Metastatic Crohn's Disease
Case Report of an Unusual Variant and Review of the Literature


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PURPOSE: Metastatic Crohn's disease is a rare complication of Crohn's disease that has been infrequently reported in the literature. We report a case where submammary, inguinal, and perineal disease was observed in a patient many years after a proctocolectomy. The proliferative and polypoid morphology of the cutaneous lesions has not been previously described. In addition, this case describes severe cutaneous metastatic Crohn's disease in the absence of active gastrointestinal disease, which to our knowledge has not been reported in the literature. RESULTS: A 55-year-old female with a 25-year history of Crohn's disease was investigated and treated over a 12-month period for metastatic Crohn's disease involving the submammary, inguinal, and perineal areas. These proliferative lesions with erythema and ulceration were histologically consistent with metastatic Crohn's disease. Gram and Ziehl Nielsen stains revealed no pathogenic organisms. The use of topical solutions, antibiotics, immunosuppression, and surgery failed to produce any significant benefit. A review of 42 cases of metastatic Crohn's disease in the literature is reported. CONCLUSION: Cutaneous metastatic Crohn's disease has an extremely variable macroscopic appearance and may be a source of considerable morbidity. It can be present without other significant symptomatology, although it more commonly parallels gastrointestinal disease activity. There are no trials to guide current treatment, which is mainly based on anecdotal reporting. [Key words: Crohn's disease; Metastatic; Crohn's disease, cutaneous; Proctocolectomy; Immunosuppression; Granuloma]


Metastatic Crohn's disease (MCD) is a rare complication of Crohn's disease that has been infrequently reported in the literature. The features are those of cutaneous involvement, noncontiguous with the gastrointestinal tract, which bears the pathologic features of noncaseating granulomas consistent with Crohn's disease. Most cases are characterized by ulcerative skin lesions which usually occur on limbs or in skin flexure creases and may be difficult to heal. The skin lesions may precede the diagnosis of Crohn's disease but are rarely present without active disease in the gastrointestinal tract. We report an unusual case of MCD with polypoid granulomatous skin lesions occurring during a period of quiescent gastrointestinal involvement.

REPORT OF A CASE

A 55-year-old female with a long history of Crohn's disease presented through the outpatient department with extensive erythematous skin involvement and edematous, polypoid lesions involving the perineum, labia, bilateral inguinal region, and submammary creases. She had an extensive past history of complicated Crohn's disease initially diagnosed 25 years previously at laparotomy for cecal perforation.

The early management of her Crohn's disease involved laparotomies for persistent abdominal pain with disease confined to the large bowel and extensive perianal abscesses, which initially responded to prolonged courses of oral steroids. Subsequently, her gastrointestinal and perianal disease required a diverting ileostomy. Perianal abscesses and a watering can perineum were treated during an inpatient stay of 11 months and ultimately led to a proctocolectomy 15 years before her current problems.

Despite some peristomal abscesses, she remained relatively well, gaining weight from 50 to 75 kg over a five-year period before developing further extensive perineal abscesses and sinuses. These were managed surgically during a period of many months before symptomatic resolution occurred and the perineum seemed to be healed. A period of some eight years of good health followed until her present condition arose.

During a 12-month period she experienced persistent skin lesions involving the perineum, bilateral inguinal areas, and submammary folds (Fig. 1). The areas were mixed ulcerating and proliferative polypoid areas (Fig. 2) that were not contiguous with the peristomal ulceration around the end ileostomy. She described considerable pruritus and a persistent serous ooze from the affected areas. These had not responded to outpatient dressings and had become troublesome enough to necessitate inpatient admission. Biopsy of the perineal, inguinal, and submammary lesions revealed epidermal hyperplasia with multinucleated giant-cell granulomata and deep perivascular infiltrate rich in plasma cells. Gram, Ziel

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Nielsen, and periodic acid-Schiff stains were repeatedly negative in all initial biopsies and in subsequent biopsies during six months. She had not experienced any gastrointestinal symptoms and a whole-body indium-labeled white cell scan did not reveal any active bowel disease. During this time of severe cutaneous disease, itchy and Watery eyes were also a feature of her symptomatology. Laboratory investigations indicated a raised erythrocyte sedimentation rate and C-reactive protein with depletion of iron stores but no anemia, leukocytosis, or zinc deficiency.

Initial treatment with topical agents (Burrow’s solution, sorbolene, topical steroid, and an antifungal agent to the submammary areas) failed to produce a significant improvement. Surgical excision and ablative diathermy to the inguinal areas resulted in healing but with regrowth of the proliferative polypoid lesions during a four-month period. The patient refused oral prednisolone, and treatment with azathioprine and methotrexate was initiated but abandoned because of side effects of nausea and dizziness, respectively. Intermittent courses of antibiotics had also failed to improve the cutaneous disease.

DISCUSSION

Cutaneous manifestations of Crohn’s disease are relatively common and include three distinct types. The most common is perianal and peristomal ulceration, where the pathologic process of Crohn’s disease in the gastrointestinal tract encroaches on the adjacent skin. The second type includes a variety of conditions which have been consistently reported in the literature as having a strong association with Crohn’s disease: pyoderma gangrenosum, erythema nodosum, erythema multiforme, acrodermatitis enteropathica, and epidermolysis bullosa acquista. The third type, granulomatous cutaneous lesions noncontiguous with the gastrointestinal tract is rare, first being described by Parks et al. in 1965.

This type is known as metastatic Crohn’s disease, the term coined by Mountain in 1970 when describing three patients with submammary, penile, and abdominal wall ulcerating skin lesions separate from the gastrointestinal tract and showing the pathologic features consistent with Crohn’s disease. Since the recognition of this manifestation it has been described in the world literature infrequently and seems to be an uncommon feature of Crohn’s disease. It seems to have a predilection for skin creases, where there is a moist environment and close apposition of skin as in the submammary fold, perineal and inguinal areas, and abdominal skin fold creases. However, there are many cases reported of limb lesions and it has also been described on the vulva, penis, trunk, and facial area.

Pathologic features of MCD require skin lesions, usually ulcerative, to be noncontiguous with the gastrointestinal tract and demonstrate on histopathology the granulomas characteristic of Crohn’s disease. Macroscopically, the skin lesions are most commonly described as ulcerating, but plaques, papules, and nodules have also been described. At histopathology, multinucleate giant cells and noncaseating granulomas