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Histopathology

LEPROSY MASQUERADING AS SWEET SYNDROME WITH FEW BULLOUS LESIONS – AN UNUSUAL PRESENTATION

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ABSTRACT Leprosy is a chronic granulomatous infection of the skin caused by Mycobacterium leprae. It is one of the most common dermatological infections in India presenting with hypopigmented anaesthetic patches. Some patients can present with lepra reactions as the initial manifestation, however the occurence of Sweet-like Erythema Nodosum Leprosum (ENL) with few bullae is extremely rare with only few cases reported in literature. Unusual and rare clinical presentation leads to difficulty and delay in the correct diagnosis & initiation of appropriate treatment. We hereby present a case report of a 27 year old lady, who presented with reddish raised skin lesions over bilateral upper and lower limbs for 3 days. History of fever and pain was present. O/E: Multiple well to ill defined erythematous plaques of varying sizes were present in both upper and lower limbs. A diagnosis of Sweet syndrome was made based on the clinical presentation. Histopathological diagnosis was borderline lepromatous leprosy with erythema nodosum leprosum. Though rare, Erythema Nodosum Leprosum (ENL) can be the initial clinical presentation of leprosy. Hence, in the absence of classical clinical features, an adequate skin biopsy including subcutaneous fat is required for a definite diagnosis of this condition.

KEYWORDS : Sweet-like ENL, leprosy, globi, reaction, bullae.

INTRODUCTION:

Leprosy is one of the most common dermatological infections in India presenting with hypopigmented anaesthetic patches. Whenever there is sudden changes in immune mediated response to antigens of Mycobacterium leprae, lepra reactions can occur which manifests as acute or subacute episodes of inflammation[1]. These reactions usually occur during or after treatment of leprosy with antileprosy drugs. Though rare, Erythema Nodosum Leprosum (ENL) can be the initial clinical presentation of leprosy. If there is no clinical suspicion or if biopsy is not performed, such cases can be missed. Rare and unusual clinical presentation leads to difficulty and delay in correct diagnosis. Hence, in any unusual presentations, an adequate skin biopsy including subcutaneous fat is required for a definite diagnosis of this condition. Herein we report a peculiar case of 27 year old lady who presented with sweet like ENL and few bullous lesion which was diagnosed as Borderline lepromatous leprosy with ENL on biopsy and hence treated appropriately. A clinical diagnosis of leprosy was not sought because the patient never had any pre-existing evidence of leprosy nor had any antileprosy treatment and the clinical presentation was unusual for the diagnosis of leprosy.

CASE HISTORY:

A 27 year lady presented with reddish raised skin lesions over bilateral upper and lower limbs for 3 days. History of fever and pain was present. There was history of similar lesions during the last pregnancy which resolved spontaneously. On examination, multiple well to ill defined erythematous plaques of varying sizes were present in bilateral upper and lower limbs which led to the clinical diagnosis of Sweet syndrome (Fig:1&2). Complete blood counts revealed neutrophilic leucocytosis. Punch biopsies were done from erythematous vesicle and erythematous plaque in Right forearm. Patient was started on steroids and the lesions started resolving. Histopathological examination of biopsy from erythematous vesicle showed marked dermal edema with formation of subepidermal vesicle containing eosinophilic secretions, few scattered lymphocytes and neutrophils. The superficial and the deep dermis showed nodular inflammatory infiltrates comprising of multiple loose granulomas including periadnexal, perifollicular and occasional perineural granulomas composed of epithelioid histiocytes admixed with foamy macrophages, lymphocytes, neutrophils and occasional plasma cells. The subcutis showed few loose granulomas including a perineural granuloma as described above. Few blood vessels in the subcutis showed transmural inflammatory infiltrates of lymphocytes and few neutrophils with focal fibrinoid necrosis – consistent with vasculitis. Biopsy from erythematous plaque showed dermal edema, moderate perivascular inflammatory infiltrates of neutrophils and periadnexal, perifollicular nodular inflammatory infiltrates comprising of multiple loose granulomas as described above. Special stain for acid fast lepra bacilli (Fite- Faraco) showed numerous beaded and solid intact acid fast lepra bacilli forming globi in foci. Histopathological diagnosis of Borderline lepromatous Hansen's disease with erythema nodosum leprosum and subepidermal vesicle with panniculitis was rendered based on the findings. (Fig: 3-5). Lesions resolved after initiation of treatment with steroids. Slit skin smear also showed numerous acid fast lepra bacilli (Fig: 6). Patient was started on antileprosy treatment and on follow up no new lesions appeared.

DISCUSSION:

Leprosy is one of the most common dermatological infections in India presenting with hypopigmented anaesthetic patches. India is one amongst the 22 " global priority countries" which contributes to 95% of the total cases of leprosy world wide[2]. According to World Health Organization (WHO), in 2019-20, around 114,451 new leprosy cases were detected in India accounting for 80% of the cases in Southeast Asian countries. Whenever there is sudden changes in immune mediated response to antigens of Mycobacterium leprae, lepra reactions can occur which manifests as acute or subacute episodes of inflammation. Lepra reactions can occur in 30-50% of cases with leprosy during or after treatment with antileprosy drugs[3]. Reactions in leprosy includes type-1 and type-2 reactions. Type 1 lepra reaction is type IV hypersensitivity reaction characterised by exacerbation of preexisting skin lesions which becomes edematous and erythematous or as fresh lesions. Type-2 lepra reaction or Erythema Nodosum leprosum is a type III hypersensitivity reaction which manifests as crops of new erythematous tender nodules and plaques and can be associated with fever, malaise, arthralgia and/or peripheral oedema[4]. Sweet's syndrome is febrile neutrophilic dermatosis which was first described by Robert Douglas sweet[5]. This condition presents as painful erythematous plaques, oedematous painful plaques with fever and leucocytosis and histologically characterised by dense dermal

infiltrates of neutrophils. Lepra reactions are common in lepromatous leprosy or borderline lepromatous (BL) during or after treatment, however they can rarely occur before treatment in patients experiencing stress, hormonal changes, coexisting infections or treatment with certain broad spectrum antibiotics like ofloxacin, rifampicin, etc[6]. But, few patients can present with lepra reactions as the initial manifestation de novo, however occurence of Sweet-like Erythema Nodosum Leprosum (ENL) with few bullae is extremely rare with only few cases reported in literature. Vesicles or bullae can develop in severe ENL but ENL as the first clinical manifestation in previously undiagnosed cases of leprosy is unusual[7]. When presenting as the first manifestation of leprosy, ENL often becomes as a diagnostic challenge because it mimics other disorders. The diagnostic difficulty in such a clinical setting may further be enhanced by atypical manifestations which includes pustular, ulcerated, haemorrhagic, frankly necrotic, vesicular, bullous Sweet syndrome-like and erythema multiforme-like lesions. Till 2021, only 16 cases of Sweet-like ENL have been reported in literature. Sweet like ENL is an uncommon form of clinical presentation in leprosy which has been described by Kuo and Chan in 1987[8]. Barreto and Freitas suggest that lepra reaction is a differential in manifestations of polymorphic erythema type when they relate a case with initial diagnosis of Sweet's syndrome. The Sweet's syndrome as leprosy reaction may be classified as a subtype of the type 2 reaction state[9]. It seems to be more common in patients with borderline lepromatous leprosy due to the immunologic instability. These reactions should be diagnosed promptly as they can damage the nerves and lead to disabilities. Hence, early diagnosis and treatment can help in preventing these disabilities.

CONCLUSION:

Though leprosy is characterised by hypopigmented anaesthetic patches, certain unusual and rare presentations do occur leading to diagnostic dilemma and delay in treatment. Early cases of ENL without any systemic manifestations can lead to diagnostic challenge both clinically and histologically. Hence, any atypical presentation which does not properly fit into a diagnosis warrants a biopsy especially with subcutis for exact categorisation of the lesion. This can lead to correct diagnosis and initiation of appropriate treatment, thereby preventing disabilities and morbidity.

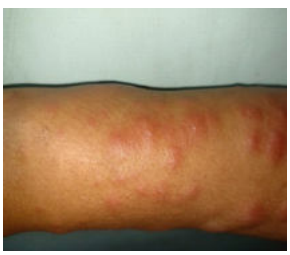

Fig 1: Erythematous nodule and plaques over the limb

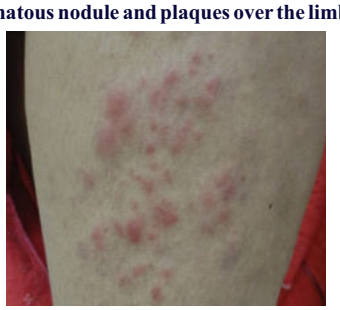

Fig 2: Erythematous nodule and plaques over the limb

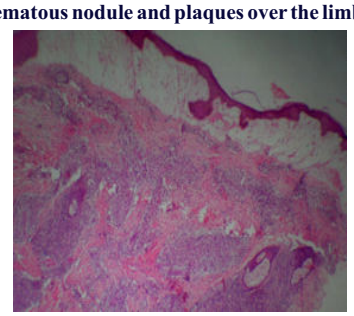

Fig 3: Skin biopsy showing marked dermal edema with formation of subepidermal vesicle

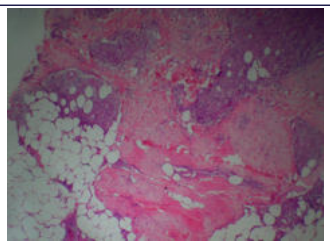

Fig 4: Biopsy showing multiple granulomas in dermis and subcutis

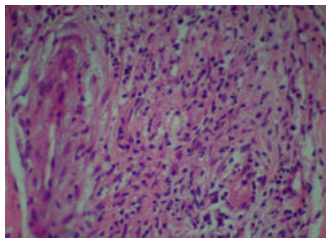

Fig 5: Biopsy showing loose granuloma admixed with foamy macrophages

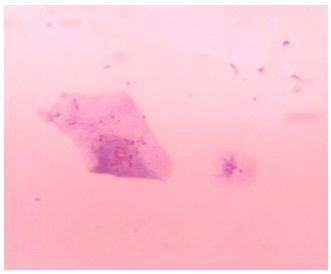

Fig 6: Slit skin smear showing numerous acid fast lepra bacilli

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