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CASE REPORTS

ACUTE FEBRILE NEUTROPHILIC DERMATOSIS (SWEET'S SYNDROME)

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A case of acute febrile neutrophilic dermatosis (Sweet's syndrome) is presented. The lesions responded dramatically to corticosteroids.

Key words : Sweet's syndrom', Corticosteroids.

Acute febrile neutrophilic dermatosis (AFND) was first described by Sweet[1] in 1964. He observed eight patients with painful, erythematous plaques involving the limbs, face and neck. A skin biopsy specimen showed a dense dermal infiltrate with neutrophils. Other important features of the disorder included fever and a polymorphonuclear leucocytosis. Since the time of Sweet's original description, some more cases have been reported in the world literature.[2-4] In the present communication, we report a case of Sweet's syndrome recently seen by us. This is the first report of this rare disorder from Libyan Arab Jamahiriya.

Case Report

A 40-year-old female patient developed an eruption of approximately 20 days duration affecting the forearms and legs. In addition to the skin lesions, the patient had noted arthralgias involving the wrists, generalized malaise and intermittent temperature of upto 39° C. Two weeks prior to the onset of skin lesions, the patient had an attack of upper respiratory tract infection. There was no history of any drug intake. However, she had had similar lesions twice during the past 2 years. She was found to be febrile. Skin showed annular, dull red and plum coloured plaques on her forearms and extensors of the legs. The lesions were studded with many vesicles and were warm and tender to palpation. There was a slight swelling and tenderness of the wrists bilaterally. The rest of the physical examination was normal.

Laboratory studies revealed ESR, 58 mm; hemoglobin, 12.9 gm/dl; TLC 13,800/c mm with 64% polymorphonuclear leucocytes, 30% lymphocytes, 5% eosinophils and 1% basophils.

Serum electrolytes, ASLO titres, liver function tests, VDRL, LE cells, blood sugar and rheumatoid factor were either negative or normal. Bacterial cultures of the skin lesions showed no growth and roentgenograms of the chest were normal.

Skin biopsy obtained from a plaque on the forearm showed a heavy perivascular cellular infiltrate composed mainly of neutrophils with some lymphocytes. There were a few areas of leucocytoclasis, but there was no evidence of vascular wall necrosis or deposition of fibrinoid material.

She was administered 30 mg of prednisolone a day along with topical betamethasone valerate cream. The lesions gradually disappeared over a period of two weeks and the corticosteroids were tapered off. Follow up for six months has not shown any recurrence.

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Comments

Our patient demonstrates the major features of AFND, i.e. painful erythematous plaques, fever, and polymorphonuclear leucocytosis. Histopathologically, the lesions demonstrated a dense, dermal infiltrate of neutrophils and lymphocytes, without true vasculitis, which is compatible with AFND.[5]

Other important features demonstrated by this patient included conjunctivitis, arthralgias and an elevated ESR. Importance of arthralgias has been emphasized by other authors also. Gunawardena et al[6] reported 18 patients of which 9 had notable joint symptoms and all but one had an elevated ESR. Thirteen of the 18 patients were also noted to have conjunctivitis or episcleritis at the time of examination.

The diagnosis of AFND should be considered in patients who present with a rather sudden onset of raised, plum-coloured, painful plaques on the limbs, face or neck and especially if accompanied by fever and arthralgias. It is likely that many cases are misdiagnosed especially in non-febrile cases.[7] Erythema elevatum diutinum and, on the face, eosinophilic granulomas, are excluded by the course and histopathology. Atypical forms of erythema multiforme may pose clinical difficulties, though, histopathology is helpful. Most of the reported cases in the literature have been middle aged women.[7] Our patient is also a female.

The cause and pathogenesis of AFND remains obscure.[5] In the original description, Sweet[1] believed that a reactive process was involved. Many of his original patients had had a preceding minor illness before the onset of the eruption. Shapiro et al[8] hypothesized that AFND was perhaps some type of hypersensitivity reaction to an infectious agent. More than half of the described patients had had febrile illness, usually an upper respiratory tract infection.[1] In our case also there was a history of preceding upper respiratory tract infection. Sweet's syndrome has also been described after acute myelogenous leukemia,[9] testicular tumours,[8] tuberculin skin tests[4] and cellulitis following vaccinations.[6] However, the dramatic response of the disorder to systemic corticosteroids and association of joint, eye and cutaneous involvement suggest an underlying immunologic disturbance.

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