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Cryptogenic organizing pneumonia in association with Sweet's syndrome

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Case Report

A 34-year-old man presented with a 4-week history of dry cough, shortness of breath, fever and weight loss following a 4-month history of general malaise, occasional night sweats and sinusitis. He had smoked 20 cigarettes daily since his twenties but had no previous respiratory history. He had no contact with tuberculosis and the only other history of note was the attendance at surgical outpatients the previous year for investigation of knee arthralgia at which time investigations were normal.

On examination he was pyrexial but had no lymphadenopathy or finger clubbing. His skin was unremarkable. Chest auscultation revealed coarse crackles at the right apex. Chest radiograph confirmed right upper lobe consolidation without cavitation or lymphadenopathy (Plate 1).

Investigations showed Hb 13·8 g dl^{-1}, WBC 11·9 nl (76% neutrophils, 11% lymphocytes, 3% eosinophils) and erythrocyte sedimentation rate (ESR) 101 mm h^{-1}. Renal and liver function were normal. Repeated sputum cultures were negative for organisms and initially direct then later culture negative for acid-fast bacilli and fungi. Serial blood cultures grew no organisms and serology for mycoplasma, chlamydia, coxiella, adenovirus, aspergillus, borrelia and legionella were negative. Urine microscopy was normal. Auto-antibody screen, anti-neutrophil cytoplasmic antibody, serum angiotensin converting enzyme and rheumatoid factor were negative.

Eythromycin therapy was commenced orally, but the patient remained pyrexial with respiratory symptoms, and chest radiograph showed progression of consolidation to involve the right middle and lower lobes. Computed tomographic scan demonstrated dense consolidation in the apical segment of the right lower and middle lobe with alveolar infiltration at the right apex. Peripheral low density infiltrates were seen in the left lung. There were no features suggestive of malignancy and no mediastinal lymphadenopathy. Fibre optic bronchoscopy was performed showing no endobronchial abnormality and washings from the right lower lobe revealed no organisms or acid-fast bacilli. Histological examination of the transbronchial biopsy showed an interstitial inflammatory cell infiltrate consisting of plasma cells and lymphocytes with some neutrophils and occasional eosinophils. Buds of granulation tissue (Masson

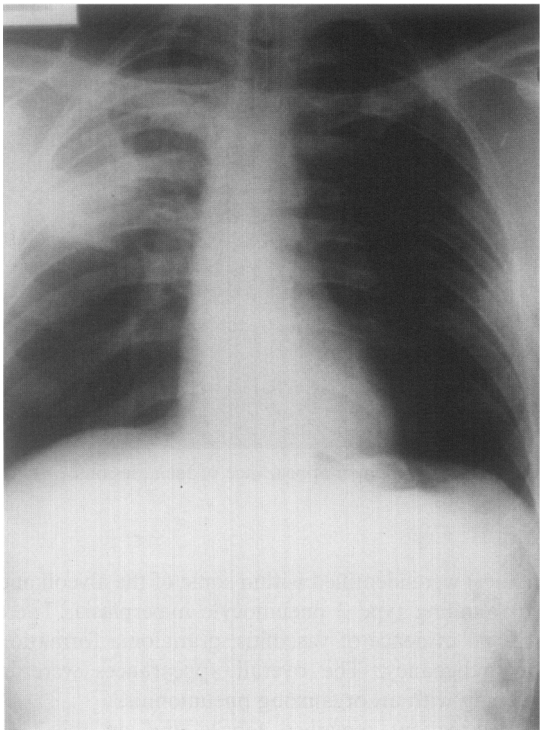

Plate 1 Chest radiograph on admission.

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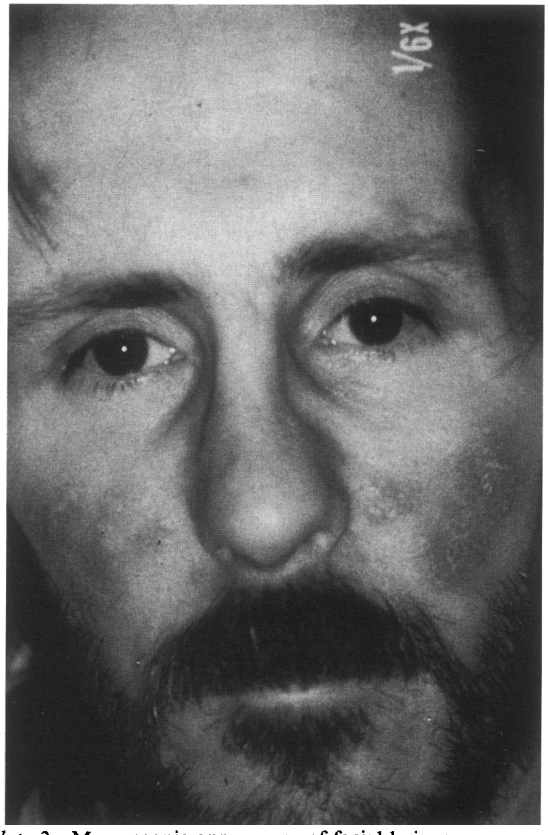

Plate 2 Macroscopic appearance of facial lesions.

bodies) were identified within some of the alveoli and surrounding type 2 pneumocyte hyperplasia. There was no evidence of vasculitis, granuloma formation or malignancy. The overall appearances were in keeping with an organizing pneumonitis.

The patient remained febrile and on the 11th hospital day developed discrete painful maculopapular eruptions on the face and neck (Plate 2). Skin biopsy showed a dermal infiltrate of neutrophils and eosinophils with accompanying oedema and a marked scattering of nuclear dust. The features were consistent with a diagnosis of Sweet's Syndrome (acute febrile neutrophilic dermatitis).

A diagnosis of Sweet's syndrome with cryptogenic organizing pneumonia was made and treatment with prednisolone (1 mg kg^{-1} day^{-1}) commenced. This produced a resolution of symptoms with clearing skin and pulmonary lesions, improved pulmonary function tests and resolution of the elevated ESR. Follow-up for 2 yr has shown no evidence of underlying malignancy, the respiratory function tests have reverted to normal. Prednisolone therapy has been discontinued and his arthralgia was unaltered by treatment.

Discussion

Sweet's syndrome (acute febrile neutrophilic dermatosis) is characterized by fever, painful erythematous plaques on the face, neck and limbs, arthralgia, neutrophilia and histological evidence of dermal neutrophilic infiltrate in the absence of infection (1). Most cases are associated with a viral upper respiratory tract infection but up to 15% of cases are associated with an underlying haematological malignancy (2).

Seven cases of Sweet's syndrome with pulmonary involvement have been described (3–8), six cases with a predominantly neutrophilic interstitial inflammatory process and one case associated with bronchiolitis obliterans organizing pneumonia with an eosinophilic, neutrophilic and plasma cell infiltrate (7). Other cases have highlighted pulmonary infiltrates that cleared with corticosteroid treatment without any diagnostic procedure performed (9, 10).

Cryptogenic organizing pneumonitis was first described by Davidson *et al.* (11) in 1983 and is characterized by the presence of buds of granulation tissue in the alveoli and alveolar ducts (Masson bodies). It is essentially the same condition described by Epler as bronchiolitis obliterans organizing pneumonia (12).

Our patient demonstrates many features characteristic of cryptogenic organizing pneumonitis: the febrile illness, microbiologically negative pneumonia, high ESR, restrictive lung defect and the prompt and dramatic response to corticosteroids.

The aetiology of either condition remains unknown although this case and that described by Chein *et al.* (7) suggests a common aetiological trigger may be operative. Both are recognized as tissue reactions to a variety of injurious agents, may be associated with peripheral blood neutrophilia and elevated erythrocyte sedimentation rate, and display a dramatic response to corticosteroids.

In conclusion, Sweet's syndrome may be associated with cryptogenic organizing pneumonia and cause substantial morbidity. Treatment with corticosteroids is effective for both conditions. The similarities between the two conditions may suggest a common aetiological trigger.

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