

Neutrophilic Dermatositis of the Face Associated with Aortitis Syndrome and Hashimoto's Thyroiditis

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This is the first reported case of neutrophilic dermatosis of the face (Sweet's syndrome) associated with aortitis syndrome (Takayasu arteritis) and Hashimoto's thyroiditis. The patient was a 39-year-old Japanese female for whom corticosteroid therapy was effective.

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We report a case of a woman with aortitis syndrome and Hashimoto's thyroiditis, who developed elevated erythematous plaques on the face. Histological findings were compatible with the diagnosis of neutrophilic dermatosis of the face, a variant form of Sweet's syndrome.

Sweet's syndrome is associated with a variety of systemic conditions; however, we are not aware of any previously published report in English on these associations.

CASE REPORT

A 39-year-old Japanese woman was admitted to our hospital. She had a 5-year history of intermittent fever of undetermined origin, cough and general fatigue. On admission, she complained that an eruption had appeared on her face. Skin examination revealed several discrete erythematous and edematous plaques, 1 cm in diameter, some of which were pustular in appearance, on the right cheek (Fig. 1). Skin biopsy showed an intact epidermis, edema of dermal papillae and a diffuse, moderately dense dermal infiltrate of predominantly polymorphonuclear leukocytes.

The major abnormal laboratory data were as follows: erythrocyte sedimentation rate, 124 mm/h; leucocyte count, 9300/mm³ (neutrophils 82%); serum protein, 8.3 g/dl, with gammaglobulin, 36.6% (IgG 3358 mg/dl, IgA 633 mg/dl, IgM 215 mg/dl); thyroid test 1280 (normal <80); microsome test 300 (normal <100); TSH 4.01 (0.5-4.0) U/ml; T₃ 133 (75-160) ng/dl; T₄ 14.6 (4-11.5) µg/dl; free T₄ 1.87 (0.8-2.1) ng/dl.

Hashimoto's thyroiditis was diagnosed on the basis of the presence of goiter and the high thyroid test and microsome test titers. The thyroid scintigram with ⁹⁹Tc showed swelling of the thyroid gland, but there were no space occupied lesions.

Aortitis syndrome was suggested by the presence of murmurs in the carotid arteries. The angiographic findings of arterial ectasia of the thoracic artery and right carotid artery and arterial ectasia and stenosis of the left carotid artery were consistent with aortitis syndrome.

The patient was treated with prednisolone at an initial dose of 50 mg daily. Systemic corticosteroid therapy led to the resolution of both the systemic complaints and the cutaneous lesions. The dose of prednisolone was gradually decreased to 20 mg daily, and the patient was discharged in good condition.

DISCUSSION

Our case showed clinically edematous erythematous plaques

on the face, and histologically a dense neutrophilic infiltrate in the dermis was observed. Therefore the lesions were diagnosed as a neutrophilic dermatosis of the face, a variant form of Sweet's syndrome.

Although the etiology of Sweet's syndrome remains unknown, it is most commonly suggested to be a hypersensitivity reaction to various agents. Sweet's syndrome has been found to be associated with a variety of systemic conditions. Notable among these are respiratory tract infections and myeloproliferative disorders (especially acute myeloid leukemia). This syndrome is also associated with such auto-immune diseases as Sjögren's syndrome (1), rheumatoid arthritis (2) and subacute cutaneous lupus erythematosus (3).

Hashimoto's thyroiditis is an auto-immune disease. From the clinical stand-point, the most important findings for the diagnosis of this disease are the presence of antithyroglobulin and antimicrosomal antibodies. In this disease, the thyroid function may be reduced, normal or even increased depending on the residual thyroid cell function, and early in the disease, the patient is metabolically normal. Our patient showed thyroid enlargement in conjunction with a high thyroid and microsome test, the serum TSH mildly elevated, but the free T₄ index in the normal range. So we diagnosed that our patient was in the early or subclinical phase of Hashimoto's thyroiditis. Recently, it has been established that both Basedow's disease and Hashimoto's thyroiditis have as their underlying cause a deficiency in suppressor T-lymphocyte function (4). Although the association of Sweet's syndrome and Basedow's



Fig. 1. Discrete, erythematous and edematous plaque on the right cheek.

disease has been reported (5), to our knowledge, the association of Sweet's syndrome and Hashimoto's thyroiditis has never been reported.

Aortitis syndrome is a rare chronic inflammatory arteriopathy affecting mainly the aorta and its branches. Many skin manifestations have been reported to accompany this syndrome, such as erythema nodosum, erythema induratum and pyoderma gangrenosum (6,7), and most cases of pyoderma gangrenosum associated with aortitis syndrome have been observed in Japan (6). The clinical and histological similarities of pyoderma gangrenosum and Sweet's syndrome have led to the suggestion that these diseases might represent two extremes of one neutrophil-mediated hypersensitivity reaction (8). We conclude that neutrophilic dermatosis of the face (Sweet's syndrome) should be considered among the skin manifestations which can be associated with aortitis syndrome.

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