Skin lesions with sinister associations

Introduction

Many serious diseases declare their presence with dermatological features. These include rashes, pigmentation changes, vasculitic lesions and on occasions highly characteristic skin lesions. This article describes a case of Sweet's syndrome which demonstrated typical skin lesions, and discusses the presentation, management and frequent sinister associations of this syndrome.

Discussion

First described in 1964, Sweet's syndrome is an acute neutrophilic dermatosis. The condition is characterized by the appearance of raised red tender skin plaques, occasionally papular, nodular or sometimes bullous. Microscopically there is dense infiltration with neutrophils. Patients feel unwell and may have a fever, headache and usually a neutrophilia. It is most common in women between the ages of 30 and 50 years, but both sexes and all ages including children can be affected.

In some patients no cause is found, but many reports describe associations with inflammatory bowel disease, myelodysplastic conditions (commonly acute myelogenous leukaemia), malignant tumours, certain drugs and pregnancy (Cohen, 1993, 2007).

This patient has the appearance of someone with a malignancy or significant inflammatory process as the underlying cause. The authors were unable to find a solid tumour but several other potential causative associations exist. The paraneoplastic character of Sweet's syndrome results in an association with solid tumours in around 7–20% of cases (Chan et al, 1994; Cohen, 2007).

Myelodysplasia has the most consistent relationship to Sweet's syndrome, in which case the syndrome may be atypical, chronic or recurrent (Vignon-Pennamen et al, 2006). Repeated evaluations become necessary in these patients. Many reports

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Figure 1. Lateral aspect of the left thigh showing multiple circular erythemato-violaceous plaques with central echymotic changes.

Figure 2. Hypothenar eminence of the palmar aspect of the right hand showing a circular purpuric plaque.

Case Report

A 78-year-old Caucasian woman was admitted to hospital with anorexia and weight loss. Six days later she developed facial swelling with multiple tender erythematous raised plaques on the buttocks, thighs (*Figure 1*), hands (*Figure 2*) and face.

The past medical history was complex and included previous stroke, ischaemic heart disease, paroxysmal atrial fibrillation and pulmonary emboli, the latter two requiring long-term warfarin. Twelve months earlier a hemicolectomy had been performed for a vesico-colic fistula of likely diverticular origin. Inflammatory bowel disease could not be excluded as the colonic biopsy had shown non-specific inflammatory changes. The patient had recently been prescribed antibiotics for pneumonia and repeated urinary tract infections. The urinary tract infections were associated with haematuria, albeit while taking warfarin.

Clinical examination revealed no evidence of a tumour, specifically no lymphadenopathy, and normal breasts. A full blood count revealed a normochromic normocytic anaemia (haemoglobin 99 g/litre, white cell count 3.9×10^9/litre, neutrophils 2.6×10^9/litre, platelets 269×10^9/litre).

Ferritin, vitamin B_{12}, folate, complement and immunoglobulin levels were normal. A tumour marker screen and extensive autoimmune screen was unremarkable. The C-reactive protein level was elevated at 149 mg/litre and erythrocyte sedimentation rate was 47 mm/hr.

Abdominal ultrasound and computed tomography scanning of the abdomen and pelvis showed moderate enlargement of the spleen. Renal ultrasound and cystoscopy found no abnormality in the kidneys or bladder. A chest radiograph and computed tomography scan of the neck and thorax showed a 14 mm right apical lung lesion. This and lymph nodes at the right hilum and sub-carinal position were felt to represent previous, now inactive, tuberculosis. A bone marrow trephine and aspiration showed changes of myelodysplasia but normal cytogenetics and immunophenotyping.

Histology of a skin biopsy from the thigh showed a neutrophilic infiltrate of the dermis with considerable leukocytoclasis. A small number of eosinophils and inflammation was seen in the subcutaneous fat. The infiltrate surrounded the small blood vessels. A diagnosis of Sweet's syndrome was made as a result of these skin features. Treatment consisted of a 7-day course of prednisolone 20 mg daily.

Six months later, the patient remained systemically well but had had two further episodes of Sweet's syndrome which had responded to short courses of oral prednisolone.

describe an association with ulcerative colitis and Crohn's disease (Foster et al, 2005).

Drugs reported to be associated with the development of Sweet's syndrome include granulocyte colony-stimulating factor, trans-retinoic acid, hydralazine, frusemide, oral contraceptives, lithium, co-trimoxazole, minocycline and, paradoxically, non-steroidal anti-inflammatory agents.

Of note, Sweet's syndrome associated with malignancy does not show the usual and still unexplained female preponderance (Von den Driesch, 1994).

Corticosteroids are usually the first line of treatment and result in resolution of skin lesions and associated systemic symptoms (Cohen and Kurzrock, 2002). Topical or intra-lesional use of high strength steroids can be used for resistant skin features. Other treatments that have been used include dapsone, colchicine, potassium iodide, indomethacin, naproxen, ciclosporin, methotrexate, metronidazole and more recently thalidomide.

Conclusions

Despite several potential reasons for the occurrence of Sweet's syndrome in this patient, the most likely explanation appears to be myelodysplasia. Continued vigilance is required to determine if these dermatological features represent a paraneoplastic harbinger of an even more serious condition. **BJHM**

Chan HL, Lee YS, Kuo TT (1994) Sweet's syndrome: clinicopathologic study of eleven cases. *Int J Dermatol* **33**(6): 425–32

Cohen PR (1993) Pregnancy-associated Sweet's syndrome: world literature review. *Obstet Gynecol Surv* **48**(8): 584–7

Cohen PR (2007) Neutrophic dermatoses occurring in oncology patients. *Int J Dermatol* **46**(1): 106–11

Cohen PR, Kurzrock R (2002) Sweet's syndrome: a review of current treatment options. *Am J Clin Dermatol* **3**(2): 117–31

Foster EN, Nguyen KK, Sheikh RA, Prindiville TP (2005) Crohn's disease associated with Sweet's syndrome and Sjogren's syndrome treated with infliximab. *Clin Dev Immunol* **12**(2): 145–9

Vignon-Pennamen MD, Juillard C, Rybojad M et al (2006) Chronic recurrent lymphocytic Sweet syndrome as a predictive marker of myelodysplasia; a report of 9 cases. *Arch Dermatol* **142**(9): 1170–6

Von den Driesch P (1994) Sweet's syndrome (acute febrile neutrophilic dermatosis). *J Am Acad Dermatol* **31**(4): 535–56