Metastatic Cutaneous Crohn Disease of the Face: A Case Report and Review of the Literature

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The estimated time to complete this activity is 1 hour.

GOAL

To understand metastatic cutaneous Crohn disease to better manage patients with the condition

LEARNING OBJECTIVES

Upon completion of this activity, you will be able to:

1. Discuss mucocutaneous manifestations of Crohn disease.
2. Recognize the histologic findings of metastatic cutaneous Crohn disease.
3. Evaluate treatment options for metastatic cutaneous Crohn disease.

INTENDED AUDIENCE

This CME activity is designed for dermatologists and general practitioners.

CME Test and Instructions on page 12.

This article has been peer reviewed and approved by Michael Fisher, MD, Professor of Medicine, Albert Einstein College of Medicine. Review date: December 2009.

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Metastatic cutaneous Crohn disease is a rare entity with an uncertain etiology. Few cases of facial involvement have been reported. We describe a 45-year-old woman who presented with several large, dusky, erythematous plaques and draining sinuses on her cheeks in the setting of chronic intestinal Crohn disease. A review of the literature regarding metastatic cutaneous Crohn disease also is provided.

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Case Report

A 45-year-old woman with a history of biopsy-proven intestinal Crohn disease presented with several large, dusky, erythematous plaques on both cheeks (Figure 1).

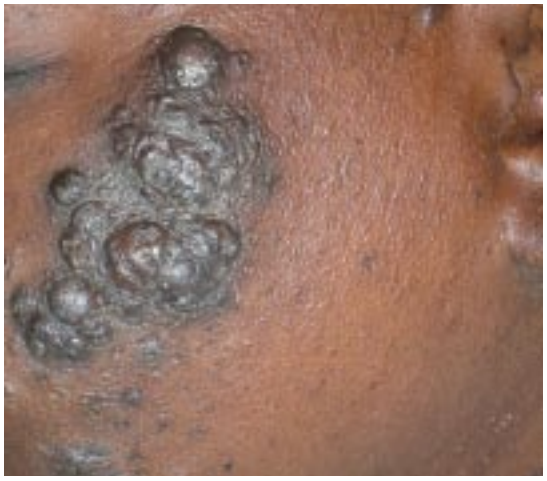

Figure 1. Dusky, erythematous, fistulous nodules in a cobblestone pattern on the patient's malar area. These lesions drained a turbid yellow-tinged serous fluid when palpated.

Similar lesions were found on the patient's glabella, brow, retroauricular areas, and back. Many of these plaques contained fistulous nodules in a cobblestone pattern that drained a turbid yellow-tinged serous fluid when palpated.

The patient reported that the facial lesions began as small papules several years prior that slowly enlarged. She initially did not seek specific treatment for the lesions but reported a mild improvement in the color and size of the skin lesions when taking infliximab for intestinal Crohn disease in 1999. Over the years, she noted an increase in the number and size of these facial lesions.

The clinical differential diagnosis of the facial lesions included metastatic cutaneous Crohn disease, deep fungal infection, mycobacterial disease, cutaneous sarcoidosis, foreign body reactions, and other granulomatous disorders. Tissue cultures were negative for bacteria and mycobacteria. A biopsy specimen from a facial lesion revealed lymphohistiocytic inflammation with noncaseating granulomas and multinucleated Langerhans-type giant cells consistent with metastatic cutaneous Crohn disease (Figures 2 and 3).

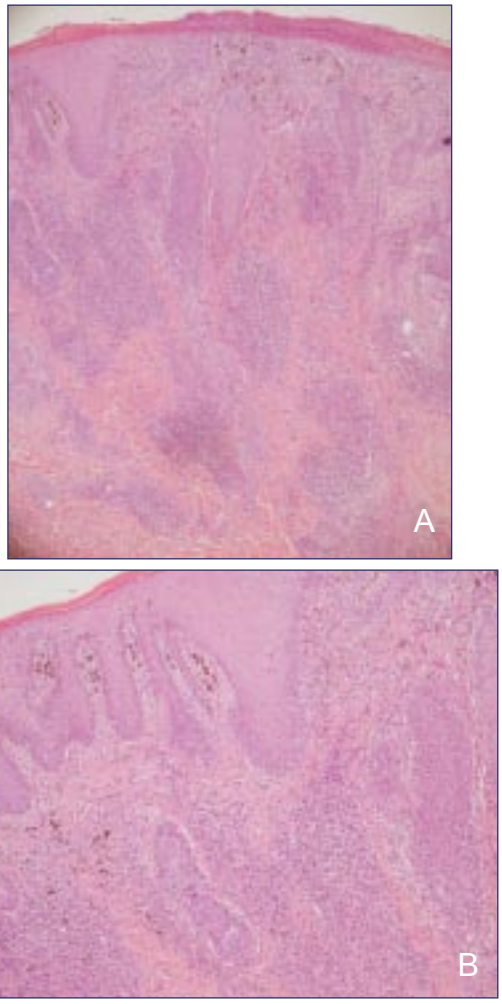

Figure 2. Histopathologic features of a skin biopsy specimen from the malar area demonstrating superficial and deep lymphohistiocytic inflammation with noncaseating granulomas (A and B)(H&E; original magnifications ×5 and ×10, respectively).

Comment

Mucocutaneous manifestations of Crohn disease can occur in 22% to 44% of patients.[1] The most common lesions are nonspecific reactive skin findings such as erythema nodosum and pyoderma gangrenosum as well as cutaneous manifestations of nutritional deficiencies. Despite its initial description by Parks et al[2] in 1965, metastatic cutaneous Crohn disease is a rare entity with fewer than 100 cases reported in the literature and only a small percentage of reported cases involving the face.[3-6] It is characterized by noncaseating granulomatous cutaneous lesions lacking a connection to diseased areas of the gastrointestinal tract.[7] Metastatic cutaneous Crohn disease usually presents after manifestation of gastrointestinal tract symptoms, though 20% of patients have skin lesions that precede the diagnosis of intestinal Crohn disease by up to 8 years.[1,8] Metastatic cutaneous Crohn disease most commonly is seen in patients with involvement of the colon that is unrelated to bowel disease.[9]

The macroscopic appearance of metastatic cutaneous Crohn disease can take several different forms. Genital disease initially presents as erythema and swelling. Nongenital disease usually presents as dusky, erythematous to purple plaques or nodules, followed by ulceration with undermining edges, draining sinuses and fistulae, and ultimately scarring.[10] The clinical differential diagnosis includes granulomatous disorders such as deep fungal infection, mycobacterial disease, cutaneous sarcoidosis, and foreign body reactions. The most frequent sites of involvement include the intertriginous and flexural

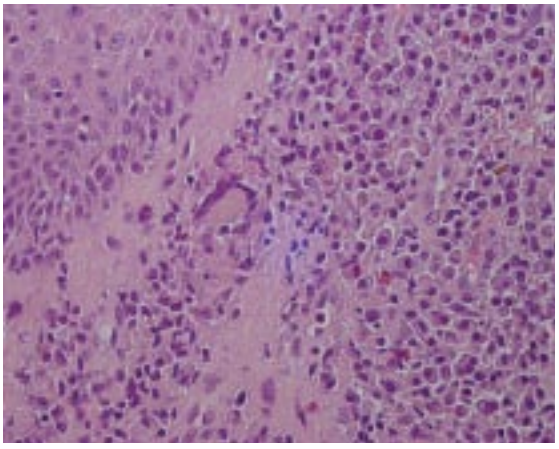

Figure 3. Histopathologic features of a skin biopsy specimen demonstrating a multinucleated Langerhans-type giant cell; many of these cells were found scattered within the lymphohistiocytic inflammation (H&E, original magnification ×40).

areas of the body, with a predilection for the lower extremities.[11] Involvement of the vulva, penis, trunk, upper extremities, and face (in order of decreasing frequency) also has been described.[1] Multiple sites, as seen in our patient, rarely are involved.[9,12,13]

Histologically, metastatic cutaneous Crohn disease looks identical to intestinal Crohn disease. The lesions consist of nodular, noncaseating, epithelioid tubercles surrounded by lymphocytes in the superficial and deep dermis that sometimes extend into the subcutaneous fat. A few scattered multinucleated Langerhans-type giant cells and a sparse perivascular lymphohistiocytic infiltrate also can be seen. Microscopically these perivascular lymphocytes and monocytes appear similar to vasculitis and have been called granulomatous perivasculitis by Burgdorf and Orkin.[14]

Treatment of metastatic cutaneous Crohn disease is largely anecdotal because there have been no controlled trials and few case reports. Oral metronidazole 250 mg 3 times daily has been shown to be effective and frequently is combined with topical or intralesional corticosteroids. Some success also has been reported with azathioprine, oral corticosteroids, infliximab, sulfasalazine, and 6-mercaptopurine.[7,15] Surgical resection also has been used with success in a handful of cases, particularly those refractory to medical management.[15] Cutaneous lesions also have been reported to respond to treatment of the underlying intestinal disease.[11]

REFERENCES

1. Burgdorf W. Cutaneous manifestations of Crohn's disease. *J Am Acad Dermatol.* 1981;5:689-695.
2. Parks AG, Morson BC, Pegum JS. Crohn's disease with cutaneous involvement. *Proc R Soc Med.* 1965;58:241-242.
3. Biancone L, Geboes K, Spagnoli LG, et al. Metastatic Crohn's disease of the forehead. *Inflamm Bowel Dis.* 2002;8:101-105.
4. Chen W, Blume-Peytavi U, Goerdt S, et al. Metastatic Crohn's disease of the face. *J Am Acad Dermatol.* 1996;35:986-988.
5. Graham DB, Jager DL, Borum ML. Metastatic Crohn's disease of the face. *Dig Dis Sci.* 2006;5:2062-2063.
6. Kolansky G, Kimbrough-Green C, Dubin HV. Metastatic Crohn's disease of the face: an uncommon presentation. *Arch Dermatol.* 1993;129:1348-1349.
7. Berkowitz EZ, Lebwohl M. Cutaneous manifestations of inflammatory bowel disease [letter]. *J Eur Acad Dermatol Venereol.* 2000;14:349-350.
8. Ploysangam T, Heubi JE, Eisen D, et al. Cutaneous Crohn's disease in children. *J Am Acad Dermatol.* 1997;36 (5, pt 1):697-704.
9. Guest GD, Fink RL. Metastatic Crohn's disease: case report of an unusual variant and review of the literature. *Dis Colon Rectum.* 2000;43:1764-1766.
10. McCallum DI, Kinmont PDC. Dermatological manifestations of Crohn's disease. *Br J Dermatol.* 1968;80:1-8.
11. Tavarela Veloso F. Review article: skin complications associated with inflammatory bowel disease. *Aliment Pharmacol Ther.* 2004;20(suppl 4):50-53.
12. Schwab RA, Giblin WJ, James WD, et al. Multiple cutaneous ulcerations. cutaneous ulcerations in Crohn's disease (metastatic Crohn's disease). *Arch Dermatol.* 1993;129:1067-1068, 1610-1611.
13. Shum DT, Guenther L. Metastatic Crohn's disease. case report and review of the literature. *Arch Dermatol.* 1990;126:645-648.
14. Burgdorf W, Orkin M. Granulomatous perivasculitis in Crohn's disease. *Arch Dermatol.* 1981;117:674-675.
15. Kafity AA, Pellegrini AE, Fromkes JJ. Metastatic Crohn's disease. a rare cutaneous manifestation. *J Clin Gastroenterol.* 1993;17:300-303.