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A Case of Sweet's Syndrome in Patient with Dermatomyositis

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Sweet's syndrome (SS) has been reported as an association with malignant neoplasms[1] and autoimmune diseases, e.g., Behçet's disease[2], Sjogren's syndrome[3], and rheumatoid arthritis[4]. But dermatomyositis (DM), one of the rare autoimmune diseases, was not reported as an associated disease of SS. We describe an interesting case of SS associated with DM. Diagnosis was made by skin biopsy, and subsequent clinical resolution occurred after institution of prednisolone.

Key Words : *Sweet's syndrome, Dermatomyositis*

INTRODUCTION

Sweet's syndrome (SS) is an acute febrile neutrophilic dermatosis characterized by fever, leukocytosis, tender cutaneous plaques or nodules and dense infiltration by neutrophils. Some cases have been reported as an association with malignant neoplasms[1] and autoimmune diseases, e.g., Behçet's disease[2], Sjogren's syndrome[3] and rheumatoid arthritis[4].

Dermatomyositis (DM) is an inflammatory muscular diseases of unknown cause. Many lines of evidence suggest that both cellular and humoral mechanisms play a role in the pathogenesis of dermatomyositis[5, 6]. But dermatomyositis, one of the rare autoimmune diseases, was not reported as an associated disease of SS.

We describe an interesting case of SS associated with DM. Diagnosis was made by skin biopsy, and subsequent clinical resolution occurred after institution of prednisolone. It is necessary to be concerned that in patients with several autoimmune diseases, including DM, SS is present as an associated finding.

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CASE REPORT

A 57-year-old man with known DM was admitted to our hospital due to an 8-day history of fever, chills and painful cutaneous lesions on neck, left arm and trunk. The fever did not respond to antibiotics and non-steroidal antiinflammatory drugs. He denied having any other recent illness or symptoms preceding this acute episode of symptoms. On admission, he appeared acutely ill and his body temperature was 39.7℃, blood pressure 120/70 mm Hg, pulse rate 100/min, respiratory rate 25/min. On physical examination, he was febrile, alert, and well-oriented. He exhibited numerous erythematous, indurated, slightly tender plaques ranging in size from 0.7 to 6 cm, located on the posterior neck, left arm and trunk. Vesicles and/or pustules were present on the surface of some plaques (Fig. 1). There were no petechiae, purpura or ulcerated or necrotic area, and the lesions did not blanch with pressure. Oral cavity and ophthalmologic examination was done, and revealed no abnormalities.

About 12 months ago, he was admitted to our hospital due to fever and proximal muscle weakness of both extremities, erythematous/violaceous rash over the eyelids and erythematous, scaling rash over the knuckles of both hands. Laboratory test revealed elevated muscle enzymes: AST 95 IU/l (normal value : NV 5-40 IU/l), ALT 112 IU/l (NV 5-35 IU/l), CK 259 IU/l (NV 32-187 IU/l), LDH 649 IU/l (NV 218-472 IU/l), aldolase 37.9 U/ml (NV 2.0-8.0 U/ml). Antinuclear antibody was present in a

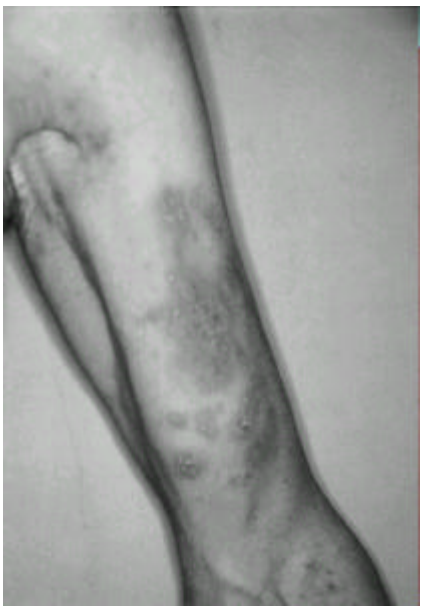

Fig. 1. Sharply demarcated, multiple, erythematous plaques on the left arm. Vesicles and/or pustules were present on the surface of some plaques.

homogenous pattern in a titer of 1:80. The Westergren ESR and cold reactive protein (CRP) was elevated.

The patient's symptoms and laboratory results were suggestive of inflammatory myopathy and electromyographic evaluation (EMG) and muscle biopsy were done. EMG showed typical findings of idiopathic myopathy, irritability of myofibrils (fibrillation potentials) on needle insertion and, at rest and short duration, low amplitude, polyphasic potentials on contraction. Histologic findings of muscle biopsy showed an area of degeneration and necrosis of myofibers in association with interstitial lymphocytic and histiocytic cellular infiltration (Fig. 2). Chest roentgenogram showed fine reticulonodular infiltration on both lower lungs and a biopsy of the lung revealed usual interstitial pneumonia. The upper gastrointestinal endoscopy and a barium enema examination for underlying neoplasm of DM were all normal.

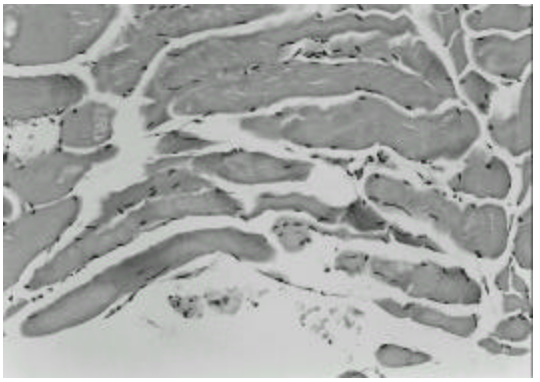

Fig. 2. Histologic findings of muscle biopsy showed an area of degeneration and necrosis of myofibers in association with interstitial lymphocytic and histiocytic cellular infiltration.

Because of the relation of the findings to the criteria of DM, he was diagnosed as DM. High dose of steroid (prednisolone 1 mg/kg/day) was started. Proximal muscle weakness was slowly improved and abnormal laboratory findings were normalized, so the dose of steroid tapered progressively. At a dose of prednisolone 30 mg/day, he was discharged and followed up. From 3 months ago, the daily prednisolone dose was tapered to 5 mg/day by himself. Recently, he stopped medication, and the above-mentioned skin lesion and fever developed.

Laboratory investigations revealed an elevated white blood cell count of 28,200/mm^3 with 83% polymorphonuclear leukocytes, 3% bands, 8% lymphocytes, 3% monocytes and 3% eosinophils, a platelet count 441,000/mm^3. The Westergren ESR was 100 mm/hr and cold reactive protein (CRP) 127 mg/l (NV: < 5 mg/l). The serum urea nitrogen, creatinine, amylase, bilirubin, uric acid and alkaline phosphatase and uric acid were all normal. The levels of muscle enzymes showed as follows: AST 56 IU/l, ALT 69 IU/l , CK 99 IU/l, LDH 349 IU/l, aldolase 17.9 U/ml. Antinuclear antibody was present in a titer of 1:40. Cryoglobulin, C3 and C4 complement levels, and hepatitis B surface antigen and antibody were all normal or negative. The urinalysis did not show any abnormal findings. Blood and urine cultures were negative.

Histologic examination of a biopsy specimen of one of the cutaneous plaques revealed dermal edema and perivascular infiltration, consisting predominantly of neutrophils and some small number of lymphocytes (Fig. 3). But blood vessels had not findings of vasculitis. So, he was diagnosed as Sweet's syndrome and flare-up of dermatomyositis. Prednisolone 50mg/day was instituted and a dramatic improvement of the skin lesions without scarring appeared within 2 weeks. Muscular weakness slowly improved with the above dose of steroid, and prednisolone was tapered. He remains without new skin

lesions, fever and other symptoms four months later, while receiving tapering doses of steroids.

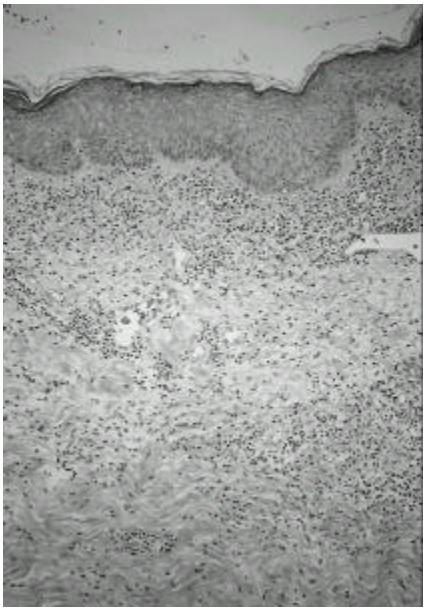

Fig. 3. Histologic examination of cutaneous plaques revealed dermal edema and perivascular infiltration, consisting predominantly of neutrophils and some small number of lymphocytes.

DISCUSSION

Sweet's syndrome was first described in 1964 by Sweet[7], and several reports appeared. It is characterized by fever, neutrophilic leukocytosis and painful, erythematous cutaneous plaque with dense dermal infiltration with neutrophils. The cause of SS is still unknown, but hypersensitivity reactions to bacterial, tumor, autoantigen were suggested as may being involved in the pathogenesis of SS[8]. SS has been noted as usually occuring in middle-aged women and often presents within two weeks of an antecedent respiratory traction infection[7, 9].

Conditions underlying SS have included malignancies and autoimmune diseases. Malignant neoplasms have been reported in approximately 10-20 % of the reported cases of SS[8]. Myeloproliferative disorders, including acute myelogenous leukemia, chronic myelogenous leukemia and multiple myeloma are the most commonly associated malignancing[8]. In addition to association with malignant neoplasms, SS has been reported in autoimmune diseases, such as Behçet's disease[2], Sjogren's syndrome[3], rheumatoid arthritis[4], Reiter's syndrome[10], subacute cutaneous lupus erythematosus[11], drug-induced lupus erythematosus[12], ANA-negative lupus-like syndrome[13] and undifferentiated connective tissue disease[14]. In a search of the literature, we did not discover any other cases of these two diseases occurring together. Our patient is the first case of SS associated with DM.

The skin lesions in DM may be a malar-like rash of the face which involves the nasolabial area (an area often spared in systemic lupus erythematosus) and erythematous/ violaceous rash over the eyelids and erythematous and scaling rash over the knuckles and dorsum of the hand. When typical, the skin lesions of DM are often virtually pathognomonic of this disease. Skin biopsy of early, active lesions of DM showed epidermal atrophy, liquefaction and degeneration of the basal cell layer, perivascular infiltration of lymphocytes and histiocytes in the upper dermis and a striking vasculopathy of small vessels[15]. The patient exhibited herein did not show evidence of the above findings and vasculitis on skin biopsy. Histologic findings and distribution of skin lesions of SS are clearly distinct from those of DM. Also, the clinical course of skin lesions in SS is different to that of DM. In SS, the skin lesions are usually recovered within several days after the institution of steroid therapy. But cutaneous rash in dermatomyositis is a more indolent course with steroid therapy.

The relationship between SS and DM may be proposed as several kinds: first, SS is an acute prodrome phase of flare-up of DM; second, the two diseases are different and occur together by chance; third, SS can be associated with several underlying diseases, of which DM and malignancy are one disease. We did not hold any one of the above three opinions. The association of malignancies and DM is well known, so the possibility of association between SS and underlying malignancy of DM may be suspected. But initial evaluations for malignant underlying DM are negative and the patient has not any evidence of malignancy during 12 months duration of DM. The onset of SS coincided with the flare-up of DM. Aggravation of muscle weakness and elevation of muscle enzymes may be evidence of flare-up of DM. This may be indirect evidence that SS and DM may be associated in this patient. In the light of

immunoregulatory abnormalities and immune complex phenomena and disease onset, consistent with precipitation by viral and bacterial infections in DM, the association of SS and DM reported further supports the hypothesis that hypersensitivity or immunological abnormality is involved in the pathogenesis of SS. Further concern about SS as an associating disease of several autoimmune diseases, including DM, is needed.

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