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CASE REPORT

Borderline leprosy masquerading as cheilitis granulomatosa: A case report

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Introduction

Chronic macrocheilia, which can be arbitrarily defined as persistent enlargement of one or both lips for more than 8 weeks, has varied aetiology, but granulomatous conditions, both infective and non-infective, comprise a significant proportion of these patients.[1] Cheilitis granulomatosa (CG) is probably the most common diagnosis entertained in such cases. Another diagnosis that is commonly entertained in these groups of patients is orofacial granulomatosis (OFG). Infective granulomatous conditions such as tuberculosis and leishmaniasis can rarely present with swelling of one or both lips.[2,3] Leprosy, a chronic infectious disease caused by an acid-fast bacillus, *Mycobacterium leprae*, with a peculiar tropism to the skin, peripheral nerves and certain mucous membranes may occasionally present with chronic macrocheilia.[4] Here, we report an unusual presentation of borderline leprosy with abnormal lip swelling.

Case Report

A 53 year old farmer presented to us complaining of asymptomatic swelling of the upper lip of 4 months' duration. Swelling had started spontaneously and progressed insidiously to reach the present size. He denied history of preceding or associated pain or itching over the

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swelling. Neither was there a history of pre-existing skin problem nor having taken any treatment for skin ailments in the past. Cutaneous examination revealed diffuse, soft non-tender enlargement of the upper lip. In addition, he also had a well-defined erythematous, infiltrated plaque (measuring 1·5 cm × 1·5 cm) at the angle of mouth on the left side (Figure 1).

A similar but smaller plaque (measuring 1 cm × 0·5 cm) was seen near the angle of mouth on the right side. There was no sensory deficit over the skin lesions; neurological examination did not reveal nerve thickening. Examination of the oral mucosae did not reveal any other abnormal finding. A differential diagnosis of granulomatous cheilitis, cutaneous tuberculosis or leprosy was considered and he was thoroughly investigated. His hematological and biochemical parameters were normal. Slit skin smear (SSS) examination (from the lesion, both ear lobules and forehead) and Mantoux test (with 1 tuberculin unit) were negative. Chest X-ray and computerised tomography (CT) imaging of the thorax were normal. Skin punch biopsy was done both from the lip and erythematous plaque. Biopsy section showed hyperkeratotic and acanthotic epidermis. Dermis showed numerous non-caseating, well defined granulomas composed of epitheloid cells, Langhans and foreign body type of giant cells admixed with dense inflammatory infiltrate of lymphocytes (Figure 2).

The granulomas were oriented mainly around the nerve bundles and appendageal structures. Special stain for *Mycobacterium leprae* (Fite-Faraco stain) revealed occasional acid-fast bacilli after careful search (Figure 3).

A final diagnosis of borderline tuberculoid leprosy was made. He was commenced on WHO multibacillary-multi drug therapy (MB-MDT) in view of the atypical presentation. On account of the lesions on the face and erythematous nature of the lesions, he was concomitantly started on oral prednisolone (30 mg/day) which was tapered in 3 months.

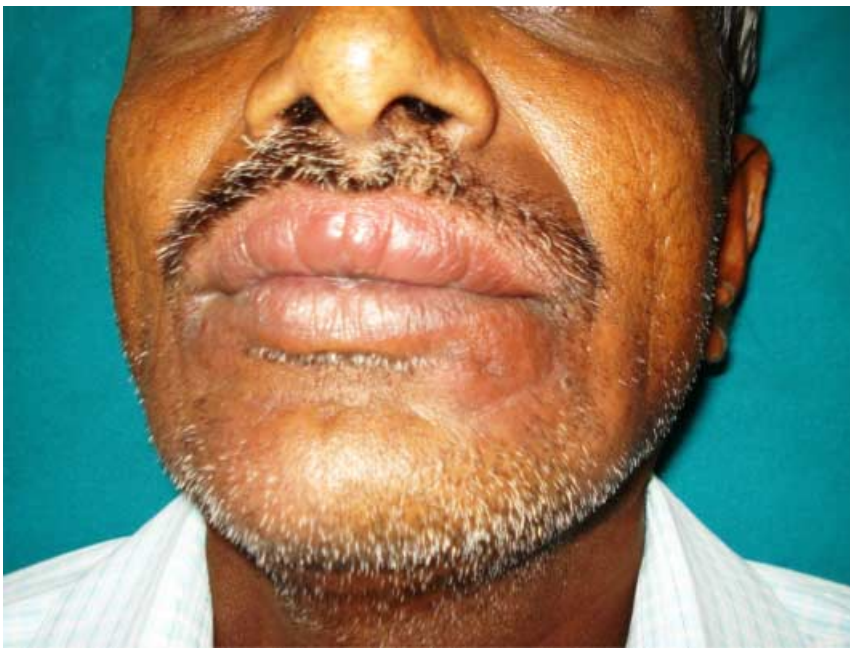

Figure 1. Upper lip edema and erythematous, infiltrated plaque on the chin on left side.

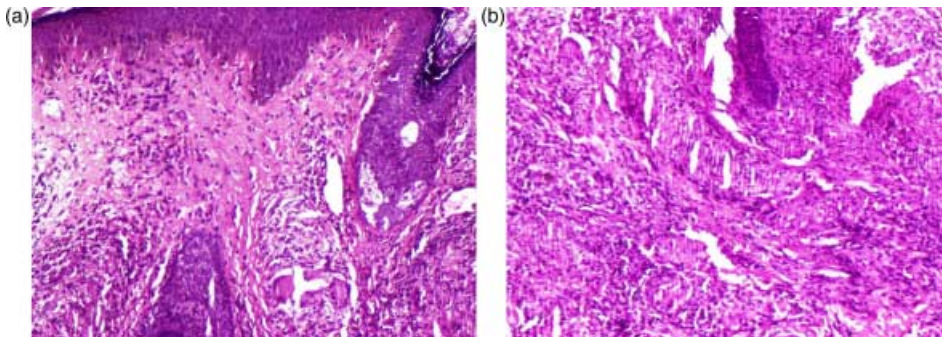

Figure 2. a) Tuberculoid granuloma in the dermis; b) Granulomatous infiltration around the dermal nerves (Haematoxylin & eosin, ×100).

At 6 months follow-up, erythematous plaques at the angle of mouth subsided and lip swelling subsided considerably. MDT was continued for 1year.

Discussion

Persistent enlargement of one or both lips is cosmetically disfiguring and socially embarrassing to the patients. It also impairs important activities such as eating and speaking. There are several conditions that can cause this disfiguring condition; however, in most, there are very few distinguishing local signs and symptoms, so they pose a great diagnostic as well as therapeutic challenge to the physician.

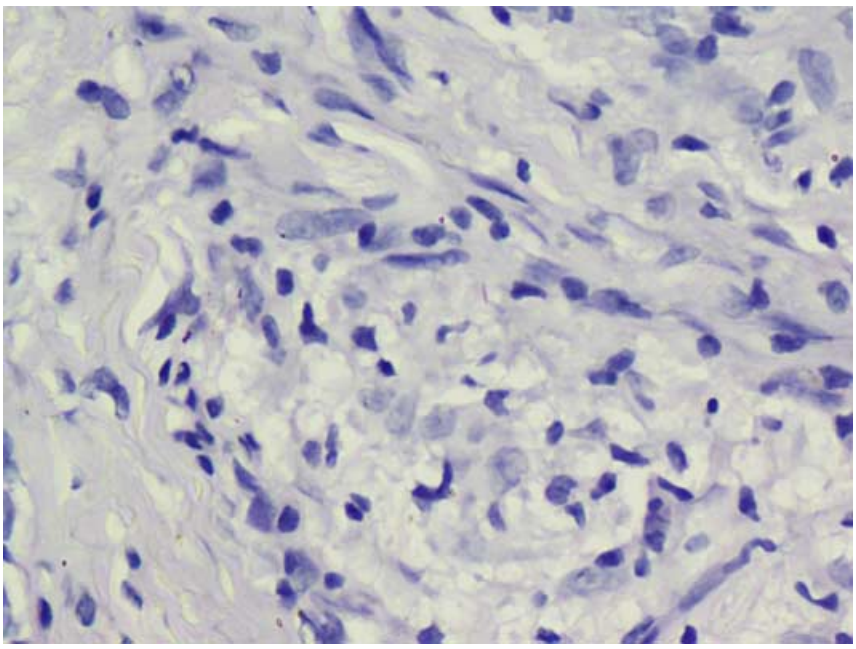

Figure 3. Sparse acid fast bacillus in the tissue section (Fite Faraco, ×400).

Oral involvement is not uncommon in leprosy. It has been reported to occur in about 20–60% of patients with lepromatous leprosy though it is quite rare in tuberculoid and borderline spectrum.[5,6] Usually it follows systemic disease, but occasionally it may present concomitantly or as a presenting manifestation of the disease. Tongue, uvula, anterior hard palate and lips are most frequently affected sites.[7] Leprosy presenting as chronic lip swelling is rare with only few case reports have been described in the literature. In a review of 28 patients with chronic macrocheilia, Handa *et al.* found leprosy as the third most important cause of macrocheilia after CG and tuberculosis. All three of their patients were diagnosed as suffering from borderline leprosy with or without Type I lepra reactions.[1]

CG is an inflammatory disease of unknown origin that was described by Meischer in 1945 and is characterised clinically by persistent upper lip swelling. CG is also an important component of Melkerson-Rosenthal syndrome (MRS) which has plicated tongue and facial palsy as the other components of the triad.[8]

The concept of OrofacialGranulamotosis was introduced by Wiesenfeld *et al.* to describe the occurrence of granulomas in the orofacial region in the absence of any recognised systemic condition.[9] There is no consensus on whether OFG is a distinct disease or initial presentation of Crohn's disease. Typically, OFG presents as recurring labial swellings that persist, resulting in enlargement of the one or both lips. OFG encompasses the previously recognised MRS and CG.[10,11]

Chronic macrocheilia due to leprosy may be confused with MRS especially if the patient has only CG which is an oligosymptomatic form of MRS. Facial nerve palsy may develop in both conditions. But the characteristic fissured tongue if present will aid in the diagnosis of MRS. Histologically, both conditions are characterised by the presence of noncaseating granuloma. But granulomas of MRS differ from others by their strong tendency to a perivascular arrangement. In contrast, tuberculoid granuloma of leprosy is seen in the upper dermis and appears to be elongated since they are oriented around the nerves.[12]

OFG is the other condition that may be confused with leprosy as both can present with labial swelling; erythematous plaques in the adjacent areas and noncaseating granulomas can be seen in both conditions. Only distinguishing feature is the demonstration of bacteria in the granulomas.[7] On special staining, occasional bacilli may be seen in the leprous granulomas. Therefore, a careful search must be made in every case to look for 'bacillary needles in tissue haystacks' since they are often sparse and easily missed. Granulomas appear to cause lymphatic blockage, leading to diffuse swellings of the lips and other sites. Therefore lip swelling may persist even after the successful treatment of underlying pathology.

To conclude, chronic lip swelling may be an atypical and rare presentation of paucibacillary leprosy. In an endemic country, leprosy should always be considered in the differential diagnosis of chronic macrocheilia. Ultimately the diagnosis depends upon correlation of historical and clinical data with histopathological findings.

References

[1] Handa S, Saraswat A, Radotra BD, Kumar B. Chronic macrocheilia: a clinico-pathological study of 28 patients. *Clin Exp Dermatol*, 2003; **28**: 245–250.

[2] Favia G, Chiaravalle G, Lacaita MG *et al.* Orofacial tuberculosis. A general and anatamico-clinical analysis of 35 cases. *Minerva Stomatol*, 1990; **39**: 261–268.

[3] Vasudevan B, Bahal A. Leishmaniasis of the lip diagnosed by lymph node aspiration and treated with a combination of oral ketoconazole and intralesional sodium stibogluconate. *Indian J Dermatol*, 2011; **56**: 214–216.

[4] Singh G, Nagaraja Prabhu S. Leprosy presenting as macrocheilia. *Indian J Lepr*, 1999; **71**: 341–342.
[5] Ghosh S, Gadda RB, Vengal M *et al.* Oro-facial aspects of leprosy: report of two cases with literature review. *Med Oral Patol Oral Cir Bucal*, 2010; **15**: e459–e462.
[6] Mishra B, Mukherjee A, Girdhar A *et al.* Involvement of lips and gums in a borderline tuberculoid leprosy patient. *Indian J Lepr*, 1996; **68**: 367–370.
[7] High AS, Lansley CV. Labial and gingival enlargement in leprosy. *Br Dent J*, 1988; **165**: 371–372.
[8] Allen CM, Camisa C, Hamzeh S, Stephens L. Cheilitis granulomatosa: report of six cases and review of the literature. *J Am Acad Dermatol*, 1990; **23**: 444–450.
[9] Wiesenfeld D, Ferguson MM, Mitchell DN *et al.* Oro-facial granulomatosis: a clinical and pathological analysis. *Q J Med*, 1985; **54**: 101–113.
[10] Grave B, M McCullough M, Wiesenfeld D. Orofacial granulomatosis – a 20-year review. *Oral Dis*, 2009; **15**: 46–51.
[11] Leão JC, Hodgson T, Scully C, Porter S. Orofacial granulomatosis. *Aliment Pharmacol Ther*, 2004; **20**: 1019–1027.
[12] Pavithran K. Melkersson-Rosenthal syndrome masquerading as tuberculoid leprosy. *Indian J Lepr*, 1987; **59**: 26–29.