Skin lesions as the presenting symptom of a gastrointestinal disease

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A 38-year-old woman developed ulcerating skin lesions on the upper and lower limbs and anterior chest, accompanied by low-grade fever. These lesions evolved rapidly from erythaematous furunculoid papulopustules to necrotic ulcers and were very painful. A trial of antibiotic treatment (for suspected furunculosis) was ineffective. One month later a migratory arthritis appeared, with no improvement on nonsteroidal anti-inflammatory agents. A few weeks later, she had numerous episodes of diarrhoea, with bright red blood rectally. At this time she was admitted to the internal medicine department for evaluation. Physical findings were unremarkable, except for necrotic ulcers on the upper and lower limbs (figures 1 and 2) and anterior chest and arthritis of left elbow and left ankle. The X-ray of the involved joints showed no erosive disease and there was no infectious aetiology demonstrable. IHA for amoeba, stool ova and parasites, and stool cultures were negative. Proctosigmoidoscopy showed granular, friable mucosa with severe ulcerations and histopathological features of acute and chronic inflammation. The biopsy of the skin ulceration showed polymorphonuclear infiltration and oedema in the epidermis associated with interstitial and perivascular infiltration with lymphocytes, and mononuclear cells in the dermis with moderate evidence of vasculitis.

Figure 1

Figure 2

Questions

- 1 What is the dermatological diagnosis?
- 2 Which group of gastroenterological diseases is this dermatological disease associated with?
- 3 Which of these gastroenterological diseases is it classically associated with?
- 4 Which other skin manifestation can be found in this group of gastroenterological diseases?

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Answers

QUESTION 1

Pyoderma gangrenosum. This is an ulcerating skin disease of unknown cause. The diagnosis is a clinical one, in as much as no specific diagnostic criteria exist. There is no consensus in the literature concerning the histopathological changes.

Although pyoderma gangrenosum may develop at any anatomic site, the majority of lesions occur on the lower extremities and less frequently on the trunk. In figures 1 and 2 there are enlarging, necrotic ulcers with bluish indeterminate borders, surrounded by zones of erythema on the upper and lower limbs.

QUESTION 2

Pyoderma gangrenosum is one of two skin manifestations of inflammatory bowel disease.

QUESTION 3

Pyoderma gangrenosum was classically associated with ulcerative colitis. Association with Crohn's disease is considered a rare extra-intestinal complication.

QUESTION 4

The other skin manifestation of inflammatory bowel disease is erythema nodosum. It is more frequently seen with Crohn's disease.

Discussion

More than 50% of patients with pyoderma gangrenosum have an underlying systemic disease. It has been associated with a number of chronic diseases (monoclonal gammopathy, chronic active hepatitis, Hodgkin's disease, polyarthritis, inflammatory bowel disease).¹

An immunologic aetiology of pyoderma gangrenosum is currently favoured. Abnormalities of humoral immunity, cell-mediated immunity and complement have all been described. Extraintestinal manifestations of Crohn's disease, such as joint and eye involvement, may all have an immunological basis.²

The incidence of pyoderma gangrenosum in patients with Crohn's disease is less than 1.5%.^{3,4} When it does occur in patients with Crohn's disease, it is more frequent when there is colonic involvement.⁵

Generally, the bowel disease precedes the onset of skin disease by about five years, but may also occur concomitantly.³ It is rarely the presenting symptom of Crohn's disease.

The activity of the pyoderma gangrenosum frequently parallels the activity of the bowel disease and treatment is directed at the underlying Crohn's disease.

We conclude that the great majority of patients with pyoderma gangrenosum have associated systemic disease. Inflammatory bowel disease is probably the most significant association while an association with Crohn's disease is very rare. Thus, all patients presenting with pyoderma gangrenosum must undergo thorough investigations of the gastrointestinal tract and be followed up carefully, even after the skin lesions have healed.

Final diagnosis

Pyoderma gangrenosum as the presenting symptom of Crohn's disease.

Keywords: pyoderma gangrenosum, inflammatory bowel disease, Crohn's disease

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