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Pyoderma gangrenosum producing saddle nose deformity

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Abstract
Pyoderma gangrenosum affecting the nose is rare and this may lead to diagnostic confusion because of the large differential diagnosis. As diagnosis is made, largely, on the basis of exclusion the treatment of pyoderma gangrenosum may be unduly delayed. The condition is often disfiguring, particularly following inappropriate surgical intervention, and early diagnosis is therefore important.

We present a case of pyoderma gangrenosum managed initially in the community with minor surgery and resulting in the rare complication of saddle nose deformity.

Key words: Pyoderma, gangrenosum; Nose deformities, acquired; Colitis, ulcerative

Case report

A 67-year-old woman presented to her family doctor with a five-day history of painful inflammation across the bridge of the nose. Her past medical history was notable for mild ulcerative colitis of seven years duration treated with oral mesalazine 400 mg b.d. The nasal lesion was treated with oral flucloxacillin and ampicillin 250 mg q.d.s. and there was a good response, with near complete resolution of the inflammation, apart from a localization of pus to the left of the nasal bridge which was incised and drained in the doctor's surgery.

Eleven days after this minor surgical procedure the patient returned to her GP with further 'acute cellulitis' of the nose but on this occasion the erythema had spread to the lower eyelids, cheeks and upper gums. A five-day course of oral phenoxymethylpenicillin 500 mg q.d.s. and metronidazole 400 mg t.d.s. failed to settle the condition and the patient was admitted to the department of otolaryngology.

On admission the patient was apyrexial with a painful violaceous ulcer across the bridge of the nose with a necrotic base (Figure 1). There was surrounding erythema spreading to both cheeks associated with a mild vestibulitis and a yellow post-nasal discharge, the rest of the physical examination was unremarkable. Blood tests revealed an elevated white cell count (WCC) 15.1×10^9/l with a neutrophilia of 11.7×10^9/l. All other parameters were normal.

A presumptive diagnosis of infective nasal cellulitis was made and the patient commenced with intravenous cefuroxime 750 mg t.d.s. and metronidazole 500 mg t.d.s.

Four days of intravenous antibiotics saw no resolution with the beginnings of a saddle nose deformity and the development of further smaller lesions on the left breast, right thumb (Figure 2) and at a venepuncture site on the left wrist. The ESR became elevated to 90 mm/hr and the patient developed frequent bouts of bloody diarrhoea. Biopsy results showed granulation tissue with no specific diagnostic features, absence of granulomata and negative stains for fungi and mycobacteria. Tests for rheumatoid factor, Wassenman reaction (WR), antinuclear antibodies and antineutrophil cytoplasmic antibodies gave negative results.

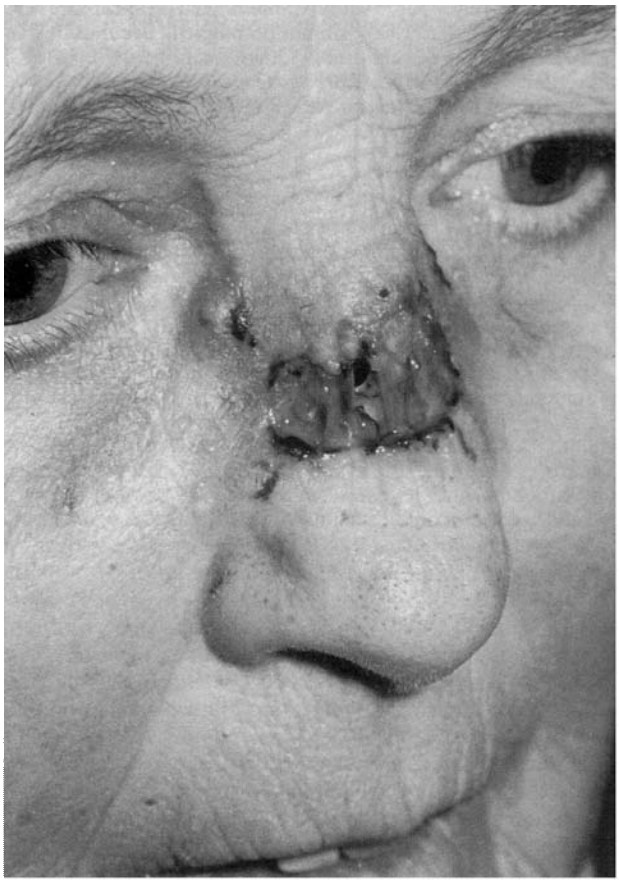

FIG. 1
Painful violaceous ulcer with necrotic base

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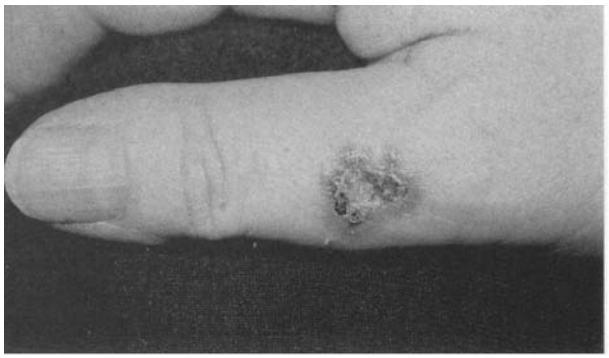

FIG. 2
Pyogenic granuloma on right thumb

A diagnosis of pyoderma gangrenosum was considered and the patient treated with oral prednisolone 60 mg o.d. together with colifoam enemas b.d. and mesalazine suppositories 250 mg b.d. A rapid improvement in the cutaneous lesions resulted. Unfortunately the patient's ulcerative colitis continued to deteriorate leading, two weeks later, to an emergency panproctocolectomy for perforated toxic megacolon. The patient made a good post-operative recovery but was left with a disfiguring saddle deformity of the nose.

Discussion

This case highlights the fact that pyoderma gangrenosum of the head and neck may mimic other chronic inflammatory conditions leading to diagnostic confusion and delay, with subsequent misguided attempts at surgical intervention.

Pyoderma gangrenosum may not be on the clinician's usual list of causes of chronic or non-healing lesions of the nose. Such a list would include the specific chronic infections, tuberculosis, syphilis, leprosy and fungal infections and the non-specific granulomata comprising Wegener's granulomatosis, sarcoidosis and high grade T cell lymphoma (formerly lethal midline granuloma).

The principal means of diagnosing pyoderma gangrenosum depends upon the recognition of the hallmark ulcerative skin lesions and the exclusion of other causes. As seen in this case the presence of inflammatory bowel disease supports the diagnosis. The ulcers are usually painful, multiple and most often located on the legs. The lesion orginates as a red papule that becomes a pustule and demonstrates central necrosis in a matter of hours. Frank ulceration occurs in one or two days (White, 1985) with the characteristic ragged, undermined, violaceous border. The aetiology of the disease is unclear but is probably due to an underlying neutrophil abnormality. Head to toe physical examination is essential because multiple lesions and lesions associated with minor trauma will suggest the diagnosis (Fowler and Callen, 1983; Prystowsky *et al.*, 1989). Other evaluation should include Gram stain and culture of the wound, serology for syphilis, autoimmune antibodies including antineutrophil cytoplasmic antigen to exclude Wegener's granulomatosis, serum vasopressin and a biopsy at the peripheral rim of erythema (White, 1985). Note that biopsy of these ulcers serves more to exclude other diagnoses than it does to confirm pyoderma gangreosum because there are no truly diagnostic histopathological markers (Su *et al.*, 1986). Fifty per cent of patients with pyoderma gangrenosum have an associated systemic disease. Ulcerative colitis is the most common association and with this in mind sigmoidoscopy +/- biopsy should be performed in all suspected cases to exclude occult inflammatory bowel disease (Hickman and Lazarus, 1980; Fowler and Callen, 1983).

Management consists of treatment of the lesion together with therapy directed at controlling associated systemic disease (Chow and Ho, 1996). With ulcerative colitis the course and response to therapy usually parallels that of the skin disease (our case is atypical in this respect) (Holt *et al.*, 1980). First line therapy consists of oral steroids sometimes complemented by immunosuppressants. In less severe cases topical or intralesional injection of steroid is indicated (Chow and Ho, 1996). Surgical intervention, however, should be avoided as this results in progression of the disease (Fowler and Callen, 1983; Prystowsky *et al.*, 1989).

Pyoderma gangrenosum affecting the head and neck is rare and if located on the nose it may result in particular diagnostic difficulty due to the extensive differential diagnosis. Resultant saddle deformity has been reported twice in the world literature (Snyder, 1986; Kramer *et al.*, 1990) but disfigurement is common and poses a particular management problem in that surgical reconstruction is contraindicated. It is for this reason that early diagnosis is important and misguided attempts at surgical closure or grafting should be avoided.

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