

Leptous macrocheilia: A rare clinical presentation

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Abstract

Oral leprosy, a granulomatous disease is classified under the term oro-facial granulomatosis which comprises a group of diseases characterized by noncaseating granulomas affecting the soft tissues of the oral and maxillofacial region. The most common clinical presentation of oro-facial granulomatous conditions is persistent swelling of one or both lips. Due to its rare incidence and clinical findings overlapping with other granulomatous conditions such as sarcoidosis, Crohn's disease, and cheilitis granulomatosa; it is practically difficult for a dental surgeon to easily diagnose this condition. This study presents a case of leprosy causing macrocheilia as the only clinical presentation and diagnosed initially as Miescher's cheilitis based on exclusion criteria.

Keywords: Cheilitis granulomatosa, macrocheilia, Miescher's cheilitis, oral leprosy, oro-facial granulomatosis

Introduction

Chronic macrocheilia is swelling of one or both lips for more than 6 to 8 weeks and can be due to varied etiology.^[1] Oro-facial granulomatosis due to infective and noninfective reasons is generally the condition associated with chronic macrocheilia and the most common considered diagnosis in such cases by dental specialists is generally cheilitis granulomatosa.

Leprosy is a specific granulomatous condition caused by mycobacterium leprae that has a high predilection for skin and peripheral nerves. Occasionally, it may present with chronic macrocheilia and can be misdiagnosed as Miescher's cheilitis.^[2,3]

The aim of this case report of leptous macrocheilia is to discuss the clinical features and differential diagnosis of various granulomatous conditions which will help general dental practitioners to avoid the misdiagnosis.^[2,3]

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Case Report

A male patient aged 17 years had reported to the Department of OMDR of our institution with a chief complaint of painless swelling of both upper and lower lips since 7 months. The patient gave a history of fever 7 months ago after which the swelling had developed. The data about exact cause and nature of fever could not be retrieved from patient. Initially, the swelling was smaller in size and was constant for few months. Gradually, the swelling increased in last 2–3 months to present size.

The patient was of average built and height. His medical history was not contributory as there was no apparent history of trauma, allergy to any substance, insect bite, pain, pus discharge, etc., which may be related to the lip swelling. He gave no present or past history of lip biting and his family history was also unremarkable.

Extraoral examination showed diffuse swelling of both upper and lower lip [Figure 1]. The swelling was nontender to palpation. The upper lip showed an area of hypopigmentation but no signs of paresthesia or altered sensations were seen. No lymphadenopathy was noted. Lips were competent.

On intraoral examination, labial mucosa of both upper and lower lips showed granular appearance. The granularity on the mucosal surfaces could be felt by digital palpation. Anterior teeth in both arches showed mild inflammation and swelling of labial gingiva. [Figure 2]. Deposits on the right

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sided premolars and molars of both arches were suggestive of unilateral chewing habit [Figure 3a and b].

On the basis of clinical findings, the condition was identified as oro-facial granulomatosis and the following differential diagnosis were considered.

Cheilitis granulomatosa (of Miescher's) appears as nontender persistent swelling of one or both lips as seen in this case.

Melkersson-Rosenthal syndrome was ruled out as Bell's palsy and scrotal tongue were absent.

Crohn's disease shows lip swelling along with gastrointestinal symptoms such as abdominal pain or diarrhea, signs of malabsorption, anemia, and skin rashes, which were absent and so was omitted.

Whereas in Sarcoidosis multiple organs are involved causing symptoms such as weight loss, tiredness, arthritis, dry eyes, blurry vision and cutaneous rashes, and none were seen in this case.

After clinical examination, patients hemoglobin level, bleeding time, and clotting time were checked to perform biopsy procedure. As reports were normal, biopsy was performed

on lower labial mucosa and was sent for histopathological examination which showed multiple granulomas suggesting a chronic granulomatous inflammatory condition and diagnosis of cheilitis granulomatosa (Miescher's type) was considered [Figure 4a and b].

Treatment was started with systemic prednisolone 10 mg TID for 1 week and the patient was recalled after 7 days. On recall appointment, lesional area did not show any obvious clinical improvement, so medication was extended for more than 1 week duration and was supplemented with local steroid injections herewith. Intralesional injections of Kenacort were given in two visits (1 vial of 40 mg/ml at each appointment) in upper and lower labial mucosa at the interval of 3 days after which patient showed some significant improvement. Hence, the patient was recalled after 3 days and reviewed, and at the same time, dose of prednisolone was tapered to 10 mg BD for 3 days and 10 mg OD for more three days and finally stopped.

On next visit, intralesional injection of dexamethasone was given and immediately after 24 h, patient was reported to the hospital with an exacerbated reaction [Figure 5a and b]. The swelling on lips had recurred and was clinically more severe than it was when the patient first reported to the hospital. Erythematous patches had developed on bilateral



Figure 1: Extraoral view showing lip swelling more prominent with the upper lip. Note the hypopigmented patch in the center part of upper lip



Figure 2: Gingival edema of upper and lower teeth. Note the loss of stippling at some areas and change in shape of marginal gingiva of upper incisors

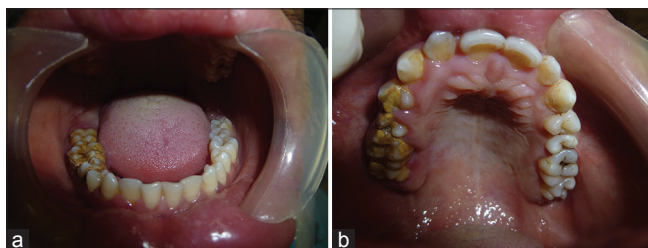


Figure 3: (a) Mandibular arch showing severe calculus formed on right-sided teeth until occlusal level. (b) Maxillary arch showing severe calculus formed on right-sided teeth until occlusal level

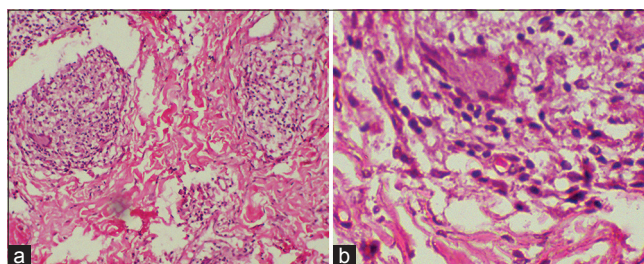


Figure 4: (a) Low power view is showing multiple granulomas in connective tissue. (b) High power view showing Langhans type of giant cell

cheek regions extraorally. The swelling was initially painless but patient now had the symptoms of pain in the lesional area. As the reason for such reaction was hard to conclude opinion of dermatologist was sort.

Dermatologist could observe the features such as scaling along with erythema in bilateral cheek region, nerve thickening bilaterally over the angle of mandible and hyperesthesia in chin area which were unfortunately missed by us because we are not familiar with such kind of lesions. On the basis of clinical findings, the dermatologist suspected the condition to be leprosy.

Skin biopsy of the patient was performed from extraoral cheek region from the site of erythematous patch which showed the evidence of foci of ill-defined granulomas with few macrophages in the dermis. Furthermore, dense, chronic inflammatory infiltrate composed predominantly of lymphocytes and plasma cells were seen around perivascular and periadnexal structures.

Diagnosis of borderline lepromatous (BL) leprosy with type 1 skin reaction was made and the patient was referred to Bombay Leprosy Programme Centre for slit skin smear examination [Figure 6]. The test appeared negative confirming the noninfectious nature of the lesion. The patient was started with Multibacillary-multidrug therapy (MB-MDT).

After 3 months follow up the lip swelling of the patient was reduced considerably [Figure 7]. MDT was continued for 1 year.

Discussion

Wiesenfeld introduced the concept of oro-facial granulomatosis in 1985 to classify diseases causing granulomas in the oro-facial region with the absence of any systemic manifestations.^[4] It includes a variety of clinical conditions such as tuberculosis, leprosy, sarcoidosis, Crohn's disease, and cheilitis granulomatosa and all are considered in the differential diagnosis as they show similar clinical presentation as persistent swelling of one or both lips. The clinical incidence of such cases is very less and the epidemiological data is also scarce with most reported data related to cheilitis granulomatosa.^[5,6]

Leprosy, a granulomatous disease is caused by mycobacterium leprae also called as Hansen's *bacillus* after the person who discovered the organism in 1874. The annual cases of leprosy detected in India is (53/100,000).^[7] Mycobacterium leprae is an acid fast *bacillus* similar to *Mycobacterium tuberculosis*. The organism affects almost any part of the body but has more predilection for skin and superficial nerves.

D. S. Ridley and William Jopling proposed their classification for leprosy in 1962 which includes:

- Tuberculoid (TT)
- Borderline tuberculoid
- Mid borderline
- BL
- Lepromatous (LL).

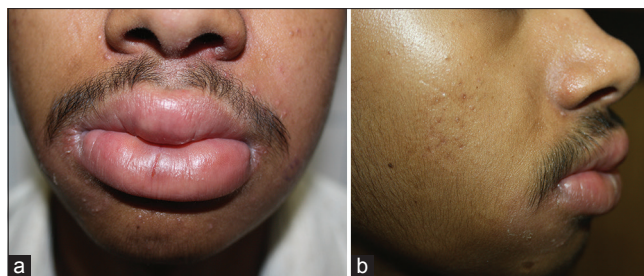


Figure 5: (a) Recurrent swelling of lips after intralesional steroids. (b) Erythematous patch on right side cheek. Note the scaling of skin near corner of the mouth



Figure 6: Slit skin smear examination



Figure 7: Reduction in the lip swelling after 3 months of start of treatment

However, in India leprosy is classified and simplified into the following types:

- TT
- Borderline
- LL
- Intermediate
- Neuritic.

Leprosy mostly affects the skin and peripheral nerves. It causes the gradual loss of sensations, especially apparent in the hands and feet; therefore, the chief complaint may be a burn or ulcer in an anesthetic extremity. It may affect eyes and earlobes and may cause multiple nodular eruptions on the face. Oral involvement is secondary and is seen in 20–60% cases of LL leprosy^[8] though is rare in TT leprosy.^[9] In general, oral lesions in leprosy are of great epidemiological significance as a source of infection^[10] and are late manifestations of the disease. Contrary to the fact in present case lip swelling was the only manifestation and patient gave no history of any past lesion elsewhere in the body.

In oral cavity, the most frequently affected site is hard palate^[11] and tongue, uvula, and lips are less frequently affected sites.^[12] Literature on leprosy macrocheilia, when reviewed, shows reports of isolated cases, but very few studies being done. Handa *et al.*^[13] in 2003 made extensive efforts to study 28 patients of this rare entity and reported leprosy to be the third most common cause of chronic macrocheilia after cheilitis granulomatosa and tuberculosis affecting 3 out of 28 patients studied.

Following features can be seen in different anatomical locations:

- Hard palate: As infiltration, reddish yellow nodules, ulceration, or perforation can be seen
- Uvula: Fibrosis with partial or complete destruction
- Lips: Macrocheilia due to infiltration or microstomia due to lip ulceration and subsequent fibrosis
- Gingiva: Lesions in contiguity with hard palate are seen, chronic gingivitis and periodontitis can occur
- Teeth: Odontodysplasia leprosa causing shortening of roots mostly maxillary anterior teeth. Chronic lepromatous infections can cause pinkish discolorations of crowns due to the granulomatous invasion of pulpal tissues.

Following are the salient features for diagnosis of leprosy:

- Enlargement and/or tenderness of a peripheral nerve
- Loss of sensation in plaque type skin lesions
- Finding of acid fast bacilli in smears.

Differential diagnosis of other granulomatous lesion must always be considered in cases with asymptomatic lip swellings.

MDT is the mainstay of leprosy treatment as it not only cures the patient, but also reduces the reservoir of infection thereby interrupting the spread of disease.

Two types of drug regimens are generally used for treating leprosy patients and depending on the severity patients are divided as:

- Paucibacillary leprosy - 2 drug regimen for 6 months: Rifampicin (600 mg) monthly supervised and dapsone (100 mg) daily unsupervised
- MB leprosy - 3 drug regimen for 1 year: Rifampicin (600 mg) with clofazimine (300 mg) monthly supervised and dapsone (100 mg) with clofazimine (50 mg) daily unsupervised.

Symptomatic and supportive measures such as adequate oral hygiene maintenance. Anesthetic mouthwashes or topical steroids for local control of pain and inflammation due to ulcers along with healthy diet and vitamin supplements are recommended.

Conclusion

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

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Conflicts of interest

The authors have obtained the necessary patient consent forms where the patients have given their approval for participation in the investigation, followed by representation in the concerned article. The patients do understand that the authors will ensure that their identities won't be revealed, however anonymity cannot be guaranteed.

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