

METASTATIC CROHN'S DISEASE

S Criton

Crohn's disease is a granulomatous inflammatory disease of the gastrointestinal tract. It can involve other sites of the body also. When such lesions are separated from Crohn's lesion of the intestine by normal skin, they are referred to as metastatic Crohn's disease. Two such cases are reported.

Key words: Crohn's disease, Metastatic

Introduction

Crohn's disease is a granulomatous inflammatory disease which can affect any part of the gastrointestinal tract, most commonly involves the terminal part of the ileum. The process of Crohn's disease is not confined to the gastrointestinal tract but can involve other sites also.¹ When such lesions are separated from Crohn's disease of the intestine by normal skin, they are referred to as metastatic Crohn's disease. Two such cases are reported.

Case Reports

CASE 1

A 23-year-old unmarried woman was seen with painful ulceration of the perineum, genitalia and medial aspect of both thighs. She developed these as erythematous painful raised lesions which gradually increased in size; some of the lesions turned vesicular and others became ulcerated. The vesicles ruptured and turned out to be ulcers. Along with these skin lesions she had fever, arthralgia and arthritis involving all larger joints. There was no history of similar episode previously.

From the Department of Dermato-Venereology,
Medical College Hospital, Trichur-680 596, India.

Address correspondence to Dr. S Criton

She was diagnosed as having Crohn's disease in November '92 when she had fever, vomiting, diarrhoea, arthritis and erythematous nodules below both knees. She was given salazopyrine and with this treatment she improved. For the past 6 months, before the episode, she discontinued the treatment on her own. There was no history of recurrent oral ulceration, conjunctivitis, sexual contact or intake of any form of iodides or bromides.

Examination revealed a sick patient with fever, tachycardia and pallor. There was no cyanosis or jaundice. She was normotensive.

There were multiple tender nodules with ulceration in the perineum and medial aspect of both thighs. There were two ulcers in the inner aspect of both labia majora, one on either side. The margins were sloping and border well defined. The floor was covered with pus. Inguinal groups of lymphnodes on either side were enlarged, discrete and mobile. Perianal region was normal. The vagina was normal.

The standard haematological and biochemical work up were normal. The blood VDRL test and HIV screening were also non reactive. The ulcer smear and tissue smear were negative for

organisms. Smear for amoeba from the edge of the ulcer of the vulva was negative. Skiagram of the chest was normal.

Histopathological study of the biopsy taken from the ulcer of the thigh showed ulcerated skin and granuloma composed of epithelioid cells, giant cells and lymphocytes. There was no vasculitis.

Case 2

A 24-year-old woman was seen for evaluation of recurrent genital ulceration since one year. The lesion started as a papule and then ulcerated. The ulcers developed at different sites on the external genitalia. Sometimes it was seen after sexual intercourse. There was no history of oral ulceration or redness of eyes. The patient was previously diagnosed as having Crohn's disease and was on treatment with salazopyrine. She looked ill and appeared pale. There were two tender oval ulcers of size 2x1cm involving the inner aspect of both labia majora. The margins were well defined and there was no undermining of edges. The floor was covered by granulation tissue and it was not attached to deeper structure. There was no bleeding on manipulation. The regional lymphnodes were not involved.

The patient was investigated and no haematological or biochemical abnormalities could be detected. Ulcer smear, tissue smear, dark field microscopy were all negative. Serology for syphilis and HIV were non reactive. The biopsy of the ulcer showed tissue covered with stratified squamous epithelium showing ulceration and mild dysplastic changes. The subepithelial region showed granulation tissue and a few non caseating granuloma with giant cells.

Both the patients were given tab pred-

nisolone 60mg in divided doses for a month and responded well.

Discussion

The cutaneous manifestations of Crohn's disease are present in 22% to 44% of cases.^{3,4} These manifestations may be divided into two categories; those in which histologic examination demonstrates characteristic features of Crohn's disease and those in which the pathologic findings are not specific for any diagnostic entity. The commonest lesions are perianal and peristomal ulcers, as well as fistula and abscess formation.² Other dermatological diseases described in association with Crohn's disease include pyoderma gangrenosum, erythema nodosum, oral aphthae, and skin changes secondary to malabsorption. The uncommon lesions claimed to be associated with Crohn's disease are erythema multiforme, epidermolysis bullosa acquisita, polyarteritis nodosa,⁵ and vitiligo.

There are instances where the process of Crohn's disease was not confined to the bowel, but extended to sites remote from gastrointestinal tract.¹ When such lesions are separated from Crohn's disease of the intestine by normal skin, they are referred to as metastatic Crohn's disease.² Metastatic cutaneous Crohn's disease (MCD) developed during quiescent as well as active periods of intestinal disease and presented as ulcers, nodules, crusted nodules or plaques.⁶

The clinical appearances of MCD is relatively distinctive and it is possible to distinguish it from other dermatologic entities associated with Crohn's disease.⁶ However, a variety of other cutaneous conditions primarily granulomatous, show resemblance to MCD.⁶ Hence a biopsy is indicated in the evaluation of nodulo-ulcerative cu-

taneous lesions in patients with Crohn's disease.

Microscopically, MCD is characterised by granulomatous inflammation, often with giant cells and discrete non-caseating granuloma.⁷ Usually there will not be any vascular involvement.

In our case no. 1, the cutaneous lesions appeared when the disease was not active, but in case 2, it appeared when there was active disease. This shows that the cutaneous Crohn's disease can occur any time irrespective whether the disease is active or not, so also whether the patient is on treatment or not.

Many treatment modalities were tried in metastatic Crohn's disease such as oral metronidazole,¹ intralesional,⁸ and systemic steroid. In our patients treatment with systemic steroid was very effective. Although there is no treatment of choice, treatment of the underlying gastro-intestinal disease has been associated with healing of skin lesions in several cases.⁶ But one of our patients was on treatment for Crohn's disease when

the cutaneous lesions appeared. It may point out that the treatment of intestinal Crohn's disease alone may not be sufficient for cutaneous metastasis.

References

1. De Vroede G, Schaefer G, Sanchez G et al. Crohn's disease of the vulva. *Am J Clin Pathol* 1975; 63:348-358.
2. Duhra P, Paul C J. Metastatic Crohn's disease responding to metronidazole. *Br J Dermatol* 1988; 119:87-91.
3. Samitz M H, Dana A S, Rosenberg P. Cutaneous vasculitis in association with Crohn's disease. *Cutis* 1970; 6: 51-55.
4. Mc Callum D I, Kinmont P D. Dermatological manifestations of Crohn's disease. *Br J Dermatol* 1968;80:1-8.
5. Goslen J B, Graham W, Lazarus G S. Cutaneous polyarteritis nodosa - Report of a case associated with Crohn's disease. *Arch Dermatol* 1983;119:326-29.
6. Sutphen J L, Cooper P H, Mackel S E, et al. Metastatic cutaneous Crohn's disease. *Gastroenterology* 1984;86:941-944.
7. Levine N, Bangert J. Cutaneous granulomatosis in Crohn's disease. *Arch Dermatol* 1982;118:1006-1009.
8. Algre V A, Pujol C, Calatayud A, et al. Cutaneous Crohn's disease: treatment with intralesional steroid. *Int J Dermatol* 1989; 28:552-554.