ACUTE FEBRILE NEUTROPHILIC DERMATOSIS (SWEET'S SYNDROME)

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Two patients had classical clinico-pathological features of Sweet's syndrome. One of them had associated polycythaemia vera, myelofibrosis, mucosal lesions and haemorrhagic blisters. The patient showed a prompt response to corticosteroids. A long term haematological follow up is imperative in all patients.

Key words : Acute febrile neutrophilic dermatosis, Sweet's syndrome.

Acute febrile neutrophilic dermatosis, originally described by Sweet in 1964,[1] consists of characteristic raised, tender, reddish-brown plaques with a peculiar pseudovesicular border and an intense dermal polymorphonuclear infiltrate in biopsy specimens. It is usually associated with fever, malaise, peripheral polymorphonuclear leucocytosis and arthralgia, though it is neither always acute nor always febrile, and may lack leucocytosis.[2,3] Leucocytosis may follow the skin lesions by several days to weeks.[4] Mucosal lesions,[5] conjunctivitis and episcleritis are uncommon.[3] Nearly 80% of the patients are females, and antecedent infectious episodes are recorded in 90% of the patients.[4]

We are reporting two cases of this syndrome.

Case Reports

Case 1

A 57-year-old housewife was an established patient of polycythaemia vera for the last 18 years, treated with radioactive phosphorus (^{32}P) and phlebotomies. She had lately developed myelofibrosis which was confirmed on trephine bone marrow biopsy. The diagnosis of polycythaemia vera had been confirmed by erythrokinetic studies employing ^{51}Cr which showed haemoglobin 20.2 gm%, haematocrit 61%, red cell volume 55.6 ml/kg (Normal upto 32 ml/kg), total blood volume 105 ml/kg (Normal 60-80 ml/kg) and plasma volume 49.4 ml/kg. One month after an episode of mild trauma, flu like symptoms, general malaise and fever, she developed dusky red, raised and tender lesions on the right hand and face. This was followed by painful oral ulcers. She had ruddy cyanosis of skin and mucosae, and hepato-splenomegaly of 10 cm and 24 cm below the costal margin respectively. Multiple, erythematous, elevated and tender plaques of variable sizes with a smooth, uneven surface and central clearing were located on the dorsum of right hand, forearm, face (Fig. 1), ears and legs. The margins of the lesions appeared vesicular, though these were firm upon palpation and did not exude fluid.

Haemoglobin was 14.3 gm% and PCV 55%. The total leucocyte count was 68,200/mm^3 and the thrombocyte count 60,000/mm^3. Differential white cell count showed 66% neutrophils, 4% lymphocytes, 3% eosinophils, 1% basophils, 1% blast cells, 1% promyelocytes, 18% myelocytes and 6% metamyelocytes. Blood urea, creatinine, blood sugar, cholesterol, uric acid, serum proteins, bilirubin and liver enzymes were within the normal range. Urine, stools and skiagrams were normal. Skin biopsy showed uneffected epidermis and a dense, predominantly polymorphonuclear infiltrate in the upper and mid-dermis (Fig. 2). The infiltrate was angiocentric and consisted of neutrophils, nuclear dust, scanty lymphocytes and histiocytes (Fig. 3).

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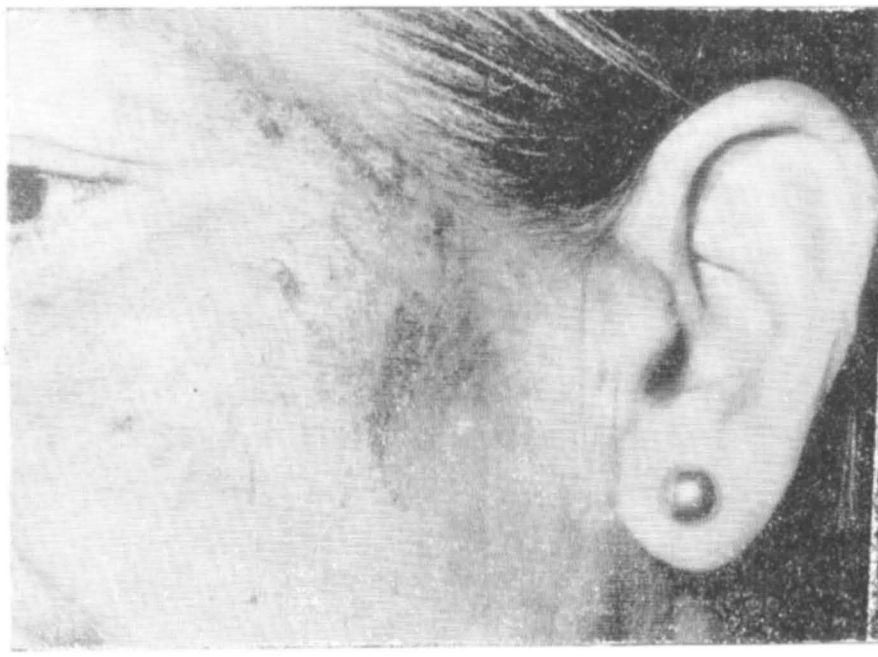

Fig. 1. An erythematous plaque with elevated and pseudo-vesicular margins.

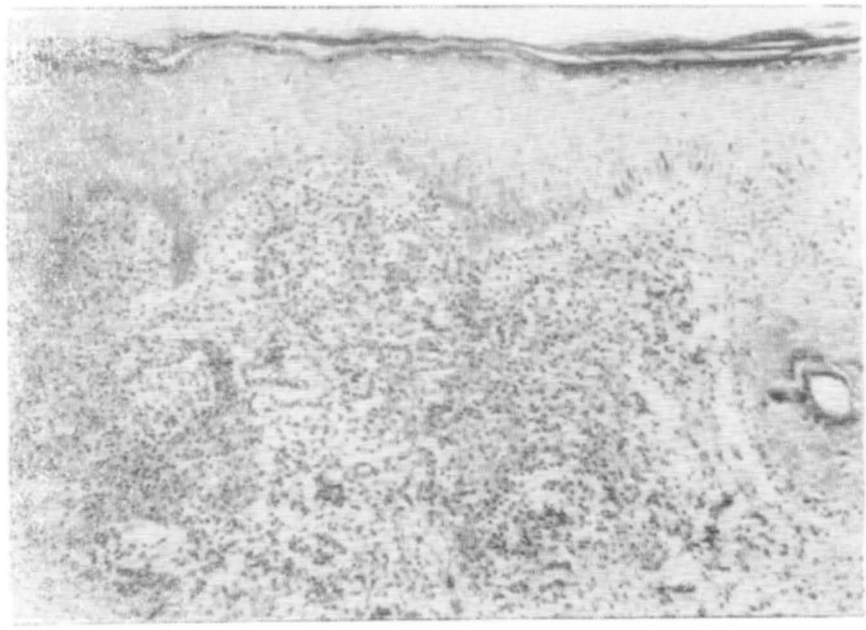

Fig. 2. Dense polymorphonuclear infiltrate in upper and mid-dermis (H & E×140).

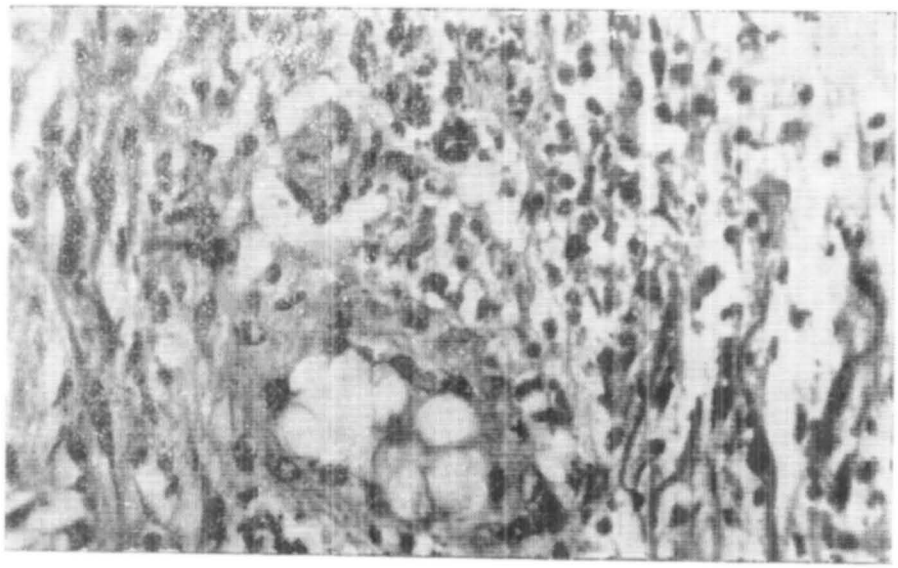

Fig. 3. Undamaged vessel wall showing endothelial swelling, perivascular neutrophilic infiltrate and nuclear dust (H & E×330).

The vessel walls were well preserved but there was a marked endothelial swelling. Direct immunopathology of the biopsy tissue revealed IgA and complement deposits around the dermal vessels. Treatment with 40 mg prednisolone a day led to a prompt regression of the skin lesions within a week. However, haemorrhagic blisters and new lesions appeared on a daily maintenance dose of 5 mg. Presently, she is in complete remission with 15 mg daily prednisolone.

Case 2

A 29-year-old male had an abrupt onset of a slowly increasing, red, tender swelling on the forehead without any prodromal illness, fever, malaise, eye trouble, arthralgia or mucosal lesions. General physical examination was within normal limits. Cutaneous examination revealed a single, shiny, dusky erythematous, raised, tender plaque measuring 8 cm×5 cm in the centre of the forehead. The margins gave an illusion of vesiculation and the surface was uneven, resembling a relief map of mountain range. The total leucocyte count was 16000/mm,^{3} with 76% neutrophils, 20% lymphocytes, 2% monocytes and 2% eosinophils. Other investigations were normal. Histopathological examination of the biopsy specimen showed changes similar to the first patient. Within a few days of biopsy, the lesion started improving on its own and regressed in two weeks without residual scarring.

Comments

The exact etiology of Sweet's syndrome is unknown, frequent preceding infection, self-limiting course, histopathological features of vascular alterations, electron-microscopic evidence of repeated vascular damage and regeneration, prompt response to corticosteroids, arthritis and renal involvement point towards a hypersensitive process, possibly a localised immune complex disease of Arthus type.[4,6] One of the three patients studied earlier[6] revealed perivascular deposits of IgG, IgM and fibrin. Nunzi et al[7] demonstrated the presence of polymorphonuclear cells with membrane-bound

and intracellular IgM and IgE, and a defective chemotactic response of circulating neutrophils.

Apart from fever, malaise and prostration, other systemic features like anaemia, elevated ESR, arthritis, renal and hepatic involvement may also occur.[3,4,6] Of more than a hundred reported cases of Sweet's syndrome,[16] patients with co-existent leukaemia have been reviewed.[8] Such a high incidence of leukaemia following the skin lesions by a few months to two years may justify the classification of Sweet's syndrome as a cutaneous marker of malignancy and a careful haematological follow up is recommended in all cases.[8] Incidently, our case 1 is the only reported case of polycythaemia vera associated witn Sweet's syndrome. Tikjob et al[9] reported a patient with an abnormal chromosomal pattern in the bone-marrow aspirate who developed acute myeloid leukaemia six months later. He recommended chromosomal analysis as a mandatory procedure in every patient. Pyoderma gangrenosum, ulcerative colitis, and benign monoclonal gammopathy are other reported associations.[6]

Typical cases are easy to diagnose, though certain atypical cases may show clinicopathological overlap with pyoderma gangrenosum.[10] Sweet's syndrome and pyoderma gangrenosum may represent two ends of a nosological continuum with a similar underlying pathogenesis.[10] Corticosteroids and potassium iodide result in complete resolution.[6] Untreated, the skin lesions may persist for a few weeks to a few months and recurrences are common.[6]

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