Sweet's Syndrome Associated with Acute Erythema Nodosum

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A 44-year-old man had Sweet's syndrome (acute febrile neutrophilic dermatosis), accompanied by erythematous tender subcutaneous nodules resembling erythema nodosum(EN). The EN-like lesions histologically showed a septal panniculitis with predominantly neutrophilic infiltrates. The association of Sweet's syndrome with EN seems to be uncommon and only a few cases have been reported until the present. We describe a patient with Sweet's syndrome associated with acute EN. (Ann Dermatol 10:(3) 208~211, 1998).

Key Words : Sweet's syndrome, Erythema nodosum

Sweet's syndrome is an uncommon, recurrent, fairly dramatic skin disease characterized by the abrupt onset of pyrexia, neutrophilia, and typical skin lesions of rapidly extending, tender, erythematous, painful, elevated papules and plaques appearing on the face, neck, and extremities. The cause of Sweet's syndrome is unknown, but it is considered to be a reactive dermatosis. There have been several reports of Sweet's syndrome in association with other disorders, such as inflammatory bowel diseases[1,2], autoimmune diseases[3-5], and various malignant tumors[6,7]. Although EN is also considered to be a reactive dermatosis, the appearance of Sweet's syndrome and EN in the same person is known to be rare. The association of Sweet's syndrome with EN has been reported in only 12 patients with biopsy confirmed cutaneous lesions of both dermatoses[8-19]. We report the first case of Sweet's syndrome associated with EN in Korea.

CASE REPORT

A 44-year-old man had erythematous, tender papules and plaques on the upper and lower extremities, and trunk (Fig.1-A), and erythematous, tender, subcutaneous nodules on both legs (fig.1-B). These symptoms had been present for 15 days. His temperature was 38.9℃ and he felt polyarthralgia. His medical history revealed that he had a total gastrectomy due to stomach cancer 14 years ago. A total leukocyte count was 25,800/mm^3 with 85% segmented neutrphil. A whole body bone scan revealed an abnormal hot uptake on the multiple articular joints but no metastatic adenocarcinoma. A skin biopsy from the back and dorsum of the hand showed the typical findings of Sweet's syndrome (Fig.2-A,B) and one from a subcutaneous nodule on the leg showed neutrophilic septal panniculitis (Fig.3-A,B). With the treatment of systemic corticosteroids, the high fever and skin lesions disappeared rapidly but recurred after decreasing the dosage of the drugs. In spite of the steroid and potassium iodide treatment for a period of 14 months, all the symptoms of polyarthralgia, mild fever, and neutrophilic leukocytosis have continued to persist apart from the skin lesions.

DISCUSSION

Sweet's syndrome, first described by Sweet in the early 1960s[20], is characterized by painful, erythematous papules and plaques on the face, neck and

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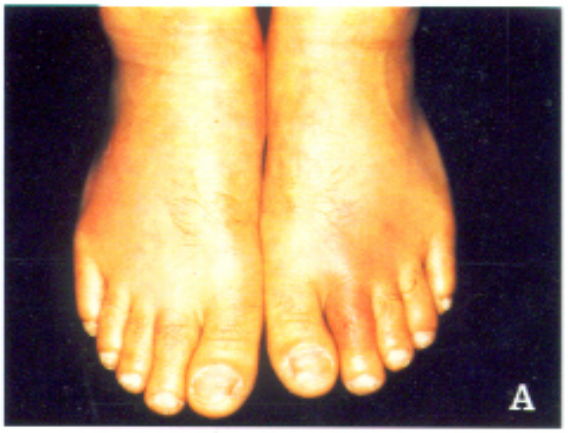

Fig. 1-A. Erythematous, tender papules and plaques on lower extremities.

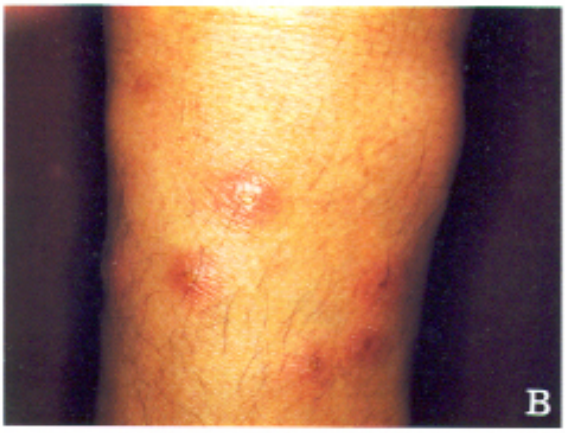

Fig. 1-B. Erythematous, tender, subcutaneous nodules on the dorsum of both feet.

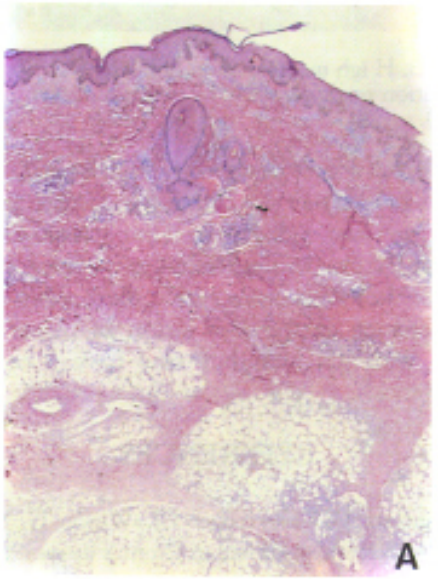

Fig. 2-A. Dense inflammatory infiltration in the upper and middermis (H-E stain : ×40).

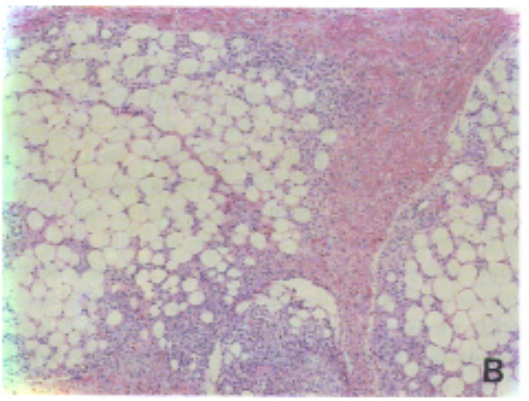

Fig. 2-B. High magnification of Figure 2-A. dermal inflammation predominantly by neutrophils and marked edema of upper dermis(H-E stain ; ×200).

extremities in an asymmetric distribution. The eruptions may be accompanied by arthralgia, general malaise and headache. There is fever and severe leukocytosis, with a high percentage of neutrophils. Histopathologically, the lesions reveal a perivascular infiltrate predominantly of neutrophils in the upper and mid dermis, but there is no true evidence of vasculitis.

An acute form of EN is characterized by tender, erythematous, subcutaneous nodules usually confined to the anterior surface of the lower extremities. The typical histological picture in early lesions of EN consists of septal inflammation with lymphohistiocytite infiltration. Neutrophils rarely predominate the infiltrate[21].

Unusually patients with Sweet's syndrome have been described as having skin lesions involving the subcutaneous tissue[19,22-29]. These patients had a diffuse neutrophilic infiltration of lobules of subcutaneous fat similar to that usually found in the dermis in Sweet's syndrome[19,22-28]. In some cases both the fat and dermis were diffusely infiltrated[29]. Blaustein et al[24] suggested that involvement of the panniculus in Sweet's syndrome occurs in two different patterns ; a neutrophilic lobular panniculitis or a septal granulomatous panniculitis.

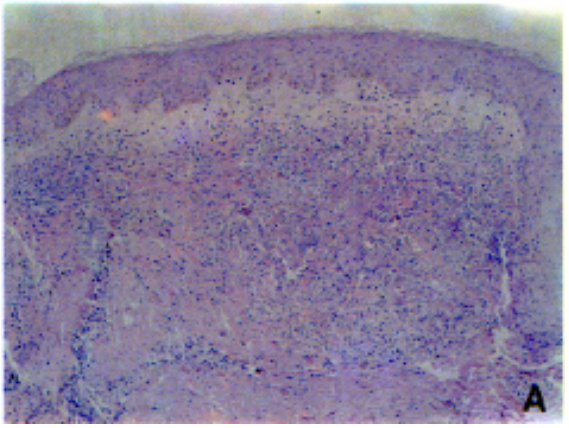

Fig. 3-A. Inflammatory infiltrations limited to the septal portion of subcutaneous tissue (H-E stain : ×40).

However, in our patient with Sweet's syndrome, the EN-like lesions in the lower extremities showed neutrophilic septal panniculitis histologically. We think that the neutrophilic septal panniculitis in our patient represents an EN associated with Sweet's syndrome rather than an involvement of panniculus by Sweet's syndrome. Cohen et al[6] compared the reported cases of the concurrent occurrence of Sweet's syndrome and EN with the same patient, whose case showed neutrophilic septal panniculitis in EN lesions. Until now, only 12 cases with biopsy confirmed cutaneous lesions of both dermatoses in the same person have been reported[8-19], and our case is the first case in Korea.

It is well known that Sweet's syndrome is associated with several diseases, especially leukemia of the myelogenous type. Although Sweet's syndrome and erythema nodosum are associated with several common disorders such as upper respiratory infections, inflammatory bowel disease, Behcet's disease and lymphoma, the appearance of both dermatoses in the same person is known to be rare. Recently Cohen et al[19] suggested that Sweet's syndrome and EN are the same reactive dermatoses which share common clinicopathological features and the pathogenesis may be related to cytokines secreted from stimulated epidermal/dermal cells.

Even though there is a distinction between EN and Sweet's syndrome, both diseases have common features ; a history of frequent upper respiratory tract infection, effectiveness of steroid and potassium iodide therapy, and the association with

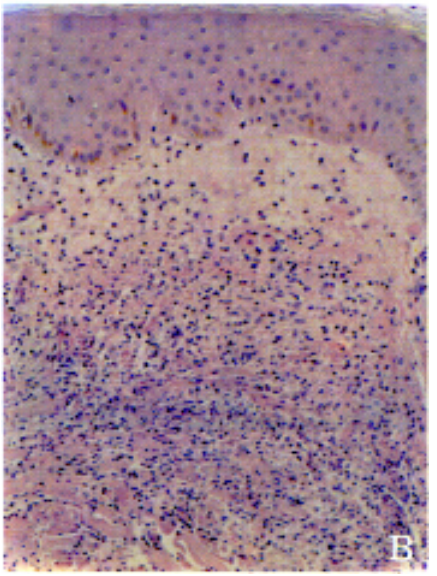

Fig. 3-B. H igh magnification of Figure 3-A. septal inflammation predominantly by neutrophils (H-E stain : ×200).

arthralgia and fever. In our patient, not only the typical lesions of Sweet's syndrome present, but also EN-like lesions responded to systemic steroid therapy. Our patient also had arthralgia at the same time.

In conclusion, we suggest that there seems to be a good chance of the association of Sweet's syndrome with EN and they may have a common pathogenesis.

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