

Cheilitis granulomatosa: A case report

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ABSTRACT:

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Cheilitis granulomatosa (CG) is a rare inflammatory disorder of the lip. The cause of CG is unknown. When CG is accompanied by facial palsy and plicated tongue, it was referred to as Melkersson–Rosenthal syndrome. We reported a 34-year-old Thai female patient presented with painless lower lip swelling and fissured tongue for a few months. Confirmed by histopathologic study of the lower lip, cheilitis granulomatosa was diagnosed.

Key words: Cheilitis granulomatosa, Melkersson–Rosenthal syndrome

บทคัดย่อ:

มติ เชื้อมโนชาญ วีรณัฐ โพธิ์ทอง รายงานผู้ป่วยโรค CHEILITIS GRANULOMATOSA วารสารโรคผิวหนัง 2557; 30: 181-185.

ภาควิชาตจวิทยา คณะแพทยศาสตร์ศิริราชพยาบาล มหาวิทยาลัยมหิดล

Cheilitis granulomatosa เป็นโรคที่เกิดจากการอักเสบบริเวณริมฝีปากซึ่งพบได้ไม่บ่อย ในปัจจุบันยังไม่ทราบสาเหตุที่ชัดเจน ผู้ป่วย Cheilitis granulomatosa ที่พบการอักเสบของกล้ามเนื้อบริเวณใบหน้าและลิ้นยื่นรื้อจะทำให้การวินิจฉัยเป็น Melkersson–Rosenthal syndrome ผู้นิพนธ์ได้นำเสนอผู้ป่วยหญิงอายุ 34 ปี มาพบแพทย์ด้วยเรื่องริมฝีปากล่างบวมร่วมกับลิ้นยื่นรื้อมาเป็นระยะเวลา 2-3 เดือน ผู้ป่วยได้รับการตัดชิ้นเนื้อบริเวณริมฝีปากล่าง เพื่อขึ้นชันการวินิจฉัยโรค Cheilitis granulomatosa

คำสำคัญ: โรค Cheilitis granulomatosa, Melkersson-Rosenthal syndrome

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Introduction

Cheilitis granulomatosa (CG) is a rare idiopathic inflammatory disorder clinically characterized by diffuse painless enlargement of one or both lips.¹ The cause of CG is unknown.² Herein we reported a case of CG in 34-year-old female presenting with painless lower lip swelling. She responded well with monthly intralesional triamcinolone.

Case report

A 34-year-old Thai female presented with painless lower lip swelling for a few months which attacked her periodically. Her lips never completely resolved. She could eat and breathe normally. She denied any new cosmetic product or drug use. There was no other abnormal systemic symptom. There was no family history of similar skin condition.

Physical examination revealed non-tender, diffuse swelling of the lower lip with fissured tongue (Figure 1). Neither eyelid swelling nor other skin lesions was found. Cranial nerves examination was normal and revealed no facial palsy. Other systems were within normal limits.

The histopathologic findings of the lower lip demonstrated multiple non-necrotizing granulomas in the dermis, composed of mononuclear cells, giant cells, epithelioid cells and few plasma cells (Figure 2). The overlying epidermis revealed confluent parakeratosis with pale glassy keratinocytes. No organism was found from the acid fast bacilli, GMS and PAS stains.



Figure 1 swelling of the lower lip

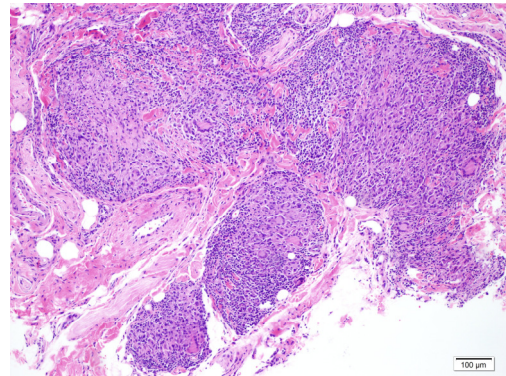


Figure 2 The histopathologic findings of the lower lip demonstrated multiple non-necrotizing granulomas in the dermis, composed of mononuclear cells, giant cells, epithelioid cells and few plasma cells. (H&E, X40)

Discussion

Cheilitis granulomatosa (CG) is a rare idiopathic inflammatory disorder clinically characterized by diffuse painless enlargement of one or both lips.¹ It was first described by Miescher in 1945.³ The age of onset is usually young adulthood with no racial or sexual predilection. The patients usually present with acute diffuse swelling involving the upper lip and-less often-the lower lip as the first manifestation which subsides completely in hours to days. Recurrent painless attacks

which increase in duration lead to persistent firm swelling. The swelling may slowly regress after some years. Spontaneous resolution is uncommon, but has been reported.⁴ The cause of CG is unknown.²

CG accompanied by facial palsy and plicated tongue referred to the Melkersson–Rosenthal syndrome (MRS).² However, the diagnosis of MRS is often difficult because the classic triad is not always complete at the time of presentation. Some authors suggest the use of the term oligosymptomatic MRS when the

patients have only two symptoms of the triad.⁵ The median time from first symptom to the diagnosis of the full triad of MRS was 4 years.⁶ Therefore, the presence of oligosymptomatic forms must be contemplated in advance in order to advise patients of the possibility of a full symptomatic triad and requires repeated observations and follow-up visits. Other associated symptoms that were reported in MRS patients together with the prevalence were shown in table 1.⁵

Table 1 Associated symptoms in Melkersson–Rosenthal syndrome patients⁵

Associated symptoms	Percentage
Facial pain	90.5%
Taste changes	42.8%
Headache (including migraine)	23.8%
Migraine	14.3%
Dizziness	19%
Tinnitus	4.8%

Table 2 Medical treatment of granulomatous cheilitis⁷

Medication	Dose
Triamcinolone ⁹	10–20 mg intralesionally with the interval between weeks to months
Clofazimine ^{10, 11}	100–300 mg daily or every other day
Minocycline ¹²	100 mg daily
Roxithromycin ¹³	150–300 mg daily
Metronidazole ¹⁴	750–1000 mg daily
Infliximab ¹⁵	3–5 mg/kg per infusion
Thalidomide ¹⁶	100 mg daily to every other day
Methotrexate ¹⁷	5–10 mg weekly, administered orally
Fumaric acid esters ¹⁸	120–720 mg daily
Tranilast ¹⁹	200–400 mg daily

Histopathologic findings in the early stages of the disease may only show interstitial edema and perivascular infiltration of lymphocytes. In the patients with long-standing lip swelling, the infiltrate becomes denser and focal non-necrotizing granulomas are formed with epithelioid cells and Langhans' type giant cells in the submucosal connective tissue. Special stains for mycobacteria and fungal organisms are recommended to exclude specific infectious diseases.⁴ The differential diagnosis of persistent upper lip swelling includes other granulomatous diseases such as a foreign body reaction, mycobacterial infection, sarcoidosis, Crohn's disease, Wegener's granulomatosis, histoplasmosis, amyloidosis, rosacea, medications such as ACE inhibitors and calcium channel blockers which can cause persistent angioedema, atopic reaction to a wide variety of allergens, and hereditary diseases such as C1 esterase deficiency.²

The treatment of CG is difficult. Until now, there is no systematic review of current treatments. Overview of current treatment from available literatures, mostly case reports and case series, is shown in table 2. Among them, corticosteroids are considered as a mainstay of treatment regimens, either as a sole treatment or an adjunct to other medical or surgical options.⁷ There were 2 case series, 13 patients in each study, demonstrated that almost all patients responded well to topical, intralesional or systemic corticosteroids depending on the severity of disease.^{1, 8} Cheiloplasty is

reserved for severe or deforming CG.⁷ Our patient was treated with intralesional triamcinolone 10 mg/cc monthly for 3 courses. Until now, the response is fairly good.

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