PYODERMA GANGRENOSUM AND INFLAMMATORY BOWEL DISEASE- CASE REPORTS

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SUMMARY
Two cases of ulcerative colitis preceded by pyoderma gangrenosum in two female patients are described. One patient developed leg ulcers, presumed to be due to sickle cell disease; the other developed crops of ulcers all over the body for several years. These patients attended and were managed at the Medical and the Reconstructive Surgical Units of the Korle Bu Teaching Hospital. The unusual presentation of pyoderma gangrenosum and the importance of diagnosing underlying probable inflammatory bowel disease is highlighted.

Keywords: Pyoderma gangrenosum, inflammatory bowel disease, bloody diarrhoea, infliximab.

INTRODUCTION
Inflammatory bowel disease usually presents with chronic diarrhoea, usually bloody. There are many other extra-intestinal modes of presentation, including erythema nodosum, pyoderma gangrenosum, spondylitis, stunted growth and delayed puberty. These may sometimes precede bowel symptoms by several weeks or even years or develop during the course of active bowel disease and may result in delayed or even missed diagnosis. We report on two such patients who presented with pyoderma gangrenosum.

Case One
A 14 year old girl was referred from a private herbal clinic in May 2003 with iron deficiency anaemia. She had bilateral leg ulcers and was suspected to be a sickle cell disease patient.

The patient had started developing multiple ulcers which started as boils, on the lower legs, about one month before referral. There were associated fever and chills and the patient was unable to walk on account of severe pains. She had previously been admitted to another hospital in Accra because of anaemia.

On admission the ulcers were described as oval shaped with necrotic and haemorrhagic sloping edges oozing with pus; one on the left shin measured about 10 x 4cm and one on the right shin 5 x 3cm. The surrounding skin was darkly pigmented. Palpable lymph nodes in the groins were tender. Her haemoglobin was 7.9gm/dl with a microcytic picture; there was neutrophilia. Her sickling was negative and Hb electrophoresis was AA.

The patient was transfused and given antibiotics. The ulcers continued to enlarge with exposure of the underlying subcutaneous tissues (Figure 1). Swab of the ulcers yielded Enterobacter species sensitive to gentamycin and amikacin; she was treated with the latter drug intravenously. At this point pyoderma gangrenosum or vasculitic ulcers was suspected; biopsy submitted for histology confirmed the diagnosis of pyoderma gangrenosum. She was started on oral Prednisolone 30mg daily.

Further history revealed that the patient had had episodes of loose stools for several years, and that she had not attained menarche. She was stunted for her age (weight 24kg) and lacked any development of secondary sexual features. Stool examination showed several pus cells and serum protein was low. A diagnosis of inflammatory bowel disease was made and Sulphasalazine 1gm tds was added to her therapy.
Flexible colonoscopy revealed changes consistent with chronic ulcerative colitis which was confirmed on histology of colonic mucosa. Double contrast Barium enema showed total chronic ulcerative colitis (Figure 2).

**Figure 2** Barium enema X-ray of large colon showing chronic ulcerative colitis changes

The skin ulcers were grafted by the plastic surgeon. The patient improved but has been lost to follow up.

**CASE TWO**

A 43-year old female was referred to the Dermatology clinic with an 18 year old history of recurrent multiple ulcers. These started as painful boils which later ulcerated and were associated with fever and joint pains. During physical examination the whole body was covered with numerous roundish scars of different sizes (Figure 3) as well as punched out ulcers on the back and left foot (Figure 4).

**Figure 3** Pyoderma gangrenosum multiple scars on legs

**Figure 4** Ulcerated pyoderma gangrenosum on the foot.

There were no buccal lesions. Pyoderma gangrenosum was diagnosed and the patient was started on Prednisolone. Dapsone and Azathioprine were introduced later. The patient complained of diarrhoea which later became bloody, requiring admission. Further history revealed that for about 5 years she had had intermittent loose stools but without blood or mucus. The recent episode was the first time of passing bloody mucoid stools. Her haemoglobin ranged between 10 to 12 gm/dl, ESR was raised at 72 mm fall/hr; other laboratory tests were normal. Flexible sigmoidoscopy revealed changes compatible with active ulcerative colitis confirmed by histology. Sulphasalazine was added to the treatment, the patient improved and was discharged home to be followed up as an outpatient.

**DISCUSSION**

Pyoderma gangrenosum is a relatively uncommon skin condition of unknown aetiology but suspected to be an autoimmune disorder. It usually starts as a blister, swelling or pustule which then ulcerates with a necrotic centre and undermined edges. The ulcer may be single or in groups, is painful and slow to heal. Four main types are described - ulcerated, pustular, bullous, vegetative. About 25% to 50% of pyoderma are related to inflammatory bowel disease, of which 1% to 2% are affected, and may predate the bowel symptoms for weeks to several years. In one study of 116 patients with ulcerative colitis, 2.4% had associated pyoderma gangrenosum. Sometimes the lesions may be associated with flare up bowel symptoms but may also develop or worsen when bowel activity is quietest. There are other conditions associated with pyoderma gangrenosum; these include rheumatoid arthritis and other connective tissue disorders, chronic autoimmune hepatitis, myeloid blood dyscrasias. Cases associated with Takayasu’s arteritis in Japan, Wegener’s granulomatosis, collagenous colitis, and colonic adenocarcinoma have all been described. The incidence of pyoderma gangrenosum associated with inflammatory bowel disease in Ghana or West Africa is not known; however a case has been described in Ibadan, Nigeria. Treatment includes dressing of small ulcers, oral steroids, sometimes with immunosuppressive drugs such as Azathioprine, Methotrexate or Cyclosporine. Antibiotics are not effective unless there is evidence of bacterial suprainfection. Lately Infliximab, an anti alpha-tumour necrosis factor (α-TNF) monoclonal antibody, has been given as intravenous infusion to treat resistant ulcers with good results.
may require skin grafting as in the first patient. The underlying cause must, of course be treated.

The first patient had bouts of chronic diarrhoea for many years probably from childhood, resulting in stunted growth, chronic anaemic and delayed puberty. She developed the skin lesions relatively recently which then led to the diagnosis of inflammatory bowel disease. Her ulcers had been attributed to sickle cell disease and the diarrhoea treated empirically. Pyoderma gangrenosum is noted to be rare in under 15 year olds but cases have been reported. In one such case the patient developed crops of skin lesions over several months followed by aggressive acute ulcerative colitis. Both settled only after total colectomy.

This second patient, on the other hand, had had numerous skin lesions all over the body over a period of 18 years leaving extensive multiple scars; the diagnosis of pyoderma gangrenosum had been missed until recently. She had had intermittent non-bloody stools for about 5 years and had only recently developed severe bloody diarrhoea. Her haemoglobin had been reasonable the lowest being 10gm/dl whereas the first patient required blood transfusion for severe anaemia. Chronic non-healing ulcers may be misdiagnosed as sickle cell ulcers as shown in case one. Both patients had sought treatment at various health facilities including traditional healers. The care of such patients requires careful expert and long term management.

Chronic inflammatory disease has the potential for malignant change especially of colon in those who have had lesions for over ten years; however malignant change appears to be uncommon in Africans. Regular colonoscopic examination with mucosal biopsies to detect early malignant or dysplastic changes is necessary. Elective colectomy is recommended for patients with disease over 10 years although this may not be acceptable in our local setting. It is suggested that skin ulcers with unusual features which fail to heal must be further investigated to exclude pyoderma gangrenosum. Confirmed cases should be investigated for underlying systemic disease particularly inflammatory bowel disease.

ACKNOWLEDGEMENTS

We are grateful to the following: Mr. Albert Paintsil, Consultant Plastic Surgeon, International Reconstructive Plastic Surgery & Burns Centre, for performing skin grafting for patient Case 1; the Nursing staffs of the Medical Units, Endoscopy Unit for their care of these patients; the Pathology and Radiology Departments for the histology and x-ray reports respectively. The photographs were taken by Medical Illustration Department and Dr. Patrick Adjei of Korle Bu Teaching Hospital.

REFERENCES


