Behçet’s syndrome presenting as myocardial infarction with impaired blood fibrinolysis

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SUMMARY A 34 year old man with no known coronary risk factors presented with acute myocardial infarction. Shortly after admission, and over the next eight years, he had recurring clinical features consistent with a diagnosis of Behçet’s syndrome. Coronary angiograms recorded six months after infarction were normal. Although he was well with maintenance steroid treatment, blood fibrinolysis was impaired. Myocardial infarction has not previously been reported as a presenting feature of the syndrome.

In 1937 Behçet described a triad of orogenital ulceration and ocular lesions. Since then, protean manifestations of Behçet’s syndrome have been recognised, including cutaneous, joint, neurological,1 intestinal, and vascular involvement.2 It has been estimated that vascular involvement occurs in 7–29%4–6 of patients with a 20% mortality in those severely affected.7 Veins and peripheral arteries are usually involved, but more recently cardiac complications have been reported in the form of pericarditis, myocarditis,8 atrial fibrillation, and, in one case, myocardial infarction during the course of the disease.9

To our knowledge, however, coronary artery occlusion occurring as a presenting feature of the disease has not been previously reported. In addition, our patient had decreased blood fibrinolytic activity, a feature which may be of aetiological and therapeutic importance.

Case report

FIRST ADMISSION

A previously well 34 year old male non-smoker presented in 1975 with a prolonged episode of classical cardiac pain. He was admitted to the coronary care unit, where, during the first 24 hours of admission, he received intravenous lignocaine for ventricular extrasystoles. Thereafter, his cardiovascular status was satisfactory; however, 48 hours after admission he developed a widespread erythematous skin rash, a monoarthritis of the right knee, and oral ulceration. These symptoms resolved with conservative management and he was discharged after 10 days, feeling well and taking no medication.

Serial electrocardiograms and cardiac enzymes confirmed the development of an acute anteroseptal myocardial infarction. A chest radiograph was normal. The erythrocyte sedimentation rate (Westergren) was 115 mm in the first hour on admission, falling to 10 mm at discharge. Haemoglobin concentration was 15 g/dl; the white cell count showed a polymorphonuclear leucocytosis of 20 000 × 109/l, which had resolved by the time of discharge. Tests for rheumatoid factor, lupus erythematosus cells, latex, and antinuclear factor were negative. Throat swabs and blood cultures produced no growth. Wassermann reaction, Widal test, viral studies and serological studies for the causative agents of brucellosis, toxoplasmosis, and leptospirosis were negative. Lipid concentrations were normal. Coronary angiography six months later showed normal vessels.

One year after infarction he developed painful red eyes. Iridocyclitis was diagnosed, and he was initially given steroid eye drops and then oral prednisolone for three months. Laboratory data at this time are no longer available.

SECOND ADMISSION

In 1978 (two and a half years after infarction) he was admitted to another hospital with a 10 day history of pain and swelling in the left calf. A deep vein throm-
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Behçet's syndrome was diagnosed, and he was given anticoagulants with intravenous heparin in the first instance and warfarin subsequently. During admission he developed a widespread rash consistent with erythema nodosum and an arthropathy of the left ankle. Prednisolone 20 mg a day was given with a pronounced improvement in symptoms. Warfarin was continued for two months after discharge and prednisolone reduced to 2 mg daily.

Laboratory investigations showed an erythrocyte sedimentation rate of 72 mm in the first hour. Blood cultures and tests for antistreptolysin O and antinuclear factor were negative. A Mantoux test was weakly positive. Ascending venography confirmed the presence of a deep vein thrombosis in the left calf. His condition improved, and he returned to full time work, although over the next three years he had intermittent recurrences (every 2-3 months) or oral ulcerations; plasminogen activator: (plasminogen factor) was positive. A deep vein thrombosis in the left calf. His condition improved, and he returned to full time work, although over the next three years he had intermittent recurrences (every 2-3 months) or oral and genital ulcers, which responded to a temporary increase in prednisolone dosage from 2 mg to 10 mg daily. In view of his history of rashes, iridocyclitis, and arterial and venous thrombosis, Behçet's syndrome was diagnosed. In the absence of evidence of coronary atherosclerosis and embolism the myocardial infarction was probably a consequence of coronary arteritis.

FOLLOW UP

After review in 1983 he still had orogenital ulcers, and since fibrinolytic activity of blood may be diminished in this syndrome, blood fibrinolysis was measured (plasminogen activator: 50%, 53% of normal pool using fibrin plate technique measured on two occasions; plasminogen 36 g/l (normal) and fibrinogen 30 g/l (normal); plasma and whole blood viscosity and red cell deformability normal).

Since fibrinolytic activity was decreased a fibrinolytic enhancing agent, stanozolol was given.

Discussion

Behçet's syndrome is a multisystem disorder in which vasculitis is considered to be the unifying pathological process. The vascular system is often involved, most frequently the venous side.

1. Aneurysm formation is the most notable arterial abnormality and myocardial infarction has only been reported. As our patient fulfilled the criteria for a diagnosis of Behçet's syndrome we consider that this case reports acute myocardial infarction as the presenting feature of this syndrome, secondary to coronary arteritis. By the time of angiography the obstruction had cleared.

Furthermore, we found decreased blood fibrinolysis in this patient, which agrees with the findings of Cunliffe et al. and Chajek et al. Interestingly, this decreased fibrinolytic activity persisted despite treatment with steroids, which have been reported to increase fibrinolysis in the blood. Cunliffe et al. and Chajek et al. suggest that impaired fibrinolysis might be an important factor in the pathogenesis of Behçet's syndrome and reported three patients whose symptoms responded to fibrinolytic enhancing agents such as phenformin, ethylloestrenol, and stanozolol. A search for clues to the aetiology of Behçet's syndrome is of prime importance since response to treatment is less satisfactory in this disease than in many of the rheumatological vascular disorders. Immunosuppressive agents, including steroids, are commonly used but rarely provide a complete cure. One of the difficulties in assessing response to treatment in this disease is the variable natural history, with spontaneous remission occurring not infrequently. Because of this these preliminary reports of patients responding to fibrinolytic enhancing therapy should be interpreted with caution. Nevertheless, our case adds further weight to the argument that impaired fibrinolysis is a pathogenic mechanism, and further work in this area should be encouraged.

References

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