

PEDIATRIC DERMATOLOGY PHOTOQUIZ

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ASYMPTOMATIC UPPER LIP SWELLING IN AN 8-YEAR-OLD BOY

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An 8-year-old boy presented to our tertiary care center with a 10-month history of an asymptomatic enlarged swollen upper lip that was initially episodic but had recently become persistent (Fig. 1). He denied any exposure to new oral

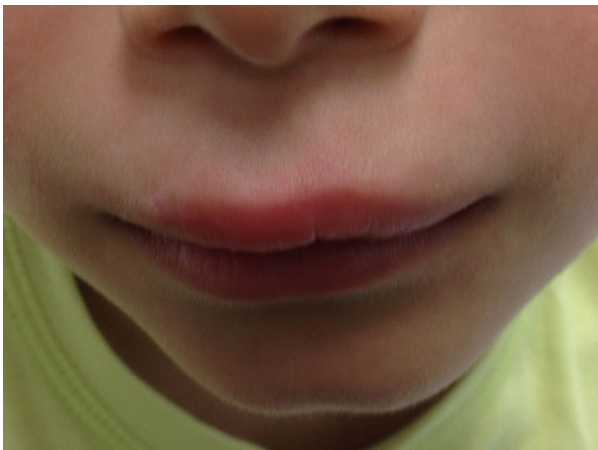


Figure 1. Asymptomatic, persistent, firm swollen upper lip.

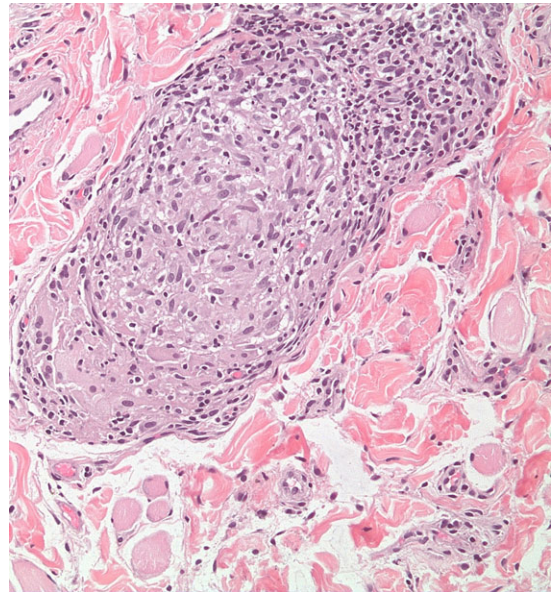


Figure 3. Close-up of the epithelioid granulomas.



Figure 2. Microscopy showed scattered, predominantly epithelioid histiocytic granulomas in the dermis.

agents such as toothpaste, lip balm, or cobalt-containing products when the swelling had initially started. He had no history of abdominal pain, blood or mucus in his stools, weight loss, or shortness of breath. He was initially diagnosed with angioedema and treated with cetirizine hydrochloride, with no improvement. He was otherwise healthy and taking no medications. Family history was noncontributory.

Physical examination revealed a smooth, nontender, edematous upper lip. There was no tongue fissuring or facial neurologic abnormalities. The findings from the remainder of the examination were normal. A punch biopsy specimen of the upper lip was taken (Figs. 2 and 3).

What is the diagnosis?

Diagnosis: Cheilitis granulomatosa

Microscopic Findings

Microscopy showed scattered predominately epithelioid histiocytic granulomas in the dermis (Figs. 2 and 3). Periodic acid–Schiff and acid-fast stains and polarization microscopy were negative. Laboratory values for complete blood cell count, angiotensin-converting enzyme, C1 esterase inhibitor, and erythrocyte sedimentation rate were within normal ranges. Purified protein derivative skin test was negative and chest radiograph was normal. Treatment with triamcinolone acetonide intralesional injections was initiated, with significant improvement after one session.

Discussion

First described in 1945, CG is a rare idiopathic, asymptomatic, persistent, firm swelling of the upper, lower, or both lips (1–4). CG has no racial or sex predisposition. The incidence of CG has been reported to peak between 15 and 25 years of age (1–4). Rarely CG has been reported in younger children, with the youngest patient being a 3-year-old boy (1–4). Similar to adults, there is no sex, racial, or ethnic predilection. Initially the lip swelling episodes are transient, subsiding in hours or days, but the frequency and duration of episodes usually increase with time until they become persistent.

The pathogenesis of CG has not been clarified. Suggested underlying causes include autoimmune mechanisms, allergic reactions to cobalt and food additives, local autonomic nervous system disturbance, infectious odontogenic foci, and tuberculosis (1–5). CG is regarded as one of the conditions of orofacial granulomatosis (OFG), which describes orofacial swelling secondary to noncaseating granulomatous inflammation (5). It can occur as an isolated condition or in the setting of Melkersson–Rosenthal syndrome (MRS), which is also characterized by facial palsy and fissured tongue (2). Some consider CG to be a monosymptomatic MRS form, especially since up to 25% of cases initially presenting as CG progress to the complete form of MRS (2–4). Orofacial swellings of the lip, chin, cheeks, and periorbital region are the most common symptom of MRS in childhood (2). In adults and children, CG has also been reported in association with Crohn's disease many years before or after onset of the disease (2–4). Especially in

children, early diagnosis of CG and its possible associated conditions is essential to avoid long-lasting functional complications and related psychological problems (2).

The differential diagnosis of CG includes hereditary angioedema, drug eruptions to medications such as angiotensin-converting enzyme inhibitors, and several diseases that may show granulomatous inflammation such as sarcoidosis, foreign body reaction, mycobacterial infection, and Crohn's disease (1–5). These can be excluded according to the clinical context, histopathologic evaluation, and relevant laboratory testing. Determining C1 esterase inhibitor levels can help exclude hereditary angioedema, and chest radiography and purified protein derivative test can exclude sarcoidosis and *Mycobacterium* infection. Gastrointestinal tract radiography and endoscopy may be performed to exclude Crohn's disease and patch testing to assess for allergies.

There is no standard treatment for CG (1–4). Treatment is usually tailored based on the severity of the condition. Mild swelling may improve with topical steroids, but intralesional or oral glucocorticoids are usually required in severe cases. Most pediatric cases of isolated CG have been reported to respond well to intralesional steroid therapy (1–4). For more severe gross disfigurement, surgery is an option. Variable effectiveness has been seen with other agents such as dapsone, clofazimine, hydroxychloroquine sulfate, metronidazole, tumor necrosis factor- α inhibitor, and sulfasalazine. Regardless of the therapeutic modality used, relapses are common.

References

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