

LETTERS TO THE EDITOR

Concomitant Sweet's Syndrome and Relapsing Polychondritis

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Sir,

Sweet's syndrome (SS) was first described by Sweet in 1964 (1). It is a neutrophilic dermatosis, characterized by a sudden onset with fever, leukocytosis and erythematous plaques, papules or pseudo-vesicles on the face, neck, chest and extremities. The patients may also show ocular involvement (conjunctivitis, episcleritis and iridocyclitis) and arthralgia, arthritis and myalgia. Skin biopsies show a neutrophilic infiltration mainly in the upper dermis. Leukocytoclasia with nuclear dust formation is usually present, but no true vasculitis with fibrinoid necrosis of the vessel wall is seen. SS may be associated with infections in the upper respiratory tract and in the gastrointestinal tract, autoimmune diseases, vaccination, pregnancy, inflammatory bowel disease, and it may be a paraneoplastic phenomenon or idiopathic. Symptoms resolve rapidly after treatment with systemic steroids.

Relapsing polychondritis (RP) is a disease characterized by inflammation and degeneration of cartilaginous tissues, especially auricular, nasal and laryngeal cartilage. Characteristically the earlobe is usually spared (2). Biopsies from affected areas show degeneration of the marginal chondrocytes and a perichondritis dominated by numerous neutrophils. In time, more lymphocytes, plasma cells, histiocytes and a few eosinophils form the infiltrate – and fibrotic scarring appears. RP may have dermatological manifestations, such as aphthosis, nodules, purpura, pustules, papules, superficial phlebitis and livedo reticularis, and may involve almost any part of the eye (e.g. scleritis, iridocyclitis, optic neuritis) (3). RP may be associated with autoimmune diseases (e.g. Sjögren's syndrome, psoriatic arthritis, rheumatoid arthritis, systemic lupus erythematosus), inflammatory bowel diseases, endocrine diseases (diabetes, Hashimoto's thyroiditis), systemic vasculitis and neoplastic syndromes. RP also responds promptly to treatment with corticosteroids (2).

We wish to report an unusual case of coexisting SS and RP.

CASE REPORT

A 41-year-old Caucasian woman developed throat angina and high fever (39–40°C) upon return from a holiday in Turkey, and was started on penicillin treatment by her family doctor. The patient had a history of hyperthyroidism and received 250 mg tiamazol and 100 µg levothyroxin daily, but was otherwise healthy. After 2 days she developed a sore red rash on her neck

concomitantly with a severe inflammation of her ears located to the helix and anthelix, but sparing the earlobe. She was admitted to the department of rhino-laryngo-otology. At this time the patient still had a high fever, C-reactive protein (CRP) of 206 (normal <75) but no increase in circulating leucocytes. Penicillin treatment was stopped and the patient was given dicloxacillin 1 g i.v. four times daily. However; due to phlebitis and a sinus tachycardia dicloxacillin treatment was stopped, and treatment with cefuroxime 1 g four times daily was instituted. This treatment had no effect on the pain, rash or fever and the patient was referred to the department of dermatology.

The patient presented with an erythematous pseudo-vesicular rash on the neck, the face and the upper part of the trunk. The outer ear was tender and inflamed, except for the earlobe (Fig. 1). Clinically, the patient had concomitant RP and SS. The patient had fever and increased CRP (876, normal <75) but normal leucocyte count. Furthermore the patient had scleritis, but no signs of additional ocular involvement, as ascertained by an ophthalmologist.

A skin biopsy (Fig. 2A) showed marked dermal oedema and an interstitial inflammatory infiltrate with numerous neutrophils and a few eosinophils. Vessel-related inflammation was present, but no true vasculitis was found. A biopsy from the ear including cartilage showed a severe, subacute, partly granulomatous inflammatory cell infiltrate containing many histiocytes, as well as lymphocytes, plasma cells, eosinophils and a few neutrophils. These changes reached the perichondrium and the superficial cartilage (Fig. 2B). The findings were compatible with SS and RP.

Antinuclear antibodies were marginally increased (titre 1:160). IgM rheumafactor and an M-component were not present. Blood cultures were negative. During the entire course of the disease no increase in circulating neutrophils could be detected. The level of thyroid stimulating hormone was suppressed by the tiamazol treatment, but normal levels of T3 and T4 were found.

The patient was treated with prednisolone 50 mg/day p.o. for 5 days then tapered with 10 mg every 7th day. The treatment had an immediate effect on the symptoms in the skin and in the outer ear.

After 6 days of treatment only a few telangiectasies remained in the skin. The patient was followed in the outpatient clinic once every month for 3 months and again after 6 months. She experienced no relapse of the symptoms and commenced intense pulsed light (IPL) therapy for the telangiectasies.

DISCUSSION

RP and SS are very rarely found in the same patient. To date only 13 patients have been described having both conditions. Three of these patients were from a study of 48 patients with SS at the Mayo Clinic who primarily suffered from RP and subsequently developed SS (4). Seven patients were from a study of 200 cases of RP in Paris; no temporal association was described



Fig. 1. (A) Marked inflammation of the helix and anthelix, but sparing the earlobe, along with an erythematous pseudo-vesicular rash on the side of the neck. (B) Erythematous pseudo-vesicles on the back and neck.

(5). Two patients from case reports manifested RP followed by SS with an interval of months to years, and in one patient SS was followed by RP (6–8). Six of the cases were associated with malignancies, mainly haematological. Our case fulfilled the criteria for both SS and RP simultaneously. She was examined and no signs of associated diseases except for a well-treated hyperthyroidism and a preceding throat infection were found. Since RP may be associated with thyroid diseases and SS with infections this may be the reason for the coincidence.

However, in 12% of RP cases additional dermatological manifestations may be seen such as SS, pyoderma gangraenosum or erythema elevatum diutinum, especially when RP is associated with malignancies (5). Thus, the coincidence of SS and RP in this case may be due to an identical inflammatory process.

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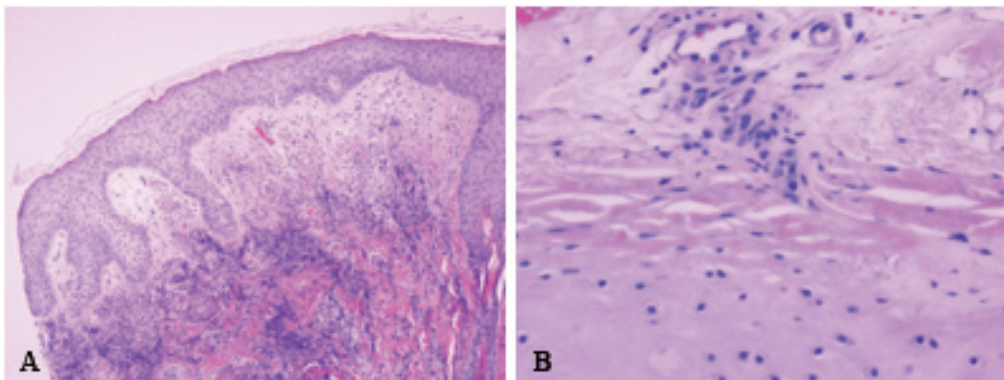


Fig. 2. (A) Histology of the pseudo-vesicular elements on the side of the neck with marked oedema and neutrophilic inflammation in the dermis. (B) Histology of the inflammation of the ear, a mixed inflammatory infiltrate affecting the perichondrial tissue.