Case report

Incomplete Behçet's syndrome with unusual manifestations

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SUMMARY We report a case of incomplete Behçet's syndrome with two major criteria plus interstitial pneumonia. The patient had also hypoplastic bone marrow and disseminated intravascular coagulation developed during the terminal stage.

Key words: interstitial pneumonia, bone marrow failure.

Behçet's syndrome is a multisystem disease which may affect many organ systems in addition to the originally described triple symptom complex. Lung involvement is one of the rare complications of the disease,¹⁻³ but bone marrow involvement has not been described so far. We have recently seen a patient who had two major symptoms of Behçet's disease (aphthous lesions and genital ulceration) combined with interstitial pneumonia and hypoplastic bone marrow without any other underlying cause.

Case report

A 32 year old female patient admitted to our hospital had experienced cough and dyspnoea for three months. Three years previously when genital ulcers and oral aphthous lesions first developed she had been diagnosed as having incomplete Behçet's disease. For the last three years, in addition to recurrences of similar lesions, she also had purpuric lesions on her legs. Two years ago, bleeding which occurred after the extraction of a tooth lasted two days and one unit of blood transfusion was necessary. Her history disclosed no lesions of the eye or skin or arthralgia. No exposure to drugs or toxic agents was reported.

On physical examination she was dyspnoeic and pale. There were several aphthous lesions in her mouth and two ulcers on the labia majora. Several

purpuric lesions were seen, mainly on the pretibial region and also on the abdomen. Bronchial rales were detected in the lungs. Ophthalmological examination was normal.

Haemoglobin concentration was 90 g/l; white cell count $1.8 \times 10^9/l$ (42% lymphocytes); platelet count $75 \times 10^9/l$; and erythrocyte sedimentation rate 104 mm/h. Routine biochemistry tests including liver function, renal function and coagulation were found to be within normal limits. The acid Ham test, purified protein derivative test for tuberculosis (5 TU), hepatitis B surface antigen, lupus erythematosus cell test, tests for antinuclear and anti-DNA antibodies; and test for rheumatoid factor were all negative. The needle prick test was negative on several occasions.

Bone marrow aspiration and biopsy showed a decrease in cellular elements with a normal erythroid/myeloid ratio and an increase in fibrosis and fat cells.

There was a diffuse reticulonodular infiltration on the chest x ray film and lung function tests showed a restrictive type of lung involvement. A lung biopsy specimen was obtained surgically and in addition to increased fibroblastic activity, a marked nodular mononuclear cell infiltration was observed in the interstitial area. There was no sign of vasculitis in the lung arterioles and venules. A biopsy specimen obtained from the genital ulcer, however, showed the presence of vasculitis. After treatment with 1 mg/kg of prednisolone the patient felt well, her dyspnoea partly resolved, her sedimentation rate decreased, and she was discharged from hospital.

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One month later the patient was readmitted with a high fever, severe dyspnoea, bleeding from the nose, genital ulcers, and pain in the abdomen. Her general condition was poor and there were widespread disseminated purpuric lesions.

Haemoglobin concentration was 70 g/l, white cell count $0.8 \times 10^9/l$ (35% lymphocytes), and platelet count $5 \times 10^9/l$. In the blood smear obvious erythrocyte fragmentation was detected. There was free haemoglobin both in the serum and in the urine and a high concentration of fibrin degradation products in the urine.

There was no sign of systemic infection, malignant processes, or any other underlying factor which might have accounted for this blood picture. All microbiological, biochemical, and immunological tests remained negative. On the eighth day in hospital the patient died of intracranial bleeding. Needle biopsy samples obtained after death showed a hypoplastic bone marrow with a marked increase of fat tissue, and interstitial pneumonia with a nodular mononuclear cell infiltration.

Discussion

This patient does not fulfil the criteria for diagnosis of definite Behçet's disease¹ as she had only two major symptoms (aphthae and genital ulceration). A diagnosis of systemic lupus erythematosus (SLE) could be considered as nodular lymphocytic infiltration in the lung, anaemia, leucopenia, thrombocytopenia, and vasculitis are well known manifestations of this disease. However, not only were the most frequently occurring symptoms like

arthralgia and skin rash absent but also the antinuclear antibody test, almost always positive in active SLE, was negative in this patient. Rarely, there are patients with SLE who are antinuclear antibody negative and anti-Ro positive, but these show photosensitive skin rashes, arthralgia, and their disease has a benign course⁵ in contrast with our patient. The clinical manifestations and biopsy findings of this patient eliminated other systemic vasculitis syndromes like polyarteritis nodosa, drug induced vasculitis, or sarcoidosis, which were considered in the differential diagnosis.

Therefore, we believe that this patient had incomplete Behçet's syndrome, combined with interstitial pneumonia and bone marrow involvement. Interstitial pneumonia has been described in Behçet's disease.^{1,3} Bone marrow involvement has not been described, however, and this finding needs to be confirmed before it is regarded as a manifestation of Behçet's disease.

References

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