Cyclosporin-responsive hidradenitis suppurativa

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Hidradenitis suppurativa, a chronic inflammatory condition of the apocrine gland follicles, may rarely be complicated by pyoderma gangrenosum (PG). We report such a case, in which the immunosuppressant cyclosporin A (CyA) was given to treat PG and a dramatic improvement occurred in the patient's intractable perineal HS.

CASE REPORT

A 48-year-old man presented in July 1984 with an 8-year history of recurrent abscesses of the buttocks, unresponsive to repeated courses of antibiotics. On examination there were multiple abscesses and sinuses on buttocks, perineum and scrotum, draining a purulent exudate. Erythrocyte sedimentation rate (ESR) was 60 mm/h, white blood count 15.0×10^9/l with neutrophilia and serum globulin 49 g/l. Swabs cultured mixed Gram-positive and Gram-negative bacteria. A diagnosis of hidradenitis suppurativa (HS) was made. He failed to respond to prolonged antibiotic treatment and a 16-week course of UVB, became depressed and was lost to follow-up.

In July 1992 he presented with weight loss and a 4-month history of two painful purulent ulcers on the right leg, clinically resembling PG. By now the area of HS had extended to upper thighs: right groin lymphadenopathy and nail clubbing were also noted. Histology of the ulcers showed dermal fibrosis, aggregates of acute and chronic inflammatory cells and lymphocytic infiltration of vessel walls, consistent with pyoderma gangrenosum (PG). Biopsy of an enlarged groin node showed non-specific reactive hyperplasia. In view of a markedly raised ESR (125 mm/h), IgA (13 g/l) and IgG (34 g/l), serum immunoelectrophoresis was performed, which showed no abnormal bands. A bone marrow examination showed reactive myeloid hyperplasia; Bence-Jones protein was negative.

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Despite oral minocycline for 5 months the PG ulcers enlarged and buttock suppuration worsened. When pustules developed on the right forearm, cyclosporin A (CyA) was introduced at a dose of 4.5 mg/kg/day. After 4 months the ulcers were considerably smaller and by 8 months they had healed. The HS markedly improved with healing of discharging sinuses and diminished pain. This clinical benefit is maintained after 15 months of treatment with CyA and continuous broad-spectrum oral antibiotics; there has been a subjective improvement in quality of life. Creatinine clearance remains within normal limits.

DISCUSSION

PG is frequently the cutaneous manifestation of a systemic disorder, particularly inflammatory bowel disease, ankylosing spondylitis, rheumatoid arthritis and haematological neoplasms. In spite of our patient's lymphadenopathy, weight loss and raised ESR and serum globulin, we found no underlying haematological malignancy and attributed the development of PG to HS. The rare association of PG with HS has been previously reported in a small number of cases[1,2]. Hypergammaglobulinaemia may occur in PG[3], but in this case may have resulted from chronic suppuration. While many defects in both cellular and humoral immunity have been documented in PG, a common defect has not been discovered[4]. CyA which suppresses lymphokine production and T-cell activation, was first reported[5] to be an effective treatment for PG in 1985 and since then has been used successfully in a number of patients.

The cause of the chronic suppuration in HS is unknown. In 1976, a study of seven patients with HS found cell-mediated immunity and neutrophil function to be normal[6]. A later study of 27 patients found seven of the most severely affected to have a marked reduction in T-lymphocyte counts[7]. An increase in circulating suppressor T-cells corrected by tolmetin sodium has been demonstrated[8]; a patient with a defect in polymorphonuclear leucocyte killing of bacteria associated with low levels of intracellular cyclic GMP has also been described[9]. Other suggested causes of HS are bacterial infection, comedonal occlusion of follicles, relative oestrogen excess or androgen deficiency, impaired glucose tolerance and genetic factors. Treatment is difficult, usually involving prolonged courses of broad-spectrum antibiotics though antiandrogens, retinoids or surgery may be used in selected cases. Gupta *et al.*[10] in 1991 reported the only previous use of CyA for HS, in a 60-year-old man in whom a moderate response was noted.

The occurrence of PG as a complication of HS indicates a possible disorder of immunity underlying the chronic suppurative process. The favourable response to CyA in this case supports this theory. Immunosuppressive therapy may be useful in refractory cases of HS.

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Primary systemic amyloid with nail dystrophy

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We report a patient with the classical cutaneous findings of primary systemic amyloidosis, due to myeloma. He had developed a nail dystrophy, which is a recognized, but rare, feature in systemic amyloid.

CASE REPORT

A 67-year-old man presented with a 1 year history of recurrent black eyes whenever he coughed or vomited. Over the previous 3 months he had developed malaise, breathlessness, nausea, weight loss, constipation, and tingling in both hands. His voice was hoarse and his tongue had become swollen, making swallowing difficult.

On examination the most striking feature was the bruising around the eyes (Figure 1). He also had purpuric lesions on the chin. His skin had a yellowish tinge, and was slightly thickened over his hands and fingers. The nails were dystrophic with longitudinal striations (Figure 2). His tongue was enlarged with waxy purpuric plaques in the centre, and the indentations of his teeth could be seen on the edges. General examination revealed an ill man with postural hypotension, and a neuropathy in the carpal tunnel distribution.

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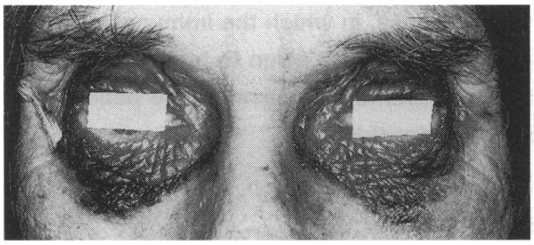

Figure 1 **Bilateral periorbital ecchymoses**

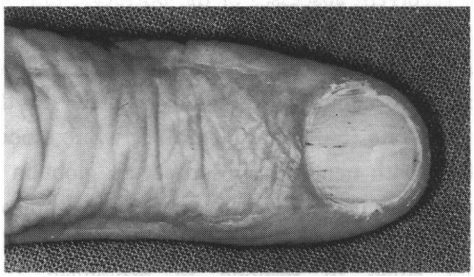

Figure 2 **Dystrophic nails, showing longitudinal striations**

The following investigations were normal: full blood count, biochemistry, clotting screen, and skeletal survey. IgG was normal at 10.8 g/l but IgM and IgA were low at 0.5 g/l and 0.7 g/l, respectively. Immunoelectrophoresis demonstrated monoclonal light chains in the blood and urine. Bone marrow examination revealed an increase in plasma cells (22% of nucleated cells) confirming the diagnosis of myeloma.

Biopsy from facial skin showed an abnormal dermis, expanded by a fissured amorphous eosinophilic material. This material showed positive staining with Sirius Red, and under polarized light the dichromic birefringence, characteristic of amyloid, was seen. This staining was preserved after exposure to potassium permanganate, distinguishing the deposits as amyloid L.

Our patient became increasingly unwell with admissions for retention of urine and gastrointestinal obstruction. Small