Postgraduate Medical Journal (July 1981) **57**, 448–449

Behçet's syndrome and neutropenia

R. C. F. LEONARD
B.Sc., M.D., M.R.C.P.

R. B. THOMPSON
M.D., F.R.C.P.

Department of Medicine, Royal Victoria Infirmary, Newcastle upon Tyne NE1 4LP

Summary

A case is described of Behçet's syndrome in which the active phases of the disease were associated with a profound neutropenia in contrast to the expected finding of normal or raised neutrophil count.

Case report

The patient was first seen at the age of 15 years in 1968 when the problem was one of severe oral ulceration of one year's duration. At that time he also had anterior uveitis. There was a marked neutropenia of less than 100×10^9/l. Because of severe symptoms, he was started empirically on a course of prednisolone with initial improvement. Subsequently, he had transient joint pains, erythematous skin rashes, and on one occasion peri-anal ulcers. Generally, however, he was well controlled on prednisolone at between 5 and 10 mg daily until the beginning of 1971 when he started to complain of headaches, and later of diplopia. In June 1971 he was admitted because of progression of these symptoms in association with intellectual deterioration. Full neurological assessment revealed paralysis of medial rotation of the left eye, intellectual impairment with marked memory loss, bilateral ankle clonus and extensor plantar responses. A lumbar puncture produced CSF with raised protein (48 mg %) and normal cells. The combination of oral, ocular, articular and now neurological symptoms suggested a diagnosis of Behçet's syndrome. A striking feature was a fall in the white cells and neutrophils accompanying this exacerbation of his symptoms (Fig. 1). The prednisolone was increased to 60 mg daily and azathioprine added at 150 mg

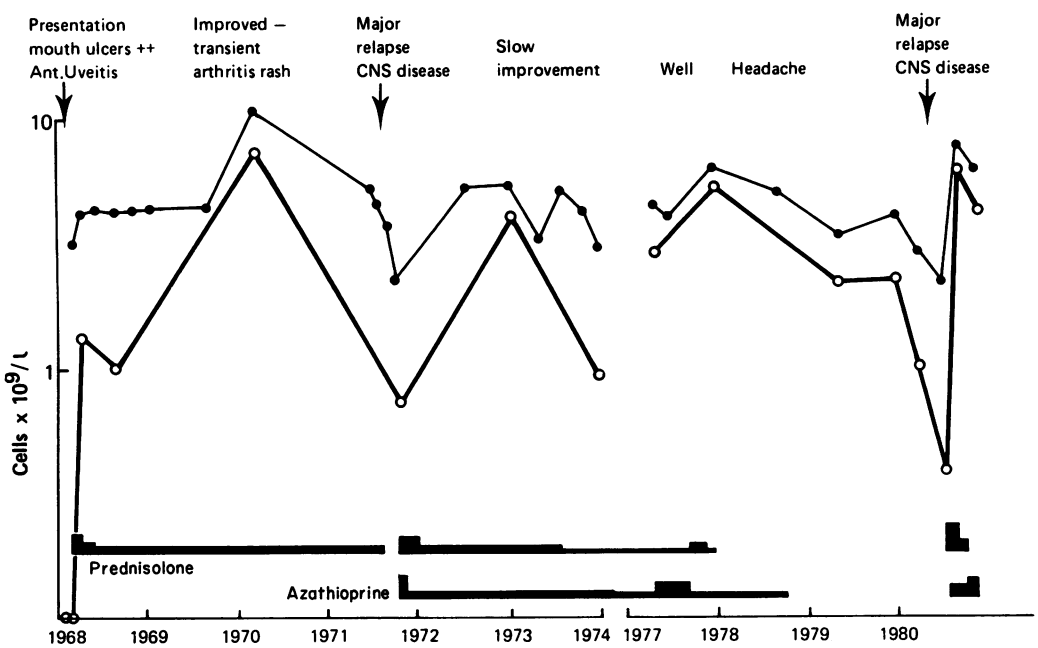

FIG. 1. Cell count of patient with Behçet's syndrome and neutropenia: ●—●, total white cell count; ○—○, neutrophil count. Prednisolone – dosage between 5 mg daily and 40 mg daily. Azathioprine – dosage between 50 mg daily and 200 mg daily.

0032-5473/81/0700-0448 $02.00

daily. There was a slow response and he remained well for 3 years on a maintenance dose of azathioprine at 50 mg daily and prednisolone 5 mg daily. The prednisolone was stopped in June 1973 and he was lost to follow-up in 1974, but when seen again 3 years later was still in good health.

Medication was eventually stopped in 1978 because of his continuing good health, but in December 1979 he complained once more of headaches which, in the absence of other features, was not regarded as good evidence of active disease. By the end of May 1980, however, it was clear that he had had a major relapse with severe headache, recurrent diplopia, mild ataxia and extensor plantar responses with brisk tendon reflexes in all limbs. This was associated with recurrent mouth ulceration, but no other external features. A lumbar puncture was normal, and prednisolone at a dose of 40 mg daily, and azathioprine at 75 mg daily were re-introduced. It was noted that over the preceding months both the neutrophil count and the total white cell count had been steadily falling and on admission the neutrophil count was less than $400 \times 10^9/l$ (see Fig. 1).

He has made an excellent clinical response to treatment and as on previous occasions the neutrophil count has responded with the clinical improvement.

Comment

Neutropenia in association with Behçet's syndrome is not well recognized and a careful review of literature (James, 1979; Sobel *et al.*, 1977; Lehner and Barnes, 1979; Matsumura and Mizushima, 1975; Mizushima, Matsumura and Mori, 1977) reveals very little information upon any haematological complications of this condition.

Disease activity is usually associated with a mild neutrophilia and an elevated ESR. Neutrophil activity has received much attention in recent years and impaired phagocytosis is a feature. It has also been postulated that the presence of immune complexes in the serum, which may be the basic pathogenetic process in the condition, affects the motility of the polymorphonuclear leucocytes (Lehner and Barnes, 1979). It was possible to find a further 3 cases out of 5 who had been studied locally and who had had adequate haematological investigations. In none of these was there neutropenia at any stage of the disease. The only other instance of neutropenia in association with Behçet's syndrome is mentioned incidentally, but was not related to disease activity (Lehner and Barnes, 1979).

References

James, D.G. (1979) Behçet's syndrome. *New England Journal of Medicine*, **301**, 431.

Lehner, T. & Barnes, C.G. (1979) *Behçet's Syndrome: Clinical and Immunological Features*. Academic Press, New York.

Matsumura, N. & Mizushima, Y. (1975) Leucocyte movement and response to colchicine treatment in Behçet's disease. (Letter.) *Lancet*, **ii**, 813.

Mizushima, Y., Matsumura, N. & Mori, M. (1977) Polymorphonuclear leucocyte function in Behçet's disease. (Letter). *Lancet*, **ii**, 1037.

Sobel, J.D., Haim, S., Obedeanu, N., Meshulam, T. & Mersbach, D. (1977) Polymorphonuclear leucocyte function in Behçet's disease. *Journal of Clinical Pathology*, **30**, 250.