

## Leprous macrocheilitis: A rare variant of oral granulomatosis

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### Introduction

Borderline tuberculoid leprosy most commonly presents with multiple asymmetric plaques over the body with enlarged peripheral nerves. Nerve damage is a common presentation, and anesthesia and motor deficit may be observed at the time of presentation.

Lesions of the oral cavity are more common in lepromatous patients, the sites being the tongue, pharynx, hard and soft palate, tonsillar pillars, and the uvula<sup>(1)</sup>.

Involvement of lips in leprosy patients is an extremely rare entity and can be mistaken for a variety of other granulomatous conditions.

### Case report

A 25-year-old man presented to our clinic with asymptomatic swelling over his lips for the past seven years. It had a waxing and waning course. The patient gave a history of taking multiple treatments such as oral tapered steroids, antibiotics, clofazimine, multidrug therapy (rifampicin, clofazimine, dapsone), ofloxacin, minocycline, methotrexate, apremilast, thalidomide, hydroxychloroquine, and colchicine.

The patient did not give any history suggestive of skin lesions or loss of sensation. No history of trauma or insect bite. No history of chronic lip biting. There is no history of comorbidities. There was no history of systemic involvement suggesting sarcoidosis or Crohns disease. Family history was unremarkable.

On examination, there was non-tender diffuse swelling of both upper and lower lips with erythema over cheeks.

Examination of the oral mucosa did not reveal any abnormal findings. There was no evidence of fissured tongue. There were no lesions elsewhere on the body.

The figure 1 shows swelling over lips with erythema



**Fig.1 Swelling over lips with erythema over cheeks**

over cheeks (pre and post-treatment over 6 months). The patient was evaluated further, and investigations like tuberculin test, X-ray Para nasal sinuses, X-ray chest, angiotensin-converting enzyme levels were done, all of which were within normal limits.

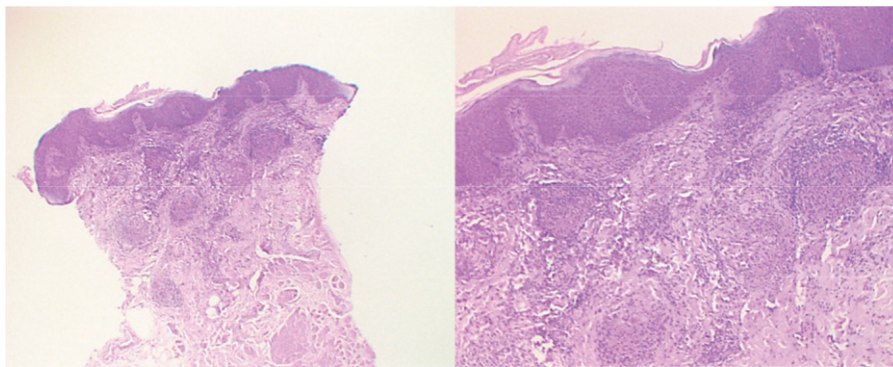
Nerve conduction tests of the left median, ulnar and sural nerve were carried out, which were normal. No abnormality was detected in the ultrasonography of the abdomen, pelvis, and scrotum. Hence, all common conditions causing orofacial granulomatosis were ruled out.

A punch biopsy of the lip was taken, which showed an unremarkable epidermis. The dermis showed papillary edema and multiple focal collections of epithelioid cells and lymphocytes around the adnexal structures and

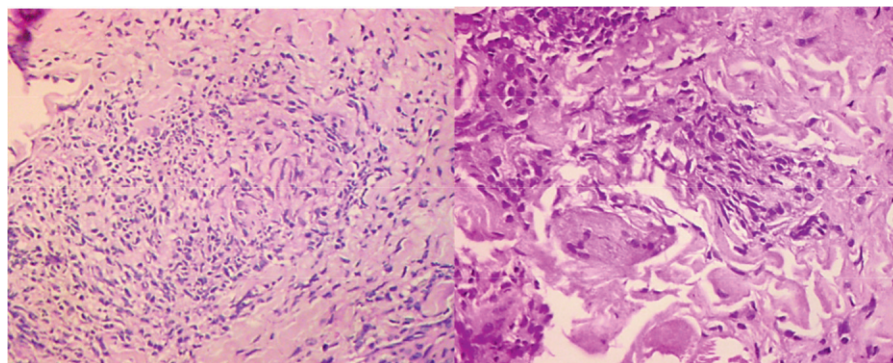
blood vessels. Such granulomas were separated from the epidermis by a thin rim of collagen. The nerve structures included in the sections also showed a similar infiltrate. Multinucleated giant cells were not evident. There was perivascular lymphocytic infiltrate and dispersion of granulomas at places. Special stains for acid and alcohol fast bacilli were negative and based on the biopsy, the patient was diagnosed as a case of borderline tuberculoid Hansens disease.

are cutaneous sarcoidosis, Rosenthal Melkerson syndrome, Cheilitis Granulomatosa. Table 1<sup>(2)</sup> shows the most common causes of granulomatous cheilitis.

Oral leprosy is a rare phenomenon and is a late presentation. It is commoner in the lepromatous pole of the spectrum than the tuberculoid pole and generally follows systemic involvement. The prevalence of oral lesions was found to be higher in males as compared to females<sup>(3)</sup>. Of the oral cavity, the chiefly affected sites are the hard palate<sup>(4)</sup>; the tongue, lips, and uvula being



**Figure 2: Granulomas in dermis (H & E at 4x and 10x magnification)**



**Figure 3: Granulomas in dermis (H & E at 40x magnification)**

Since the patient showed no improvement after being initiated on multidrug therapy for one year, he was shifted to second line drugs like ofloxacin, minocycline and clarithromycin with a tapered dose of prednisolone for 4 months after which he showed improvement.

### Discussion

The concept of orofacial granulomatosis was introduced by Weisenfeld in 1985, which classified diseases causing granulomas of the orofacial region without any systemic features<sup>(1)</sup>. A few other conditions which may also present as persistent swelling of the lips

rare sites of affection. Following features are seen in corresponding sites: Table 2<sup>(5)</sup>.

We found the following cases published to date of granulomatous cheilitis, including leprous macrocheilitis. Other cases with leprous macrocheilitis:

1. Handa et al 2003<sup>(6)</sup> – There were nine males and four females with few patients with both lips and few with single lip involvement. The mean age of patients was 28 years, with one patient having facial palsy, three patients with lingua plicata. Three out of these patients had leprous macrocheilitis.
2. Chintagunta et al 2017<sup>(7)</sup> There were two males and

three females, some with involvement of both lips, few with single lip involvement, out of which one patient was suspected of having oral leprosy.

3. Gogri et al 2015<sup>(8)</sup> – Reported a case of a male with involvement of both lips, diagnosed as Hansen's, started on multidrug therapy.
4. Gupta et al 2019<sup>(9)</sup> – Studied 200 patients with orofacial Hansen's. Out of these patients, 18% had macrocheilitis.

Cases showing other causes of macrocheilitis:

1. Dhar and Kanwar 1995<sup>(10)</sup> – Report three males and three females with Rosenthal Melkerson syndrome, with few patients with involvement of both lips and few with single lip involvement. The approximate age was around 1-20 years, with three patients having facial palsy, one patient with lingua plicata, one with MRS. Treatment consisted of injection triamcinolone acetonide and tablet clofazimine.
2. Gupta and Singh 2014<sup>(11)</sup> – Report one female aged 58 years with granulomatous cheilitis of the upper lip. She was treated with tablet metronidazole 400mg thrice daily and tablet minocycline 100mg once daily.
3. Savithri et al 2017<sup>(12)</sup> – report three females diagnosed with orofacial granulomatosis and tuberculosis.

4. Tambe et al 2018<sup>(13)</sup> report two females diagnosed with granulomatous cheilitis, one with involvement of both lips, and one with upper lip involvement. The patients were treated with tablet metronidazole 400mg thrice daily.
5. Sharma et al 2020<sup>(14)</sup> report two females with involvement of both lips and single lip diagnosed as granulomatous cheilitis who were treated with injection triamcinolone and tab clofazimine 100 mg twice daily.

### Conclusion

Leprous macrocheilitis in itself is a rare entity. It should be considered a differential diagnosis in a case of chronic macrocheilia, especially in an endemic country like India, even in the absence of the common cutaneous findings seen in cases of leprosy. Chronic lip swelling can be a cause of cosmetic and social concern for patients. Since the patients presenting with chronic macrocheilia as the sole complaint with no systemic or other cutaneous findings, it may pose a diagnostic and therapeutic challenge to the physician.

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**Conflict of interest:** Nil

**Table 1: Common causes of granulomatous cheilitis**

Etiology	Examples
Infectious	Tuberculosis, leprosy, histoplasmosis
Granulomatous causes	Sarcoidosis and foreign body reaction
Inflammatory conditions	Rosacea
Malignancies	Salivary gland tumours
Drugs	Angiotensin converting enzyme inhibitors and calcium channel blockers
Atopic reaction	Due to allergens
Hereditary diseases	Angioedema secondary to C1 esterase deficiency and Aschers syndrome
Metabolic diseases	Amyloidosis

**Table 2 Clinical features seen in corresponding sites**

Site	Clinical features
Tongue	Superficial ulcers, mild glossitis, low of papillae, fissured tongue
Lips	Macrocheilia, flat topped nodules, microstomia
Buccal mucosa	Diffuse infiltration, papulonodules and ulceration
Hard and soft palate	Erythematous papules, nodular sub mucosal infiltrate, palatal ulceration and perforation
Uvula	Miliary papules and nodules
Dental	Gingivitis, periodontitis

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