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

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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.


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).
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).

Comment

Mucocutaneous manifestations of Crohn disease can occur in 22% to 44% of patients. 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 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. It is characterized by noncaseating granulomatous cutaneous lesions lacking a connection to diseased areas of the gastrointestinal tract.

The macroscopic appearance of metastatic cutaneous Crohn disease can take several different forms. Genital disease initially presents as erythema and swelling. Non-genital disease usually presents as dusky, erythematous to purple plaques or nodules, followed by ulceration with undermining edges, draining sinuses and fistulae, and ultimately scarring. 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 areas.
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Figure 3. Histopathologic features of a skin biopsy specimen demonstrating a multinucleated Langerhans-type giant cell; many of these cells were 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.3,9,12,13

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 peri-vascular 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, sulphasalazine, 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


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