

A Rare Case Report of Leprous Macrocheilia in Borderline Tuberculoid Leprosy with Type 1 Reaction

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It is well known that *Mycobacterium leprae* tends to target the cooler parts of the body and can involve the oral cavity. Despite this, macrocheilia - a condition where the lips become enlarged - caused by leprosy is rarely documented. There are few reported cases of leprosy macrocheilia in India. We present a unique case of an elderly woman with borderline tuberculoid leprosy in type I reaction who developed leprosy macrocheilia.

Keywords : Leprous Macrocheilia, Borderline Leprosy, Type 1 Reaction

Introduction

Leprosy is a chronic, infectious disease caused by acid-fast bacilli – a mycobacterium, *Mycobacterium leprae*. Skin involvement and peripheral nerve damage are the main features, but being a multisystem disease, some degree of infiltration is seen in many other organs including the liver, kidney, gonads, lymph nodes, joints, eyes, nasal and oral mucosa (Singh et al 1999). Oral cavity involvement in leprosy is seen mainly in multibacillary cases and the hard palate is the most common site involved (Pallagatti et al 2012). Leprosy may rarely present as chronic macrocheilia which is defined as abnormal swelling of one or both the lips for 8 weeks or more (Burton et al 1998). We hereby report a

rare case of leprosy macrocheilia in a patient of borderline tuberculoid leprosy with type 1 reaction.

Case:

A 70-year female, a resident of Sonipat, presented to our Dermatology OPD with a complaint of swelling of the lower lip and a raised red lesion in her chin from the last 2.5 years. A major increase in size occurred in the last 2.5 months accompanied by mild pain over the lesions. She also had intermittent fever and myalgia for the last two months.

Other than a few topical medications from a local practitioner there was no definite drug history that could be linked to the development of the lesions. There was no other skin disease, known

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allergy, or history of insect bites. A family history of a similar illness was absent. There was no history of cough, haemoptysis, breathlessness, diarrhoea or swelling in the neck.

On examination, the patient was of regular build and height. Cutaneous examination revealed diffuse, hard and tender swelling of the lower lip. There was also a well-defined erythematous infiltrated plaque measuring 8 cm *4 cm extending from the lower lip up to the angle of the mouth on the right side of the face with an area of central sparing (Fig. 1a and 1b). Now, we made a differential diagnosis consisting of borderline tuberculoid Hansen's disease with type 1 reaction (BT-HD), sarcoidosis, granuloma annulare, Jessner lymphocytic infiltrate and

proceeded with a slit skin smear followed by punch biopsy from the lesion. There was no sensory loss over the plaque along with no nerve thickening. A slit skin smear was performed on the lesion to assess the bacillary index (BIS) and it came out as positive for acid-fast lepra bacilli (Fig. 2). Skin biopsy revealed loose (oedematous) epithelioid cell granulomas, with only occasional Langhans type of giant cells with a bacillary index of 3 plus consistent with a diagnosis of BT-HD with type 1 reaction (Fig. 3a and 3b).

As per WHO guidelines for diagnosing leprosy, one of the three cardinal features, i.e., presence of acid-fast bacilli on slit skin smear was positive in our patient. Taking into consideration the histopathology findings along with, we confirmed



Fig. 1a and 1b : Front and right lateral view of face showing swelling of the lower lip along with, a well- defined erythematous infiltrated plaque measuring 8 cm * 4 cm extending from the lower lip up to the angle of the mouth on the right side of the face with an area of central sparing.

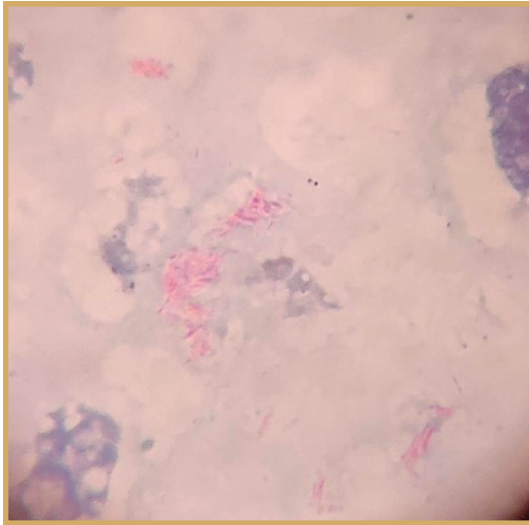


Fig 2 : Slit skin smear showing acid fast bacilli (pink) in a bluish background.

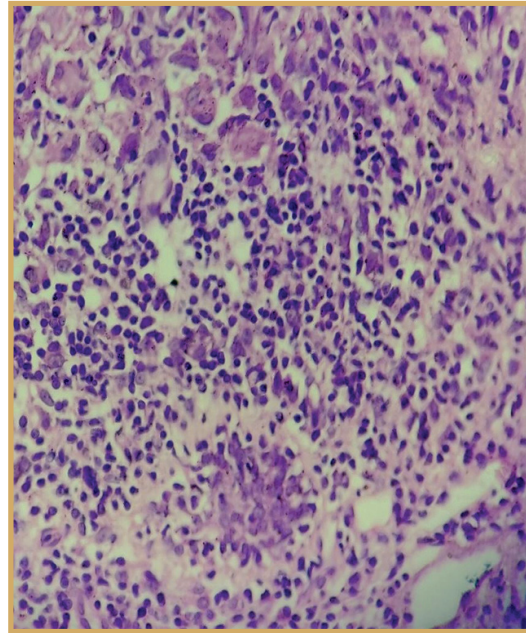


Fig. 3b : Loose (oedematous) epithelioid cell granulomas, with only occasional Langhans type of giant cells [H and E 40x].

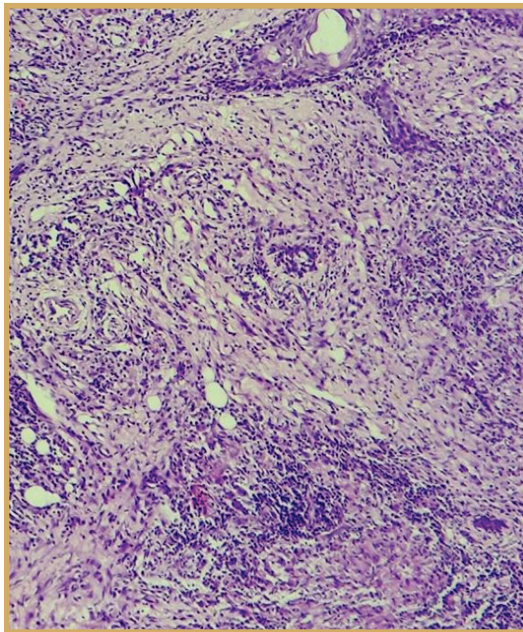


Fig. 3a : Loose (oedematous) epithelioid cell granulomas, with only occasional Langhans type of giant cells [H and E 10x].

our diagnosis as leprous macocheilia and after baseline investigations, started the patient on WHO Multibacillary Multidrug therapy (MB-MDT) along with 40 mg oral prednisolone. After 3 weeks of drug therapy, a significant decrease in size and erythema of the lesions was observed. However, as the granulomas cause lymphatic blockage lip lesions may persist even after eliminating the underlying disease, the MDT was continued for one year.

Discussion

The lepra bacilli have an affinity towards cooler areas of the body like nasal and oral mucosa and peripheral nerves. This was confirmed in a study by Hastings et al (1968) which showed the bacillary index to be higher in the skin areas with

a core temperature of 32.5 degrees celsius than that with a temperature of 34.6 degree celsius. This also explains why the hard palate is the most frequent site of involvement in the oral cavity (Scheepers 1998).

Depending upon the clinical profile of a particular group of leprosy patients, 19-60% of leprosy patients have been reported to have oral lesions (Scheepers 1998). These lesions can be non-specific enanthem of the palate, or uvula or specific lesions like papules plaque, nodules, and ulcers which show bacillary positivity. However, none are pathognomonic of leprosy (Reichert 1976). Leprosy involves the lips most commonly in the form of macrocheilia or by the presence of flat-topped nodules and microstomia. Leprous macrocheilia present as swollen and rigid lips which are cosmetically disfiguring leading to social embarrassment (Prabhu & Daftary 1981). Scheepers (1998) reported that the severity of oral lesions is higher in patients with a positive family history of leprosy and the number of lesions increases with age. The overall prevalence of lesions is higher among males although females present earlier probably due to greater cosmetic concern (Reichert 1976). Handa et al (2003) reported leprosy macrocheilia in 10.7% out of a total of 28 patients of chronic macrocheilia thereby making leprosy the third most common cause after cheilitis granulomatosa and tuberculosis.

Leprous macrocheilia is an important differential of orofacial granulomatosis (OFG) as well as cheilitis granulomatosa (CG). OFG is a concept first described in 1985 by Wisenfeld to classify all diseases causing granulomas in the orofacial region without any systemic features

(Wiesenfeld et al 1985). Both OFG and chronic macrocheilia due to leprosy may present with lip swelling, erythematous facial plaque and non-caseating granuloma on histology. But a slit skin smear or special stain like Fite Faraco will demonstrate the bacteria, i.e. *Mycobacterium leprae* from the skin lesion. Hence, in such cases, the clinician should consider leprosy as one of the important differentials that may be missed. The other conditions included in OFG are tuberculosis, sarcoidosis, Crohn's disease, cheilitis granulomatosa, and Melkerson-Rosenthal syndrome (MRS).

Cheilitis granulomatosa (CG) was first described by Miescher in 1945 as an inflammatory disease of unknown origin clinically presenting as persistent upper lip swelling. CG, if presents along with plicated tongue and facial palsy forms the Melkerson-rosenthal syndrome (Allen et al 1990). Chronic macrocheilia due to leprosy is a close differential of MRS as both may have CG, facial palsy clinically and granulomas in histology. However, in MRS, the granulomas are perivascular; in leprosy, they are more elongated and oriented around the nerve bundles (Pavithran 1987). Lip and perioral lesions have also been reported in the form of muzzle area involvement, where the patient presented with skin-coloured raised lesions that resemble post-kala-azar dermal leishmaniasis or sarcoidosis. However, skin biopsy was in favour of lepromatous leprosy (Reganti et al 2022).

Conclusion

In summary, it is rare to find borderline tuberculoid leprosy with type 1 reaction that presents as chronic macrocheilia. While leprosy macrocheilia in the tuberculoid pole

has been reported, our case is perhaps the first report to present macrocheilia in a patient with type 1 reaction. In India, a country with high leprosy prevalence, medical professionals must remain vigilant and consider leprosy as a potential diagnosis for such cases. Additionally, examining the oral cavity should be a routine part of assessment of leprosy patients, and both residents and practicing physicians/specialists should keep this in mind.

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