

Sweet's Syndrome and Pyoderma gangrenosum Associated with Ulcerative Colitis

E. C. BENTON,¹ DANIEL RUTHERFORD² and J. A. A. HUNTER¹

¹*Department of Dermatology, Royal Infirmary, Edinburgh, and*

²*Gastrointestinal Unit, Western General Hospital, Edinburgh, Scotland*

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A young woman with ulcerative colitis developed pyoderma gangrenosum during the active phase of the disease and Sweet's syndrome (acute febrile neutrophilic dermatosis) three months after panproctocolectomy. (Received June 14, 1984.)

E. C. Benton, Department of Dermatology, The Royal Infirmary, Lauriston Place, Edinburgh, EH3 9YW, Scotland.

CASE REPORT

A 24-year-old woman developed severe ulcerative colitis of the distal colon in 1979. Treatment with sulphasalazine 3 g daily and hydrocortisone acetate foam (Colifoam) administered rectally did not produce complete remission, and prednisolone 20 mg daily was also required to control her symptoms. In July 1981 she was readmitted to hospital with a severe relapse of the colitis. She was ill and pyrexial, and examination revealed on the medial aspect of her buttocks two purulent ulcers with overhanging reddish-blue edges; these were characteristic of pyoderma gangrenosum. She had also severe ulcerating anal disease which exposed the internal anal sphincter muscles. Blood transfusion and intravenous corticosteroids produced an initial improvement. However, eleven days after admission, the colon perforated and the patient underwent a subtotal colectomy, leaving the rectal stump in situ. Histological examination of the resected colon showed acute, necrotising ulcerative pan-colitis; there was no evidence of Crohn's disease. Systemic steroids were withdrawn gradually postoperatively but oral iron supplements continued. Six months later the rectal stump was excised as an elective procedure.

There was, however, no improvement in her general health. She lost 4 kg in weight in spite of an adequate diet. The ESR remained elevated at 76 mm in 1 hour Westergren, and she showed a persistent anaemia, Hb 8.9 g/l, with a serum iron of 4 μ mol/l (normal 14–22 μ mol/l) and transferrin 1.47 mg/l (normal 2–4 mg/l). Total serum proteins were raised at 98 g/l (normal 60–80 g/l) with an increase in alpha 2 and gamma globulin fractions. IgM was 454 i.u./l (normal 20–160 i.u./l), and IgG was 218 i.u./l (normal 90–170 i.u./l). A jejunal biopsy showed minimal non-specific inflammatory change. Three months after excision of the rectal stump she developed dusky-red, infiltrated plaques, some pustular in appearance on the cheeks, forehead and dorsum of the hands (Fig. 1). A clinical diagnosis of Sweet's syndrome was made. This was confirmed by skin biopsy which showed an intact epidermis, oedema of the dermal papillae and a dense perivascular infiltrate of predominantly polymorphonuclear cells, many of which showed leucocytoclasia. There was some endothelial swelling but no perivascular fibrin deposition. The total white cell and differential counts were normal. Raised levels of both IgG and IgM persisted and circulating immune complexes were detected by the C1q assay.

Prednisolone 15 mg daily was commenced and this resulted in rapid resolution of the skin lesions without scarring. Her general health improved steadily, her weight increased and the anaemia resolved. The dose of prednisolone was gradually reduced to 7.5 mg daily, and the clinical improvement was maintained eighteen months later.

COMMENT

Cutaneous signs may occur in up to a third of patients with ulcerative colitis (1), although many of these are non-specific e.g. urticaria, purpura or erythematous eruptions. More specific changes include erythema nodosum and pyoderma gangrenosum, 40–50% of patients with the latter condition having ulcerative colitis.

Sweet's syndrome or acute febrile neutrophilic dermatosis, originally described in 1964 (2), is characterised by the eruption of tender, well demarcated, dusky, erythematous plaques whose histology shows dermal oedema, a dense perivascular infiltrate of polymorphonuclear cells, but no evidence of vasculitis. Apart from the fever and skin lesions, there may be other systemic manifestations e.g. anaemia, leucocytosis, elevated ESR, arthritis, myalgia, and renal and hepatic involvement (3).

The majority of cases in the original series were preceded by infection, often respiratory. However, since then the syndrome has been reported especially in association with

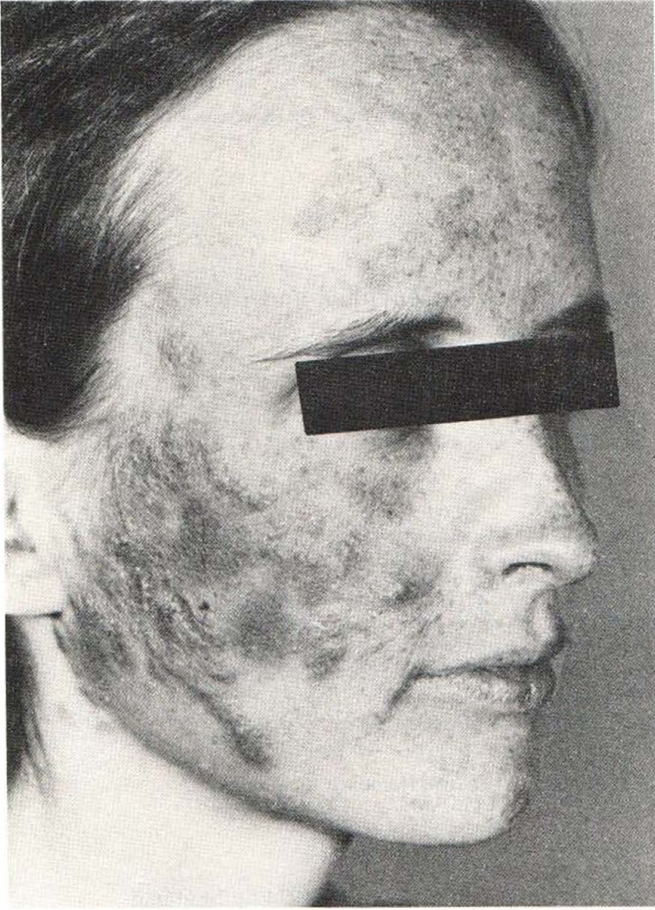


Fig. 1. Dusky red infiltrated plaques on cheeks and forehead.

myeloid leukaemia, but also with other primary and secondary malignancies and Sjögren's syndrome (3). Only two cases have been reported in association with ulcerative colitis (4).

The clinical and histological features define this syndrome as a distinct entity, but occasionally there are morphological and histological similarities between Sweet's syndrome and pyoderma gangrenosum. However, ulceration is always present in established lesions of pyoderma gangrenosum and histologically there is not only loss of epidermis but a far greater degree of dermal necrosis in association with an acute inflammatory infiltrate. Nevertheless, both conditions have been described in association with a similar range of diseases and it is possible that they represent two ends of a nosological continuum with a similar underlying pathogenesis (5). The occurrence of both pyoderma gangrenosum and acute febrile neutrophilic dermatosis in our case would support this concept. We postulate that the unusually severe degree of anal mucosal destruction which occurred in our patient, and her failure to thrive after panproctocolectomy, might also be related to the same underlying cause.

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