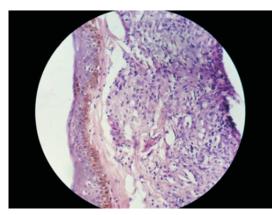


Fig 2: Epitheloid granuloma in the dermis with giant cells and moderate lymphoplasmacytic infiltration of chronic granulomatous inflammation.

chronic granulomatous inflammation. Stain for lepra bacilli was negative and fundus examination was normal. Patient was diagnosed as tuberculoid leprosy.

Case report 2

Leprosy Mimicking Lupus Vulgaris

A 55 year old female presented with ulcer over the left jaw (Figure 3) since 7-8 months and



Fig 3: Ulcer over the left jaw.



Fig 4: Red elevated lesion over the left forearm

a red elevated lesion over the left forearm (Figure 4) which was recent in onset (1-2 months). No history of fever, malaise, cough, chestpain, breathlessness, tingling, numbness or trauma was present. On examination a well defined ulcerated necrotic lesion was present over the left mandibular area with tender firm cervical lymphadenopathy and a saucer shaped erythe-

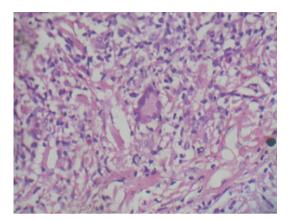


Fig 5: Foamy histiocytes with subepidermal narrow grenz zone. 5% AFB showed few acid fast bacilli. BI (Bacilli Index): 1.

matous tender infiltrated plaque with central clearing and few satellite lesions were present over the left forearm. Some red elevated lesion was present preceding the ulcer. Patient was being treated as Lupus Vulgaris and had been given AKT for 6 months with partial response.

Touch and temperature Sensations were altered over the plaque. Cervical lymph nodes were palpable, tender and firm. Other vital parameters were normal. Routine investigations were in the normal range. Serum HIV, S.VDRL, Mantoux test were negative. Chest X-ray showed no abnormality. A Punch biopsy was taken. On Histopathology (Figure 5) foamy histiocytes were seen with subepidermal narrow grenz zone. 5% AFB showed few acid fast bacilli. BI (Bacilli Index): 1.

Diagnosis was kept as Tuberculoid leprosy with type 1 reaction.

Case Report 3

Tuberculoid Leprosy Mimicking Cutaneous T Cell Lymphoma

A 53 year female presented with single infiltrated plaque 6x4 cm below chin (Figure 6) since 1 year



Fig 6 : Single infiltrated plaque 6x4 cm below chin.

and red asymptomatic lesions over the left forearm, arm and medial side of right thigh since 6 months. O/E: Multiple erythematous asymptomatic nodular lesions were present on left forearm and arm and medial side of right thigh. Single infiltrated plaque of approximate size 6*4 below the chin was seen with crusting at few places. Hot and cold sensations were normal on the lesions. No nerve enlargement was present. No complaint of epistaxis, joint pain, slipping of chappals, fever and headache was present. Differential diagnoses considered in this case were cutaneous T cell lymphoma and tuberculoid leprosy. Histopathology (Figure 7) showed thinned out epidermis at some places with dermis showing multiple granulomas comprising of lymphoplasmocytic cells, epitheloid cells and 158 Raval

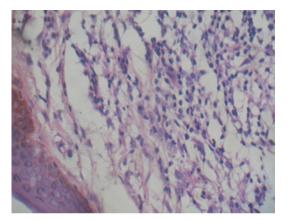


Fig 7: Multiple granulomas comprising of lymphoplasmocytic cells, epitheloid cells and langhan's type of giant cells.

langhan's type of giant cells. Fibrinoid necrosis was present suggestive of tuberculoid leprosy in type 1 reaction with 5% AFB negative.

Case report 4

Single Plaque with Lepromatous Leprosy

A 55 year old male presented with tingling and numbness over hands and feet and history of slipping of chappals since 8 months, non healing ulcer over right little finger (Figure 8) since 8 months. On examination patient had leonine



Fig 8: non healing ulcer over right little finger.

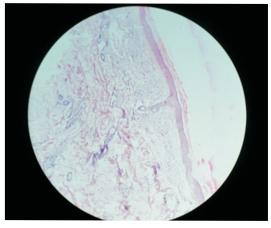


Fig 9: Dermis reveals aggregates of foamy histiocytes and subepidermal narrow grenz zone of collagen tissue was present.

facies, gynecomastia, a diffuse infiltrative plaque was present over back. Hot and cold sensations were altered over hands and feet distally and even on the lesion. Also touch sensation was altered over both extremities. Both Ulnar nerves, lateral popliteal nerves and posterior tibial nerves were enlarged. On histopathology (Figure 9) dermis reveals aggregates of foamy histiocytes. Subepidermal narrow grenz zone of collagen tissue was present. Histiocytic cell collection was seen perineurovascularly in the dermis. Acid fast bacilli are seen on 5% AFB staining. Routine investigations were normal.

Diagnosis was kept as Lepromatous Leprosy with BI: 4

Discussion

The clinical presentation of leprosy is highly variable and in all its stages it can mimic great variety of other lesions. The differential diagnosis is so wide that one has to exclude wide variety of dermatological diseases before stamping it to be leprosy as stigma is still associated to it. Even neurological conditions and lepra reactions are to

be differentiated from a number of systemic illnesses (Sharma and Mahajan 2010).

Our first case was being treated for angioedema on account of short history but patient didn't respond. Also the lesion was asymptomatic and on re-evaluation pt was having altered sensations. On histopathology granulomatous reaction was seen. So diagnosis of tuberculoid leprosy was put. Also many of cases of tuberculoid leprosy mimick angioedema and urticaria. These entities are considered to be one of the differential diagnosis of papulo-plaque lesions (Sharma and Mahajan 2010).

In our second case, patient is having tuberculoid leprosy with type 1 reaction. Patient was having well defined ulcer over left jaw area characterized by necrosis, deep ulceration and elevated plaque over the left forearm. Patient on the basis of her ulcerated lesion with lymphadenopathy was being treated as a case of lupus vulgaris but showed partial improvement. But the lesion on her forearm after being evaluated showed affected sensations and granulomatous response was seen in the biopsy which pointed to the diagnosis of borderline tuberculoid leprosy in type 1 reaction. Lupus vulgaris is common over the face, has variable morphology and has to be differentiated from tuberculoid leprosy. It is also characterized by significant tissue destruction and ulceration (Sharma and Mahajan 2010). Necrosis and ulceration are seen in severe cases of lepra reactions (Kar and Sharma 2010). Also unusual expression of BT leprosy is spontaneous ulceration of skin lesions. This is the result of exaggerated hypersensitivity in type 1 reaction. Rarely it occurs in the absence of any history of skin lesions or coexistent nerve pathology (Kumar and Dogra 2010).

In our third case, with unaltered sensations over the lesions and looking at the age of the patient our clinical expression was of cutaneous T cell lymphoma but the biopsy pointed to the diagnosis of tuberculoid leprosy. Also on further evaluation her son and daughter in law both had 2-3 welldefined hypopigmented lesions with slightly elevated borders present on their buttocks which when biopsied showed histology resembling tuberculoid leprosy with epitheloid granuloma. Mycosis fungoides may present with pruritic atrophic plaques and red shiny tumors over face and trunk but biopsy is diagnostic (Sharma and Malhotra 1994).

In our fourth case, patient had gynaecomastia but a single diffuse plaque over rt side of the back with non healong ulcer over right little finger. All his routine investigations were normal. On biopsy of the plaque it showed lepromatous leprosy.

Rarely a single nodule or plaque may be the presenting feature. It may be shiny and have sloping margins. The biopsy reveals borderline lepromatous or lepromatous histology and the lesions are full of acid fast bacilli. The rest of the skin is entirely normal and slit skin smears are negative. Such lesions may be responsible for some of the relapses in paucibacillary patirnts. This highlights the value of using slit skin amears in suspected lesions (Sharma and Malhotra 1994).

Conclusion

Leprosy mimicks many dermatological and neurological disorders. So in an endemic country like India we need to still have to an eye to recognize the varied manifestations of leprosy. We are presenting these cases on account of their rare occurrence.

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