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Metastatic Crohn's Disease

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GRANULOMATOUS LESIONS OF THE SKIN not attributable to a known foreign body reaction or infectious agents are often presumed to be sarcoidosis, a disease of exclusion. We report the case of a patient initially thought to have cutaneous sarcoidosis, who on further investigation was determined to have "metastatic" Crohn's disease.

Report of a Case

The patient, a 62-year-old man, first consulted our dermatology service for evaluation of an erythematous, scaly, nummular patch on his left lower leg that had been present for about two months.

Further history indicated that six months before he noted the lesion, he had been ill with malaise, diarrhea, vomiting, and a 9-kg (20-lb) weight loss. He was admitted to hospital and underwent an extensive evaluation, including radiographic studies that revealed tumors in the antrum of the stomach and terminal ileum (Figure 1). These were initially considered to be consistent with adenocarcinoma, but an endoscopic biopsy of the gastric lesion showed many noncaseating granulomata. No other systemic illnesses or abnormalities were identified at that time. A diagnosis of Crohn's disease was made after tuberculosis and other causes for the granulomata were ruled out. He was treated with a regimen of prednisone, 30 mg a day, with complete resolution of all gastrointestinal symptoms. The patient had been completely weaned off prednisone at the time of being seen by us.

About two months after the prednisone therapy was discontinued, he was examined in the dermatology clinic because of mild erythema and scaling that had developed on both legs. Several potassium hydroxide preparations were negative, and the patient was initially treated with mid-strength topical steroids with no apparent response. The lesions progressed, and eventually thickened, indurated plaques developed with sharply marginated ulcerations and crusting (Figure 2).

Several skin biopsies revealed sarcoidal granulomas with few giant cells and little coagulation necrosis (Figure 3). Special stains and cultures were negative for deep fungi, acid-fast bacteria, and spirochetes. No foreign body material was identified by polarization. A diagnosis of sarcoidosis was initially made, and a complete evaluation for systemic involvement was embarked on. All results were normal. The patient had no adenopathy. A chest x-ray film showed no hilar

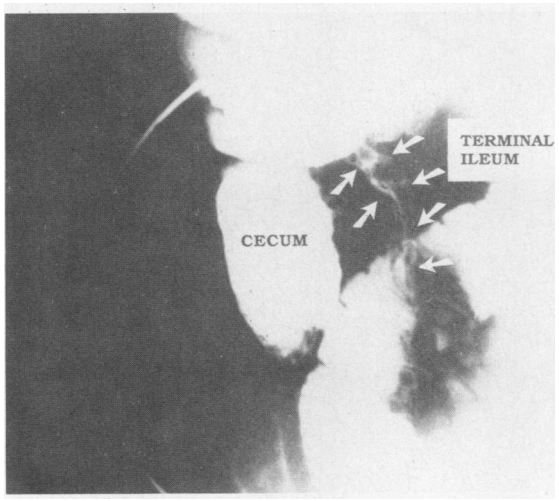

Figure 1.—A barium-enhanced radiographic examination of the intestine reveals cuffing of the terminal ileum (arrows) characteristic of Crohn's disease.

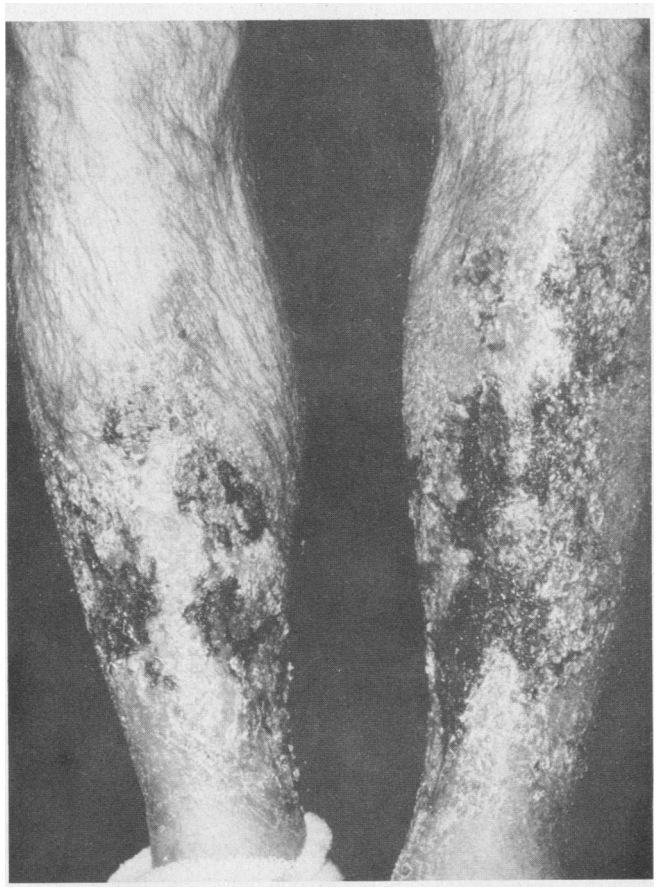

Figure 2.—The initial skin findings consisted of dermatitis and scaling of the legs within which there is ulceration and crusting.

(McGillis ST, Huntley AC: Metastatic Crohn's disease. West J Med 1989 Aug; 151:203-205)

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adenopathy nor evidence of tuberculosis. Angiotensin-converting enzyme levels were within normal limits. On routine laboratory studies, all values including serum calcium levels were normal. An ophthalmologic evaluation revealed no ocular lesions. Serum immunoglobulin levels were also normal. A repeat gastroendoscopy showed no recurrence of the mucosal lesions. No other cutaneous lesions were noted at that time.

Because of the established diagnosis of Crohn's disease involving his bowel and the lack of involvement in other target organs typical for sarcoidosis, his cutaneous reaction seemed to best fit into the category of "metastatic" Crohn's disease. He was treated with a regimen of oral prednisone with gradual resolution of the skin lesions (Figures 4 and 5). Following resolution of the leg lesions, the prednisone therapy was again discontinued. Over a period of months, histologically and clinically similar new skin lesions developed on his back. Prednisone was readministered with an excellent response, and he has been lesion-free for more than a year.

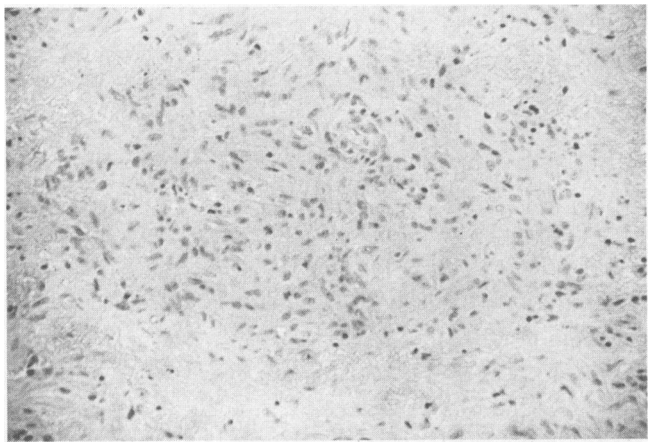

Figure 3.—A high-power field of the reticular dermis from the skin biopsy of the patient's leg shows a sarcoidal granuloma (hematoxylin and eosin, x 400).

Discussion

This case is an example of metastatic Crohn's disease and shows some of the difficulty in differentiating between Crohn's disease and sarcoidosis, two diseases of diagnostic exclusion that can have similar clinical and histologic involvement of the skin. Differentiation is based primarily on the pattern of extracutaneous involvement.

Our initial working diagnosis was cutaneous sarcoidosis. His skin findings were similar to those recently reported in two other patients with sarcoidosis,[1,2] but in both those patients systemic evidence of sarcoid was well documented; in our patient, the only other disorder was of his gastrointestinal tract. When making a diagnosis of sarcoidosis in the skin, involvement of other target locations is sought. Sarcoidosis most commonly involves the lungs (90%), followed by skin (30%), lymph nodes (27%), and eyes (27%). Gastrointestinal involvement is distinctly rare for this disease.[3] Based on the results of an endoscopic biopsy, his gastrointestinal disorder was diagnosed as Crohn's disease. The skin lesions are also clinically and histologically consistent with this process.

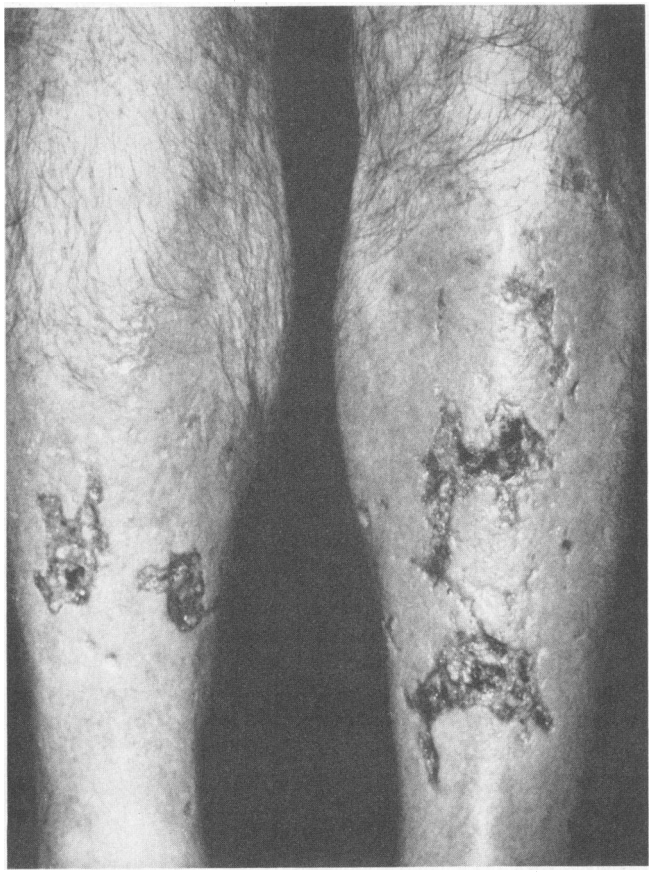

Figure 4.—Six weeks after prednisone therapy was started for his skin problem, the patient's dermatitis is moderately resolved and the punched-out nature of the ulcers is more clearly shown.

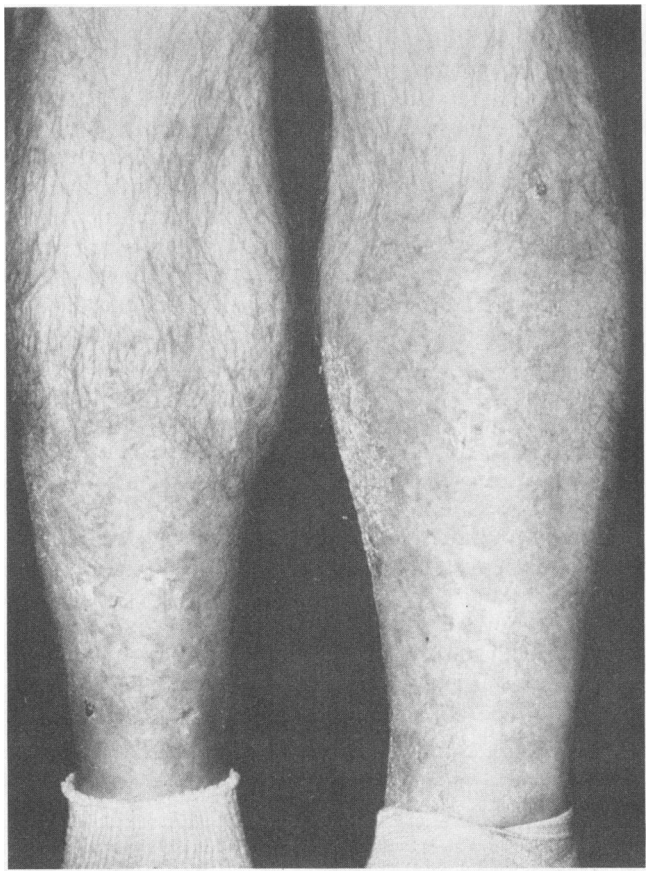

Figure 5.—Substantial improvement of the skin of the patient's legs is seen after about three months of prednisone therapy.

In fact, it is not uncommon for Crohn's disease to show extraintestinal involvement: as many as 44% of patients have skin findings.[4] Most of these lesions occur in close proximity to the gastrointestinal tract and thus are thought to be due to direct extension. These include perineal ulceration, fistula formation, and granulomatous peristomal lesions. Patients with large bowel and colon involvement are most likely to have skin disease.

So-called metastatic Crohn's disease refers to extraintestinal involvement not due to direct extension of the disease. Of 15 reported cases of metastatic Crohn's disease, specific skin lesions have included nodules, plaques with and without ulceration, ulcerated patches, lichenoid papules, intertriginous ulcerations, erysipelas-like eruptions, and vulval swelling.[5,6] There does not seem to be a relationship between bowel activity and cutaneous disease.[7] Granulomatous ulceration of the extremities, similar to our patient's initial cutaneous presentation, has been reported.[8]

Nonspecific cutaneous lesions may also occur in Crohn's disease. Such lesions include oral mucosal lesions such as aphthous ulcers and a mucosal cobblestone pattern and those lesions that occur as the result of nutritional deficiency, such as acrodermatitis enteropathica.[9] Other skin reactions noted in Crohn's disease include pyoderma gangrenosum, erythema nodosum, erythema multiforme, epidermolysis bullosa acquisita, finger clubbing, and palmar erythema.[10] Like sarcoidosis, cutaneous Crohn's disease occurs in many forms.

On histologic examination, metastatic Crohn's disease shows dermal or subcutaneous noncaseating granulomas for which infections, sarcoid, foreign body granulomas, and tuberculosis have been ruled out.

This case illustrates the important point that sarcoidosis and Crohn's disease may have similar cutaneous findings. Crohn's disease should be added to the list of diseases to be excluded in patients with cutaneous sarcoidal granulomas, especially in those patients with gastrointestinal complaints.

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