

Chronic Lip Swelling and Crohn's Disease

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INTRODUCTION

Crohn's disease is chronic inflammation in the GI tract that can involve anywhere from the mouth to anus, although generally non-contiguous and restricted to the ileum and colon. Cutaneous Crohn's is an extra-intestinal manifestation of the disease.

PATIENT PRESENTATION

HISTORY OF PRESENT ILLNESS

- 27-year-old male with 2.5 years of chronic lip swelling following an upper respiratory infection
- Reports abdominal pain and lip numbness
- Denies nausea, vomiting, diarrhea, constipation, blood in stool, lip injections or lip trauma
- Previous work-up with Dermatology, ENT and Allergist in Northern California, records not available

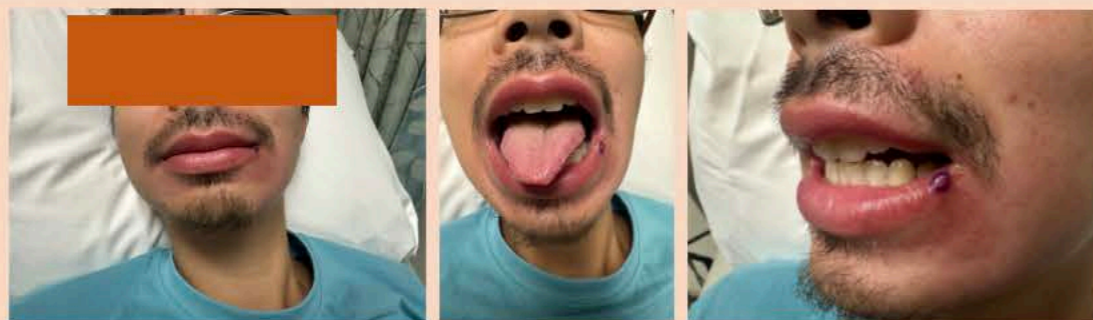
FAMILY HISTORY

- Positive: Inflammatory bowel syndrome (mother)
- Negative: Inflammatory bowel disease, connective tissue disorder, or hereditary angioedema

MEDICATIONS

- Benadryl (provides minimal improvements)
- Previous medications included betamethasone, cetirizine, clotrimazole, and short course of oral steroids

PHYSICAL EXAM

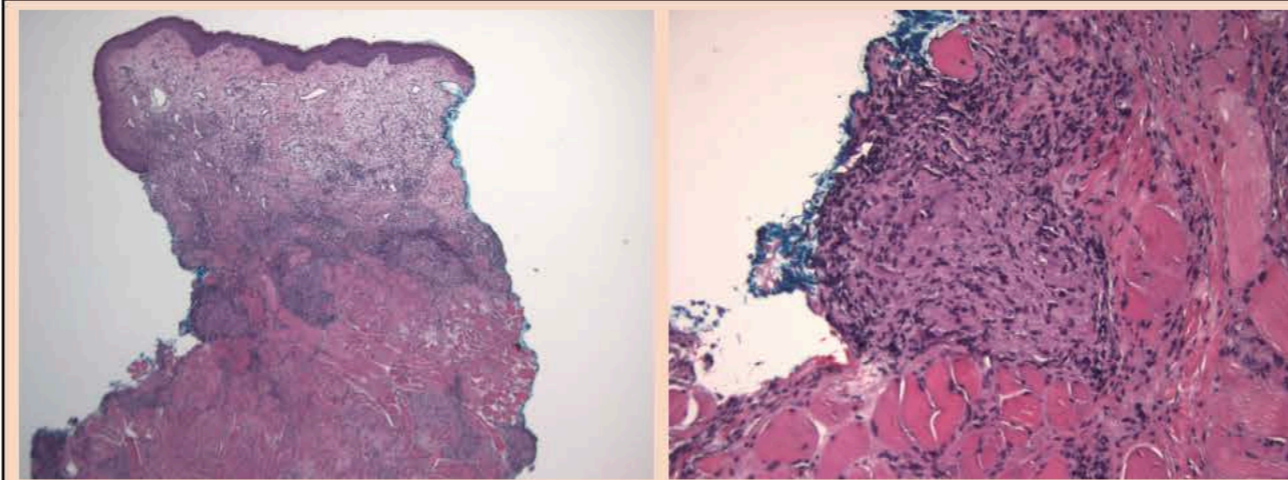


On physical exam, patient has uniform upper and lower lip swelling (when compared to photos on patient's phone from 3 years prior). No angular cheilitis or groves in the tongue present. Circled area is site of lip biopsy.

CLINICAL COURSE

- Dermatology consult
 - Biopsy performed which showed non-caseating granulomas
- Follow-up appointments patient started to complain of intense abdominal pain and diarrhea three times a week. Differential included Crohn's disease, infection (including TB, coccidioidomycosis, HIV), allergic dermatitis, and sarcoidosis
 - Diarrhea work up: elevated calprotectin, negative for infection
 - CRP: elevated
 - Coccidioides, HIV, TB: negative
 - Chest XR: negative for sarcoidosis lesions
- Gastroenterology consult
 - Colonoscopy with biopsy completed which showed "small bowel mucosa with acute ileitis, granulomas and architectural distortion," compatible with IBD like Crohn's disease
- Patient is currently on Humira with improvements in GI symptoms and lip swelling

PATHOLOGY



Left: "Shows superficial dermal edema with prominent telangiectasia and dense inflammatory infiltrate throughout the superficial and deep dermis" per Pathologist
Right: Central area showing aggregates of inflammatory cells, including giant cells, scattered lymphocytes and plasma cells without areas of necrosis, representing a granulomas

RESOURCES

1. Bennett, C. N., & Krishnamurthy, K. (2023). *Cutaneous Crohn Disease*. In StatPearls. StatPearls Publishing. Retrieved from <https://www.ncbi.nlm.nih.gov/books/NBK470311/>
2. Mackey, D. M., & Green, L. (1981). Cutaneous manifestations of Crohn's disease: Report of two cases and review of the literature. *Journal of the American Academy of Dermatology*, 5(6), 735-742. [https://doi.org/10.1016/S0190-9622\(81\)70130-0](https://doi.org/10.1016/S0190-9622(81)70130-0)
3. Safar, B., & Sands, D. (2007). Perianal Crohn's disease. *Clinics in Colon and Rectal Surgery*, 20(4), 282-293. <https://doi.org/10.1055/s-2007-991027>
4. Siroy, A., & Wasman, J. (2012). Metastatic Crohn disease: A rare cutaneous entity. *Archives of Pathology & Laboratory Medicine*, 136(3), 329-332. <https://doi.org/10.5858/arpa.2011-0020-RS>

DISCUSSION

About 40% of all patients affected by Crohn's disease experience at least one extra-intestinal manifestation including cutaneous lesions. Cutaneous Crohn's can be subdivided into three categories¹:

1. **Specific lesions** (have histological findings consistent with Crohn's disease on biopsy)¹
 - a. *Orofacial/perianal lesions* – suggest to be a direct extension of bowel disease to the skin¹
 - Orofacial lesions: lip swelling, angular cheilitis, gum inflammation, mucosal tags, and cobblestoning of the cheeks²
 - Perianal lesions: strictures, abscess, fistulae and skin tags³
 - b. *Metastatic Crohn's disease* – suggested to be antigens or immune complexes from GI tract circulating in blood and depositing in the skin¹
 - In adults may presents as papules, plaques, nodules, and ulcerations which can involve the arms, legs, genitalia, and face, often in moist environments⁴
 - In children often presents in the genitalia area⁴

2. **Reactive lesions** (do not share the same histopathological findings and suggested to be due to impaired function of neutrophil or altered cellular immunity)¹

- a. *Pyoderma gangrenosum* (more associated with Ulcerative Colitis, but can be seen in Crohn's disease)

3. **Associated lesions** (arise due to shared HLA-gene types or secondary to chronic inflammatory response)¹

- a. *Erythema nodosum* (occurs in about 6-15% of patients with Crohn's disease)

CONCLUSION

Chronic lip swelling is an atypical initial presentation of Crohn's disease. This case underscores the importance of considering Crohn's disease in the differential diagnosis of persistent orofacial swelling. Early recognition and multidisciplinary management are crucial for diagnosis and effective treatment.