Sweet's Syndrome and Non-Hodgkin's Lymphoma:

The First Report of This Association

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A 58-year-old woman developed Sweet's syndrome one week after a flu-like illness. She was later found to have a centrocytic/centroblastic non-Hodgkins lymphoma. Six courses of chemotherapy were given during which the lesions of Sweet's syndrome resolved completely. As far as we are aware this is the first report of the association of Sweet's syndrome with a lymphoma. Key words: Acute febrile neutrophilic dermatosis; Centrocy-

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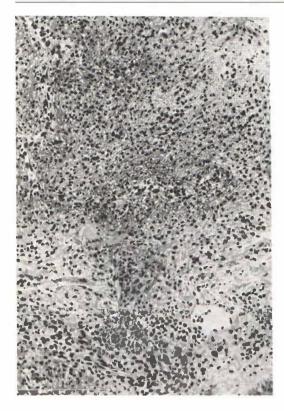


Fig. 1. A photomicrograph of the dermis showing a profuse infiltration with polymorphonuclear neutrophils permeating between collagen bundles. Fragmentation of many polymorphs has occurred and fibrinoid material has been deposited in the region of the blood vessel in the lower part of this picture. Haematoxylin and eosin, × 140.

tic/centroblastic non-Hodgkin's lymphoma; Diffuse mixed lymphocytic histiocytic lymphoma. (Received February 25, 1985.)

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Sweet's syndrome (acute febrile neutrophilic dermatosis) is a well recognised but uncommon condition, which may be associated with a variety of systemic diseases (1). Up to 10% of cases are accompanied by leukaemia which is usually acute myeloid or acute myelomonocytic in type (1, 2, 3). We report here a case in which Sweet's syndrome was associated with a non-Hodgkin's lymphoma. We are not aware of any previously published report of this association in the English literature.

CASE REPORT

A 58-year-old woman was referred to the skin clinic with a six-week history of an eruption on her leg appearing one week after a flu-like illness. She felt otherwise well and the rash had tended to wax and wane. She had noticed occasional reddish discolouration of her urine and her family doctor had discovered a significant depression of her platelet count (81×10⁹/l), but subsequent full blood counts were normal. At the skin clinic, 1 cm diameter infiltrated, erythematous and purpuric nodules were noted, scattered over the lower legs; and slightly enlarged, mobile lymph nodes were felt in the anterior cervical and supraclavicular regions. Dip-stick urinalysis showed moderate haematuria; the ESR was found to be 50 mm in the first hour (Westergren); a biochemical profile and full blood count were normal apart from slight elevation of the plasma alkaline phosphatase concentration (128 units/l, normal range 40-100 units/l) and throat swabs and antistreptolysin O titres were negative. A skin biopsy contained a florid and diffuse derinal infiltrate of mature polymorphonuclear leucocytes

extending from perivascular sites to permeate between collagen bundles. The polymorphs were fragmented with abundant nuclear debris and some small blood vessels showed necrosis of their walls. Fibrinoid material was deposited around some vessels and the infiltrate was separated from the overlying epidermis by a prominent oedematous band (Fig. 1). This picture was consistent with the clincal diagnosis of Sweet's syndrome. Some six weeks after her initial presentation the patient was found to have a palpable spleen and the liver had enlarged to 4 cm below the right costal margin. A chest X-ray showed old changes of pulmonary tuberculosis. A lymphoma was considered likely and two lymph node biopsies were undertaken. Histological examination of the excised lymph nodes showed a diffuse mixed lymphocytic/histiocytic lymphoma (Rappaport) which was centrocytic/centroblastic (Kiel) in character. Six courses of chemotherapy (cyclophosphamide, doxorubicin, vincristine and prednisolone (100 mg daily for five days) repeated every fourth week) were given over the next five months during which her eruption cleared completely. Further chemotherapy has been given for her persisting lymphoma and there has been no relapse of her Sweet's syndrome.

COMMENT

Sweet's syndrome has been associated with a variety of underlying systemic conditions (1, 2, 3). Notable among these are respiratory tract infections (especially those due to Bhaemolytic streptococci) and myeloproliferative disorders (especially acute myeloid or myelomonocytic leukaemia) which may occur in up to 10% of patients with Sweet's syndrome. The syndrome is also associated with immunisation, trauma, ulcerative colitis, Sjögren's syndrome, multiple myeloma, metastatic adenocarcinoma and seminoma (1, 2, 3). It is possible that the preceeding flu-like illness might have precipitated the Sweet's syndrome in this patient but throat swabs and antistreptolysin O titres were negative. Both Sweet's syndrome and non-Hodgkin's lymphom are unusual conditions and occurred here in the same patient. This is the first report of such an association in the English literature.

The clinical and histological similarities between pyoderma gangrenosum and Sweet's syndrome, as well as the association of both conditions with similar underlying disorders, has led to the suggestion that they might represent the two extremes of one neutrophil mediated hypersensitivity reaction (1, 3). However, no adequate pathophysiological explanation for either eruption has been put forward so far.

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