Sweet's syndrome-A case report

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รายงานผู้ป่วย Sweet's syndrome

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กลุ่มอาการสวีทเป็นโรคทางผิวหนังที่ไม่ทราบสาเหตุ และพบได้ไม่บ่อยนัก อาการสำคัญประกอบด้วยผื่นนูนอักเสบที่ผิวหนังที่เกิดขึ้นอย่างเฉียบพลัน ร่วมกับอาการไข้, ปวดข้อและมีจำนวนเม็ดเลือดขาวเพิ่มสูงขึ้นในกระแสโลหิต กลุ่มอาการนี้มีรายงานว่าพบร่วมกับโรคของระบบอื่นหลายโรค ผู้เขียนได้เสนอรายงานผู้ป่วยชาย 1 ราย ที่มีอาการและอาการแสดงของกลุ่มอาการสวีท ซึ่งพบร่วมกับโรคหนองในเยื่อหุ้มปอดจากเชื้อซัลโมเนลลา และวัณโรคของต่อมน้ำเหลืองในผู้ป่วยคนเดียวกัน ได้วิจารณ์ถึงวิธีการวินิจฉัย ลักษณะการดำเนินของโรค และผลการรักษาไว้ในรายละเอียด

Acute febrile neutrophilic dermatosis (Sweet's syndrome) is an uncommon, recurrent skin disease of unknown etiology characterized by painful inflammatory plagues with fever, arthralgia and leucocytosis. Many associated systemic diseases were reported. In this report we described a classical case of Sweet's syndrome in association with Salmonella empyema and tuberculous lymphadenitis. Diagnostic approaches, clinical course and outcome were discussed in detail.

INTRODUCTION

Acute febrile neutrophilic dermatosis (Sweet's syndrome) is an uncommon, recurrent skin disease characterized by painful plague forming and inflammatory papules associated with fever, arthralgia and peripheral leucocytosis[15]. It was first described by Sweet in 1964. The cause of this condition is unknown, but massive tissue leukocytosis strongly implies a pathogenic role of leucotactic mechanisms. Many systemic diseases such as ulcerative colitis[1,13,4,15], leukemic diseases[8,9,14,15] and various malignant tumors[7,12] were reported in association with this disease. In this report we would like to describe a classical case of Sweet's syndrome in association with another unusual systemic diseases.

CASE REPORT

A 50 year-old male patient presented with acute high grade fever and simultaneous eruption of painful skin lesions at trunk, hands and feet for 2 days. He was a known case of Salmonella B (typhimurium) empyema and tuberculous lymphadenitis and had been treated with trimethoprim/sulphamethoxazole and anti-tuberculous drugs about 1 month before the skin eruptions.

Physical examination revealed an acutely ill middle age man with high fever. There were many painful erythematous and edematous plagues and papules studded with tiny pustules on the surface over the chest wall (fig. 1), back (fig. 2) and extremities (fig. 3). Multiple small firm, non tender lymph nodes were found at cervical regions. There was decreased breath sound at right lower lung field, other systemic examination was within normal limit. Laboratory investigation consisted of CBC: Hct 34%, WC 22,300/mm, neutropohil 87%, lymphocyte 10%, monocyte 3%, and normal platelets; urine analysis was within normal limit; chest radiography revealed pulmonary infiltration at right lower lung field with calcified pleura. Pus from the skin lesion was negative for Ziehl-Neelsen and gram stain, neither was the culture.

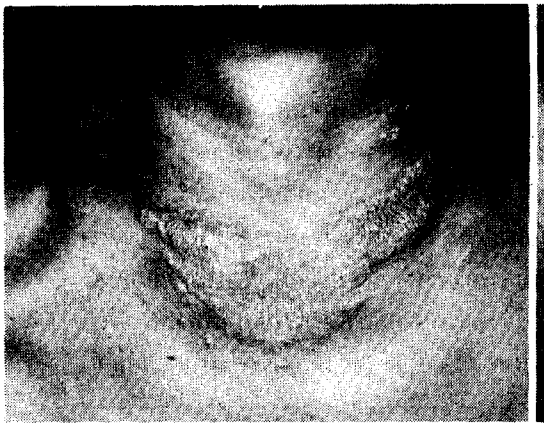

Fig. 1 a. Erythematous and edematous painful plague around the neck

Fig. 1 b. The same lesion 10 days after treatment

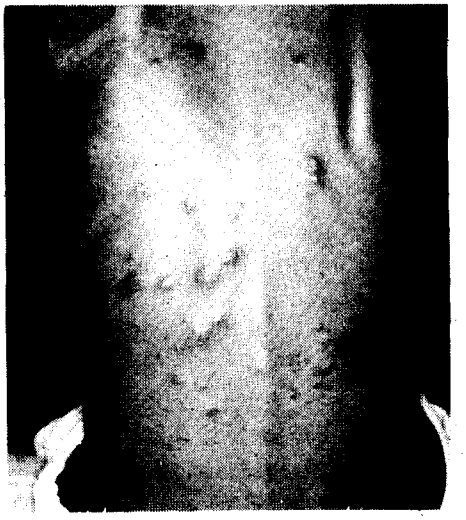

Fig. 2 a. Erythematous tender nodules at the back

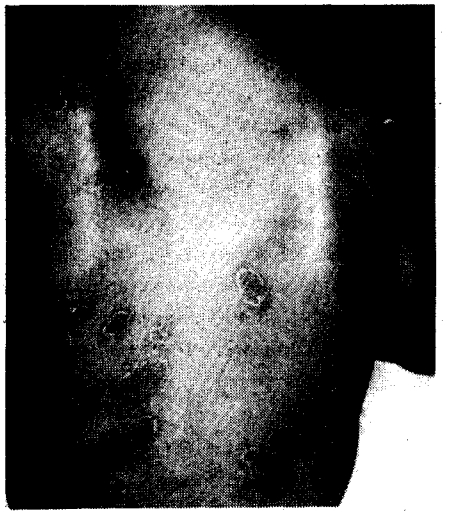

Fig. 2 b. The same area 10 days after treatment

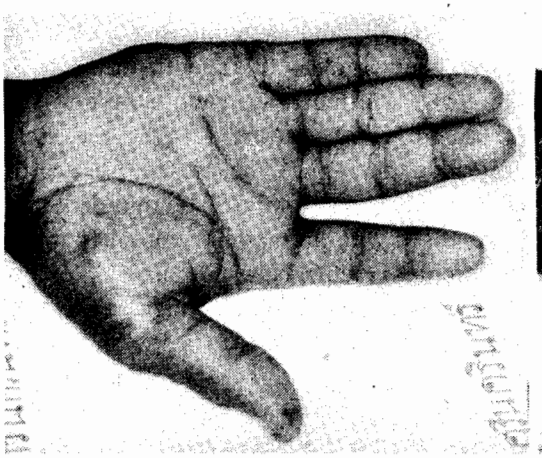

Fig. 3 a. Erythematous tender nodules at the hand

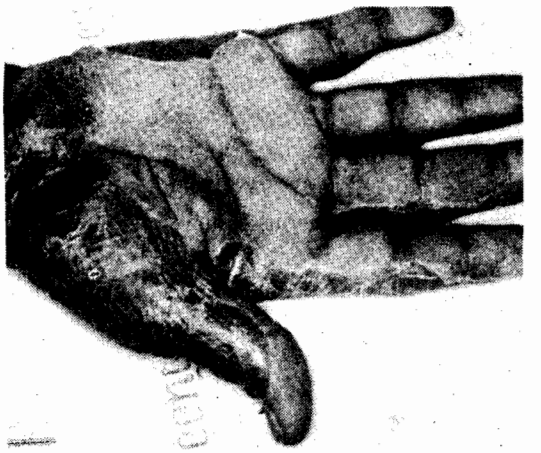

Fig 3 b. The same area 10 days after treatment

The histologic examination taken from the skin lesion at the patient's back revealed dense perivascular infiltration composed largely of neutrophils (fig. 4 and fig. 5). In addition, there were some mononuclear cells such as lymphocytes and histiocytes and a few eosinophils. The capillary walls showed no evidence of vasculitis. Subcutaneous fat was unremarkable. Consequently, acute febrile neutrophilic dermatosis (Sweet's syndrome) was diagnosed.

Fig. 4 Histologic section from the skin lesion showed deuse dermal infiltration of inflammatory cell. (Hematoxylineosin stain, orginal magnifications)

Fig. 5 Higher majuification of Fig. 4 showed that the cellular infiltration composed of polymorph with lencocytoclasia admixed with lympho-histiocyte. (Hematoxylin-eosin stain, original magnifications)

The patient was treated with oral prednisolone 60 mg/day, within 24 hours the lesions was dramatically response then the dosage was reduced to 30 mg/d. Treatment for empyema thoracis and tuberculous lymphadenitis was continued. The dosage of prednisolone was slowly reduced. Eventually the treatment was discontinued at the end of three months period without any clinical relapse.

DISCUSSION

This patient characterized the clinical features of Sweet's syndrome which composed of (1) fever, (2) neutrophilia, (3) raised painful plagues on the trunks and limbs, and (4) a dense dermal infiltration with mature neutrophils seen histologically[14]. Other associated features of this syndrome are arthritis and/or arthralgia [3], myalgia, conjunctivitis and/or episcleritis [3], renal and hepatic involvement [14]. In this patient myalgia was the only associated symptom. The majority of patients had upper[3,4,15] and lower respiratory tract infection (1) preceded the skin lesions by 1 to 3 weeks. Tuberculosis has not been shown to be associated with this syndrome. Despite exhaustive investigation in patients with lower respiratory tract infection, Sweet failed to demoustrate any tuberculosis in his series[1]. Our patient had tuberculous lymphadenitis togather with salmonella empyema which was unusal in normal host, this may indicated that there might be some other underlying immunological disturbance in this patient, even if tuberculin test have been normal. However, Storer (14) found that other immunologic studies such as quantitative T and B-cell studies, lymphocyte transformation and neutrophil studies fail to reveal any abnormality. The associated diseases which had been previously reported were ulcerative colitis[1,9,13], acute leukemia[5,8,9], pyoderma gangrenosum[13], ovarian carcinoma[12], and testicular carcinoma[6]. However, in view of the small number of reported cases associated with malignant diseases, it would be premature to consider Sweet's syndrome as a paraneoplastic condition. Nevertheless we should cautiously look for the associated malignant disease in this patient in long term follow up.

Anti-inflammatory agent such as aspirin and indomethacin[14], topical steroids [14], potassium iodide (900 mg daily for 2 weeks)[15], colchicine (15 mg daily for 7 days with gradual reduction to 0.5 mg daily over 3 weeks)[11], and systemic corticosteroids have been used as therapeutic adjuncts in Sweet's syndrome. Systemic steroid is the most frequently reported to be the most effective therapy. The dosage of corticosteroids prednisolone 30-60 mg. daily has been used with dramatic response. In order to suppress recurrences, contiunation of low doses prednisolone (10-30 mg/day) over a period of a few weeks to a few months is recommended.

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