CASE STUDY

Behçet's syndrome: a case study

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SUMMARY. This is the case report of a woman who for many years was a diagnostic puzzle. Her various problems were eventually identified as features of Behçet's syndrome, which is uncommon in this country. Besides the known clinical symptoms of the disease, this patient has suffered intermittently from dysmenorrhoea and menorrhagia, which may well be features of this disorder which have not previously been documented. There is no sign of remission for this patient, but awareness of her condition should lead to more sympathetic management of these cases.

Introduction

BEHÇET'S syndrome is uncommon in this country, with a prevalence of 0.6 per 100,000 of the population. It is more common in Mediterranean countries and the Middle East, and in Japan the prevalence is 10 per 100,000 of the population (Chamberlaine, 1977, 1978). The aetiology of the disease is unknown, but an immunological basis seems possible. The histology is of vasculitis, particularly involving venules and capillaries, with mononuclear cell infiltration. Circulating autoantibodies to oral mucosa were first described in 1963 (Oshima *et al.*, 1963).

The main clinical features of Behçet's disease are relapsing iridocyclitis with recurrent oral and genital ulceration. What is less well known is that this disease can affect most systems of the body (Geraint James, 1979).

Case history

Mrs S was born in 1955. At the age of two months she was admitted to hospital with a chest infection and was incidentally found to be anaemic. The blood picture returned to normal after two months of iron therapy. Between the ages of one and five years she had recurrent epilepsy.

She kept well until aged 17, when she presented with a swollen left knee. This effusion resolved three months later after a course of physiotherapy. A clinical diagnosis of chrondromalacia patellae had been made. She subsequently developed a painful and swollen metatarsal joint (at age 22) and a painful and swollen ankle (at age 24)

When she was 18 years old she had a severe attack of stomal and genital ulceration and every year since she has had three or four recurrences. These episodes usually last at least a week and cause severe dysuria and dyspareunia.

When she was 19 she complained of frequency of micturition without dysuria. On direct questioning she admitted to being incontinent at times and of being unable to feel anything when she voided. Full investigation, including intravenous pyelography and cystoscopy, revealed nothing. A tentative diagnosis of neurogenic bladder was made. She completely recovered her bladder function after six months.

At age 20 she had an attack of diarrhoea which lasted for six weeks. The following year she had another attack of diarrhoea, this time with blood and mucus in the stools. Full investigation revealed no abnormality. She continued to have recurrent episodes of diarrhoea for three years.

The patient had two normal pregnancies at ages 20 and 21. When she was 23 she complained of dysmenorrhoea and had menorrhagia. She suffered intermittently from this problem for five years. On two occasions she has had full gynaecological investigation with negative findings.

On several occasions the patient has suffered from severe headaches and once her headache was accompanied by neck stiffness and fever.

Since the age of 24 she has had six or seven attacks of bronchitis, with wheezing and occasional haemoptysis. Chest radiographs have shown no abnormality.

At age 25 she developed conjunctivitis, which did not respond to local treatments and persisted intermittently for three months. This was diagnosed as episcleritis and following this she had retrobulbar neuritis. Her eye symptoms resolved after two months.

Between 1972 and 1981 the patient attended the surgery about every two weeks; the Table lists the number of consultations over this period. From the involvement of one system after another, Behçet's syndrome was diagnosed.

Discussion

This patient is one of the group of frequent attenders who have major physical problems. The completion of the diagnostic puzzle led to a search of the literature on Behçet's syndrome. In a review of 23 patients, A. Thomson, D. Geraint James and N. G. R. Page (personal communication) stressed the clinical nature of the diagnosis. They found oral and genital ulceration, ocular inflammation, skin rashes and polyarthritis in the

Consultation rate for Mrs S from 1972 (aged 17) until 1981 (aged 26).

Year	Number of consultations
1972	9
1973	16
1974	17
1975	16
1976	13
1977	7
1978	16
1979	17
1980	15
1981	20
Total	146

majority of cases. Central nervous system involvement, thrombophlebitis, cardiovascular involvement, gastrointestinal lesions and renal disease are also features of this disorder.

Behçet's syndrome is not a benign disease. Ulcerating haemorrhagic lesions involving the mucosa of the oesophagus, stomach, intestine or anus can lead to fatal perforation. Involvement of the central nervous system can include mood changes and a progressive confusional state which can lead to dementia. The ocular symptoms can terminate in blindness.

This case history included recurrent wheezing bronchitis with haemoptysis; this feature of the disease has only recently been documented (Gambel *et al.*, 1979). Persistent dysmenorrhoea and menorrhagia may well be features of this disorder which have not previously been documented.

Conclusion

The disease shows no sign of remission in this case. The 'minor' features were major ones for Mrs S, who endured 'colitis' for two years and has had painful arthritis in the elbow, sacroiliac joints, knee and ankles at various times. The CNS manifestations of urinary incontinence were most unpleasant for six months, and the persistence of abnormal uterine bleeding and recurrent bronchitis is still causing her much suffering. In a protracted and difficult case such as that of Mrs S, the role of the general practitioner in the medical team is to support the patient and control her contact with specialist departments. In this case the doctor/patient relationship has been under strain for years because of the frequent attendances at the surgery with a series of baffling problems which at best have required only symptomatic treatment and at worst have proceeded for weeks and weeks without responding to any remedy.

Over the years the patient has endeavoured to relate to a succession of doctors at the local hospital, at one time or another attending no less than five different departments and being extensively investigated by each in turn. A study of the literature on Behçet's syndrome has improved our understanding of this patient's problems and we hope this will lead to more sympathetic management in the future.

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Hypertension

Ninety-nine per cent of the 2,495 employees of an insurance company in Massachusetts were screened for hypertension. The illness absenteeism of 259 hypertensives was studied for the year after they had been screened and identified. Absenteeism increased more in the 48 people who were previously unaware of their hypertension than in the 211 who were aware. Among the newly labelled hypertensives only the young (<34 years) and those with 'pure' systolic hypertension were absent more often from work; older people with diastolic hypertension were not. Those who received active follow-up and anti-hypertensive treatment had only minimal increase in absenteeism. In contrast, those who received active follow-up without medication, or only episodic follow-up, had significantly increased absenteeism.

Source: Charlson, M. E., Alderman, M. & Melcher, L. (1982). Absenteeism and labelling in hypertensive subjects. Prevention of an adverse impact in those at high risk. *American Journal of Medicine,* **73,** 165-170.