SWEET'S SYNDROME

Subhash C Bharija, Gurmohan Singh

A series of 10 cases of Sweet's syndrome observed during a period of 3 years in Eastern parts of Libya is reported.All the patients were females.Fever and peripheral neutrophilia were not found in all these cases.Dapsone was found to be very effective in the cases in which it was tried.

Key Words: Sweet's syndrome, Fever, Neutrophilia, Dapsone

Introduction

Sweet's syndrome is a rare disorder first described by Sweet in 1964.[1] Perhaps less than 200 cases have been reported in the world literature. We report a study of 10 cases of this syndrome seen over a period of 3 years in Benghazi, Libya.

Patients and Methods

All the patients with a provisional diagnosis ofSweet's syndrome were admitted to the skin words of Jamahiriya Hospital Benghazi, from 1986 to 1989. The diagnosis was confirmed by histopathology. Total and differential counts, palatelet count, erythrocyte sedimentation rate, liver function tests, renal function tests, throat swab culture and X-ray chest were dpme. The cases inwhich the diagnosis was confirmed by histopathology,were assigned to various treatments including 20-60 mg prednisolone daily (6 cases), dapsone 200 mg day (2 cases), and nonstertoidal anti-inflammatory drugs (2 cases). Progress of each case was recorded reglarly.

Results and Discussion

Sweet's syndrome is said to be rare. about 200 cases having been reported in quorter of a century. It may be due to the strict adherence to four cardinal features for its diagnosis as originally described by Sweet in 1964 which included fever, followed by painful plaques on the skin, neutrophilia in the peripheral blood and dense neutrphilic tissue infilteration. We observed 10 cases in a period of three years from amongst a small community of Benghazi, Libya. We used two most important cardinal features for its diagnosis viz; painful erythematous plaques on the extremeties or face and its histological confirmation.

Though majority of the cases in the first report of Sweet were preceeded by infection[1] and 85-90% of the patients have been reported to have high persistent faver,[2] yet only one of our 10 cases had fever.We consider that it is not an essential or consistent feature of this entity.

Majority of the patients have been reported to have leukocytosis with a high neutrphilia. In our study only three patients had leukocytosis and two were at borderline levels in their leukocyte counts. Neutrophils ranged from 58-78%. Thrombocytosis (557,000/cmm) was seen in one patient. two of the patients had over 400,000 platelet counts. Although, thrombocytosis has not been reported in general, yet a child with Sweet's syndrome was reported having thrombocytosis and he died of coronary artery disease.[3] Elevated erythrocyte

From the Department of Dermatology, Al Arab Medical University, Benghazi, Libya.

Address correspondence to : Dr S C Bharija 71/43, Prem Nagar, New Delhi - 110058, India.

sedimentation rate upto 80 mm in 1st hour, is a common feature.[4] All our patients showed elevated ESR ranging from 50 to 106 mm in 1st hour.

Our patients first reported for treatment from 4 to 15 days after the onset of the disease.All the patients were females ranging in age from 32 to 50 years (mean age 40.4 years).

The sites involved were hands only (3 cases), forearms (2 cases), and the other three had involvement of all the above 3 sites. None of our patients had involvement of face and neck.The extensors were more involved and the distribution of the lesions was asymmetrical in all the cases. In half of the cases the lesions assumed annular appearance within 2-3 days.There was no evidence of either joint or eye involvement. Liver function tests and renal function tests were within normal limits.

The treatment with systemic corticosteroids, dapsone and other anti-inflammatory drugs was effective. Lesions disappeared in a period of 8 to 16 days after instituting steroid therapy (mean 9.5 days) and it took 6-7 days to clear the lesions with dapsone therapy (mean 6.5 days). Dapsone inhibits neutrophilic chemotaxis and its effect may be more marked than prednisolone as evidenced from this limited observation.

Sweet's syndrome does not appear to be as rare as it seems though a full blown classical picture may not be observed in each case. Raised, erythematous and painful plaques, particularly if they are asymmetrical, especially in females, should arouse the suspicion of this entity. We tried dapsone in 2 patients and found it to be an effective therapy.

References

1. Sweet RD. An acute febrile neutrophilic dermatosis. Br J Dermatol 1964; 76: 349-56.
2. Shapiro L, Baraf CS, Richheimer LL. Sweet's syndrome (acute febrile neutrophilic dermatosis). Arch Dermatol 1971; 103: 81-4.
3. Muster AJ, Bharati S, Herman JJ, et al. Fatal cardiovascular disease and cutis laxa following acute befrile neutrophilic dermatosis. J Pediatr 1983; 102: 243-8.
4. Levin DL, Esterly NB, Herman JJ, Boxall LBH.The Sweet syndrome in children. J Pediatr 1981; 99: 73-8.