Sweet's Syndrome Associated with Crohn's Disease

C. BECUWE,[1] E. DELAPORTE,[2] J. F. COLOMBEL,[1] F. PIETTE,[2] A. CORTOT[1] and H. BERGOEND[2]

[1]*Department of Gastroenterology and* [2]*Department of Dermatology A, University Hospital Center, Lille, France*

The first known case involving an association of Sweet's syndrome with Crohn's disease is described. A 36-year-old woman developed a diarrhea, fever, and infiltrated erythematous cutaneous plaques on neck and limbs, consistent with a presumptive diagnosis of Sweet's syndrome. This was confirmed by a skin biopsy showing a dense dermal infiltrate of polymorphonuclear leukocytes. Crohn's disease, extending from the anus to the terminal ileum, was diagnosed as well. Prednisolone treatment resulted in the improvement of both the bowel disease and skin lesions. ***Key words: Inflammatory bowel disease; Neutrophilic dermatosis.***

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F. Piette, Department of Dermatology A, C.H.U. Lille, F-59037 Lille, France.

The systemic complications of inflammatory bowel diseases may include a number of skin disorders such as pyoderma gangrenosum, erythema nodosum, aphthous ulcers, vasculitis, and epidermolysis bullosa acquisita (1, 2). In recent years, several cases of Sweet's syndrome (acute febrile neutrophilic dermatosis) have also been described in association with ulcerative colitis (3, 4). We report here the first known case of Sweet's syndrome occurring during the course of Crohn's ileocolitis.

CASE REPORT

In October 1987, a previously healthy 36-year-old woman suddenly developed diarrhea, fever (38.5°C) and a skin eruption characterized by tender, well-demarcated, infiltrated erythematous plaques predominant on neck and limbs. The presumptive clinical diagnosis of Sweet's syndrome was confirmed by a skin biopsy showing an intact dermis, edema of the dermal papillae, and a perivascular infiltration of leukocytoclastic polymorphonuclear leukocytes into the mid- and upper dermis. There was some endothelial swelling but no perivascular fibrin deposition. Direct immunofluorescent studies failed to demonstrate immunoglobulin and complement deposits in the affected skin. Laboratory examinations revealed a leukocyte count of 9.3×10^6/l with 82% neutrophils and an elevated ESR of 60 mm in 1 hour Westergreen. A bone marrow aspiration proved normal.

The diagnosis of Crohn's disease was based on clinical and radiological data, an endoscopic score of 83 upon colonoscopy according to the evaluative method of Pera et al. (5), and colonic biopsies which revealed transmural inflammation accompanied by submucosal thickening, fibrosis and fissures. The disease extended from the anus to the terminal ileum. Stool culture for enteric pathogens and Yersinia serology were negative.

Initial treatment with prednisolone 40 mg daily resulted in both cutaneous and digestive improvement. However, when the dose of prednisolone was halved after 15 days, cutaneous lesions, fever, and diarrhea reoccurred. Dapsone (100 mg daily) was thereupon associated with the steroid without beneficial effects and was discontinued 3 weeks later because of the appearance of a hemolytic anemia. Prednisolone was then again increased to 40 mg daily for 1 month, gradually reduced over a 2-month period, then stopped. The patient's clinical improvement has been maintained until now.

DISCUSSION

The cutaneous febrile illness known as Sweet's syndrome is clinically characterized by tender, dusky red papules, nodules, or plaques and histologically identified by a dense infiltrate of polymorphonuclear leukocytes with leukocytoclasia in an edematous dermis (6). Approximately 10 to 15% of published cases of Sweet's syndrome occur in patients with a malignancy, with nearly half of these involving acute myelogenous leukemia (7). Sweet's syndrome has also been reported in association with Sjögren's syndrome, Behçet's disease, rheumatoid arthritis, and various infections. Only four cases of Sweet's syndrome have thus far been reported in patients with ulcerative colitis (3, 4, 8). This case is the first known association with Crohn's disease and showed a parallel evolution of cutaneous and digestive symptoms. Mucocutaneous complications of Crohn's disease may be related to the primary granulomatous process, nutritional deficiencies, or therapy (1). Most manifestations such as erythema nodosum, erythema multiforme, epidermolysis bullosa acquisita, necrotizing vasculitis, and pyoderma gangrenosum are idiopathic. It is worth noting that Sweet's syndrome and pyoderma gangrenosum have been described in a similar range of diseases including leukemia, myeloma, and inflammatory bowel diseases. Furthermore, they have both been reported successively in the same patient (3). This association, as well as their clinical and histological similarities, have led to the hypothesis of a nosological continuum between Sweet's syndrome and pyoderma gangrenosum based on a similar un-

derlying pathogenesis which needs further elucidation (9, 10).

ADDENDUM IN PROOF

Since submission of this article for publication, KEMMETT et al. reported two other cases in British Medical Journal 1988: 297, 1513–1514.

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Transient Acantholytic Dermatosis Associated with Lymphomatous Angioimmunoblastic Lymphadenopathy

BRIAN D. ZELICKSON,[1] AYALEW TEFFERI,[2] MORIE A. GERTZ,[2] PETER M. BANKS[3] and MARK R. PITTELKOW[1]

[1]*Department of Dermatology,* [2]*Division of Hematology and Internal Medicine, and* [3]*Section of Surgical Pathology, Mayo Clinic and Mayo Foundation, Rochester, Minnesota, USA*

Transient acantholytic dermatosis is a papulovesicular cutaneous eruption first described in 1970. There have been subsequent reports of similar disorders occurring in patients with malignancy. Angioimmunoblastic lymphadenopathy with dysproteinemia is a disorder characterized by an acute onset of generalized lymphadenopathy associated with fever, malaise, pruritus, night sweats, and hepatosplenomegaly. The patient described had a papular acantholytic dermatosis associated with the development of angioimmunoblastic lymphadenopathy with dysproteinemia-like T-cell lymphoma. The cutaneous manifestations of angioimmunoblastic lymphadenopathy with dysproteinemia are discussed. ***Key words: Grover's disease; Acantholysis; Polyclonal hypergammaglobulinemia.***

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M. R. Pittelkow, Department of Dermatology, Mayo Clinic, 200 First Street SW, Rochester, MN 55905, USA.

Several different dermatoses display acantholysis. This loss of coherence between epidermal cells may be a result of primary disruption of the intercellular substance or induced by an alteration in the structural integrity of keratinocytes. Examples of the disruption of intercellular substance include pemphigus, and examples of an alteration of keratinocytes are Hailey-Hailey disease, Darier's disease, and warty dyskeratoma.

In 1970, Grover (1) described six patients with transient pruritic papulovesicles that were mostly localized to the trunk and histologically showed acantholysis. Subsequently, other primary acantholytic disorders with histologic similarities to Grover's disease have been reported. Some of these have been related to malignancy (2, 3), actinic damage (4), or ionizing radiation (5).

The clinicopathologic entity of angioimmunoblastic lymphadenopathy with dysproteinemia (AILD) was first described 15 years ago (6) and recently has been extensively reviewed (7). It is a lymphoproliferative disorder that usually occurs in the elderly and is characterized by an acute onset of generalized lymphadenopathy associated with fever, malaise, pruritus, night sweats, hepatosplenomegaly, and cutaneous eruption. Laboratory abnormalities include an elevat-