

## Granulomatous cheilitis

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**ABSTRACT** A 19-year-old labourer presented with progressively worsening swelling of both lips. Clinical assessment and investigations led to the diagnosis of granulomatous cheilitis.

**KEYWORDS** Granulomatous cheilitis, orofacial granulomatosis, sarcoidosis

**DECLARATION OF INTERESTS** No conflicts of interest declared.

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### CASE REPORT

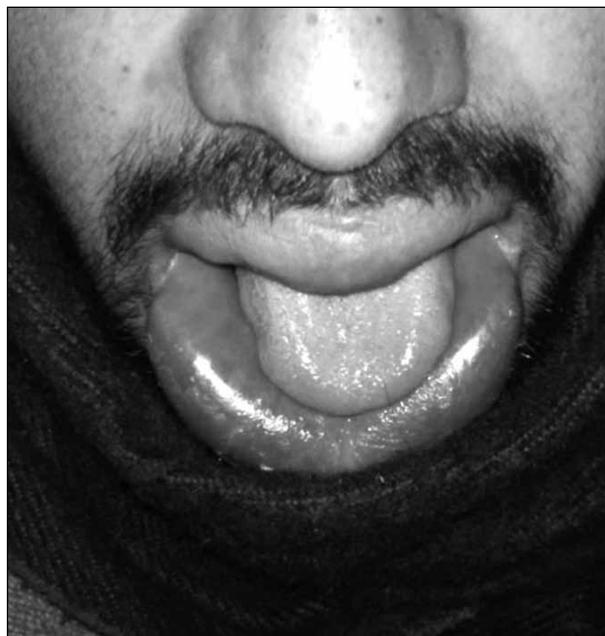
A 19-year-old man presented to the outpatient department of our hospital with progressive swelling of both lips over the preceding six years and worsening over the previous 12 months. There was no history of trauma, skin lesion, facial muscle weakness, febrile episodes, bowel disturbances or swelling in the neck. There was no significant past medical or surgical history. He was not on any medications and denied use of drugs, food or contact allergies. On examination, he had a strong physical appearance, with diffusely enlarged upper and lower lips. His lower lip was more swollen than the upper lip and his tongue was normal. Oral hygiene was good, with no evidence of dental infection. There were no palpable lymph nodes. Cranial nerves examination was normal. He was haemodynamically stable and a systemic examination was unremarkable.

### INVESTIGATIONS

Initial investigations showed a normal blood count, biochemical profile and C1 esterase level. Angiotensin-converting-enzyme (ACE) levels were raised, at 120 units/mL. The Mantoux tuberculin skin test was negative. An upper gastrointestinal endoscopy showed no abnormality. A chest X-ray and computed tomography (CT) scan of the chest did not show lymphadenopathy or any evidence of tuberculosis. A lower lip biopsy showed non-caseating granulomas.

### MANAGEMENT

The patient was referred to the dermatology department. He received weekly triamcinolone (10 mg) intralesional injections over the course of six weeks. He was advised to continue these injections for at least three months. However, due to his perceived lack of improvement, and pain induced by the injections, the patient declined any further intralesional injections.



**FIGURE 1** Swelling of upper and lower lip.

Subsequently he was given a course of oral steroids for the next two months (1 mg/kg). He did not show any improvement. The patient declined any further injections after receiving six steroid injections. Azathioprine (1 mg/kg/day) and sulfa salazine (500 mgm, every six hours) were added to the regimen. He persevered with oral medication for ten weeks with no significant improvement. The patient then requested a referral to the plastic surgery department. The patient was evaluated for surgery. The procedure was explained and he was asked to arrange the required payment. He did not attend for a follow-up appointment.

## DISCUSSION

Orofacial granulomatosis (OFG) is a rare condition of unknown aetiology used to describe swelling in the lips, face or areas within the mouth, in the absence of any recognised systemic conditions. The term was first introduced by Wiesenfeld in 1985.<sup>1</sup>

Granulomatous cheilitis (GC), first described by Miescher in 1945,<sup>2</sup> refers to persistent, painless swelling confined to one or both lips. It is regarded as a monosymptomatic manifestation of Melkersson-Rosenthal syndrome,<sup>3</sup> a triad of facial palsy, fissured tongue and GC.

The cause of OFG is unclear.<sup>4</sup> A variety of foodstuffs, including wheat and dairy products, chocolate, eggs, peanuts, cocoa, cinnamaldehyde, monosodium glutamate, cormosine, food additives, toothpaste, and dental material such as amalgam and mercury have been suggested in several case reports.<sup>4</sup> Some studies consider OFG to be the oral manifestation of Crohn's disease.<sup>5,6</sup> Differential diagnoses include acquired and hereditary angioedema leprosy, tuberculosis<sup>7</sup> and sarcoidosis.<sup>8</sup> The median age of presentation is 25–28 years old.<sup>9</sup>

The first episode of oedema subsides completely in hours or days. After recurrent attacks, which may range from days to years, swelling may persist and progressively increase, eventually becoming permanent.<sup>9</sup> Attacks are sometimes accompanied with fever and mild constitutional symptoms such as headache and visual disturbances. The main presenting complaint in a series of 119 patients was facial lip swelling.<sup>9</sup>

Depending on the patient's symptoms and a systemic review, evaluation includes a complete blood count, ACE levels, CI esterase level, tuberculosis skin tests and patch tests. Patch tests have identified sensitivities to various foodstuffs and additives, most commonly to benzoic acid.<sup>10,11</sup> Gastrointestinal endoscopy, gallium scan or positron emission tomography (PET) scanning may be performed. A panoramic scanning dental X-ray may be obtained to exclude chronic dental abscess.

Spontaneous remission is rare. Treatment has been challenging.<sup>12</sup> No standard or predictably successful modality has been established. Corticosteroids (local or oral) have been advocated<sup>13</sup> and improvement has been reported, however it is temporary. Antibiotics, sulphamethoxazole, tetracycline, isoniazid, minocycline, roxithromycin,<sup>14</sup> and metronidazole have all been tried. Immune modulators such as infliximab<sup>15</sup> and methotrexate have also been reported. Elimination diets have been recommended. Cinnamon- and benzoate-free diets have shown to be of benefit in a significant number of patients.<sup>16,17</sup> However a European series showed only a small benefit from cinnamon- and benzoate-free diets in those that patch tested positive for benzoic acid, cinnamaldehyde and cinnamyl alcohol.<sup>18</sup> Surgical treatment is an option for those patients who are severely affected or have impairment of function.<sup>19</sup>

## CONCLUSION

Chronic swelling of lips is a difficult problem to diagnose and manage. It is a socially embarrassing condition. Management depends on the underlying cause. There is no consensus on the best treatment option. A benzoate cinnamon-free diet has shown benefit in some studies.

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## Myre Sim Fund Research & Travel Grants

Dr Myre Sim (1915–2009) was a renowned and published psychiatrist who assisted many individuals and organisations through his long and successful career, in addition to making a major contribution to the medical community. He was brought up and educated in Edinburgh and graduated MBChB from Edinburgh University in 1938. Although he did not remain in Edinburgh to practice medicine, he retained a great fondness for the Edinburgh University Medical School and the Royal College of Physicians of Edinburgh. He became a Fellow of the Royal College of Physicians of Edinburgh in 1970.

During his lifetime Dr Myre Sim made a series of generous donations to the College. These are now administered by the Myre Sim Committee which meets twice a year, in May and November.

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