- READ, A.E., SHERLOCK, S. & HARRISON, C.V. (1963) Acute 'juvenile' cirrhosis considered as part of a systemic disease and the effect of corticosteroid therapy. *Gut*, **4**, 378.
- RIIS, P. & ANTHONISEN, P. (1964) Eosinophilia in peripheral blood and inflammatory exudate in non-specific proctocolitis. *Acta medica scandinavica*, **175**, 85.
- RIISAGER, P.M. (1959) Eosinophil leucocytes in ulcerative colitis. *Lancet*, **ii**, 1008.
- SHERLOCK, S. (1974) Chronic hepatitis. *Gut*, **15**, 581.
- TAYLOR, K. (1965) Immune mechanisms in gastroenterology. In: *Recent Advances in Gastroenterology* (Ed. by J. Badenoch and B. N. Brooke), p. 24. Churchill, London.
- WRIGHT, R. & TRUELOVE, S.C. (1966) Circulating and tissue eosinophils in ulcerative colitis. *American Journal of Digestive Diseases*, **11**, 831.

Postgraduate Medical Journal (February 1977) **53**, 108–110.

Behçet's syndrome with perforations of the colon

K. HAAVIK NILSEN†
M.D., M.R.A.C.P.

S. M. JONES*
F.R.C.S.

B. A. SHOREY*
F.R.C.S.

*Department of Medicine, and *Department of Surgery, University of Bristol and
†Royal National Hospital for Rheumatic Diseases, Bath*

Summary

A case of Behçet's syndrome with mucosal ulcerations of the ascending colon and three perforations of the caecum is described. It is the first documented case of this syndrome where colitis has occurred without concomitant rectal lesions. The reported cases of Behçet's syndrome with associated colonic lesions make up a heterogeneous group. It is suggested that the different patients may have suffered from pathogenetically different diseases.

Introduction

Behçet's syndrome consisted initially of recurrent iritis with genital and oral ulcerations. Since Behçet's description in 1937 many other manifestations have been added to the original triad. They include various skin lesions, arthritis, orchitis, epididymitis, venous and arterial thrombosis, pericarditis and various lesions of the central nervous system (CNS) and the gastrointestinal tract (Chajek and Fainaru, 1975). Except for the commonly occurring uveitis, which almost invariably leads to blindness (Watson, 1975), the syndrome usually runs a relatively benign course. The only reported life-threatening manifestations are some of the rare CNS (Fadli and Youssef, 1973) and gastrointestinal (Empey, 1972) lesions.

A case of Behçet's syndrome and right-sided colitis with perforations of the caecum is now described. The colonic manifestations of the syndrome are discussed and an attempt is made to

delineate the nature of colonic involvement in Behçet's syndrome.

Case report

A 28-year-old welder had suffered over the last 13 years with recurrent, painful mouth ulcers, especially frequently during the last 3 years. In 1973 and 1974 he attended a venereology clinic for associated ulcerations of the penis. Venereal disease was excluded and no cause was found for the penile ulceration. In August 1973 he suffered an attack of right-sided orchitis. A few days after treatment with cephalexin was started, he suddenly developed an erythema nodosum-like rash on the legs and the arms associated with fever and aching joints. This cleared spontaneously. No cause was found for the rash but hypersensitivity to cephalexin was suspected. He had, however, later taken cephalexin without side effects. In March 1974, he developed a left-sided orchitis. An erythema nodosum-like rash appeared this time before any antibiotic was given. It cleared following a course of prednisolone. Since then he suffered several attacks of erythema nodosum. During the months March, April and May 1975 the rash reappeared every second to third week. Each attack lasted 4–5 days, and he was treated intermittently with prednisolone. He started to be troubled with increasing backache and arthralgia of the fingers, wrists, right elbow and right knee both at the times he had the rash and in the periods between. Left-sided sacro-iliitis was found on

X-ray. Treatment was started with 150 mg of indomethacin daily. He subsequently had another attack of left-sided orchitis, and soon after became acutely ill with fever, abdominal pain, nausea, vomiting and diarrhoea. A deep, painful ulceration developed on the side of his tongue. The only abnormal blood tests were a leucocytosis of 24,000 with 90% neutrophils and a raised serum viscosity at 1.84 cP (normal: 1.50–1.72). Except for signs of peritonitis in the right iliac fossa, rectal examination was normal. At laparotomy, faecal material was found in the peritoneal cavity. There were three perforations of the caecum and a necrotic-looking area of the colon at the hepatic flexure. Apart from the peritoneal reaction the rest of the intestine felt and looked normal. A right-sided hemicolectomy was performed with an end-to-side anastomosis between the terminal ileum and the transverse colon. Several shallow, well demarcated mucosal ulcerations were found in the removed ascending colon with normal mucosa in between. Microscopy (Fig. 1) showed that the ulcerated areas had ischaemic necrotic bases surrounded by non-specific inflammation and periarteriolar and perivenular infiltration by macrophages and lymphocytes. Some of the small vessels were thrombosed. Their walls were not infiltrated to any marked degree by inflammatory cells. There was no fibrinoid necrosis or granulomatous lesion.

FIG. 1. The opened ascending colon showing the mucosal ulcerations, and a microscopy section through the edge of one of the ulcerations showing the ischaemic necrotic base.

Initially, the patient recovered well, but on the eighth postoperative day he developed fever and arthralgia. He was transfused 3 u. of fresh blood and 30 mg of prednisolone was added to his antibiotic. On this treatment he became afebrile and asymptomatic within 8 hr. One week later he again became pyrexial, but without associated arthralgia. At this time an abscess drained through the operation wound. The fever settled. As there were subsequent problems

with wound healing, the steroid dosage was reduced rapidly. During this time he developed episodes of arthralgia, and thus the reduction of the steroid was slowed down. Some weeks later he developed signs of an ileus and a further laparotomy was performed. A paracolic abscess was drained and adhesions causing obstruction of the small intestine were divided. Following this he made an uneventful recovery. He was discharged on a daily dosage of 6 mg of prednisolone, which is being slowly reduced. There is now no evidence of intestinal disease, and the earlier troublesome erythema nodosum has not recurred. He has had one more mouth ulcer lasting 3 days. This was associated with arthralgia of the fingers and a feeling of grittiness of the eyes. No ophthalmic lesion was found.

Discussion

It was first suspected that the patient had granulomatous colitis with systemic manifestations. Histology did not confirm this diagnosis, nor that of the alternatively suspected necrotizing vasculitis. Behçet's syndrome therefore seems the most likely diagnosis. Mason and Barnes (1969) proposed that to make a diagnosis of Behçet's syndrome a minimum of three major or two major and two minor diagnostic criteria are required. The major criteria are buccal ulceration, genital ulceration, eye and skin lesions. The minor criteria are gastro-intestinal lesions, thrombophlebitis, cardiovascular lesions, arthritis, CNS lesions and a positive family history. The present patient fulfills these criteria with three major and two minor manifestations, and no other single diagnosis fits the facts.

Saugmann Jensen suggested in 1944 that the intestine may get involved in Behçet's syndrome. Since then symptoms such as abdominal pain, nausea and diarrhoea have been found to occur in some 40–50% of the patients with the syndrome (Chajek and Fainaru, 1975). Colonic lesions have been properly documented in only fourteen earlier cases. The literature on these was recently reviewed by Smith, Kime and Pitcher (1973). It was noted that all these patients had left-sided colitis with associated rectal lesions. Some of them had clinical and histological features indistinguishable from those of ulcerative colitis. They may therefore have been cases of ulcerative colitis with additional, unusual systemic manifestations. Alternatively, they may have been cases of Behçet's syndrome with concomitant ulcerative colitis. Other patients showed clinical and histological features suggestive of granulomatous colitis and, again, the correct classification is uncertain. Four patients had colonic lesions similar to those found in the present patient, with a histological picture similar to that of the oral and genital lesions (Lehner, 1969). The histological

similarities make it more likely that the colonic lesions are true manifestations of the Behçet's syndrome in these patients.

Smith *et al.* (1973) found only two cases reported in English of Behçet's syndrome complicated by perforations of the colon. The case reported by Bøe, Dalgaard and Scott (1958) did, however, have a rash typical of erythema multiforme and a typical ulcerative colitis with a splenic flexure perforation. It would be difficult to classify that as a case of Behçet's syndrome, especially when the criteria of Mason and Barnes (1969) are used. Bøe *et al.* (1958) used the term 'mucocutaneous-ocular syndrome' including several different clinical entities. The case reported by Empey (1972) had features of Behçet's syndrome with a diffuse colitis, histologically involving all layers of the wall of the colon and a perforation of the sigmoid colon. The case now reported is thus the second well documented case, in English literature, complicated by perforations of the colon, and it is the first case of Behçet's syndrome with colitis that does not affect the rectum.

In conclusion, it appears that some of the reported patients with Behçet's syndrome and colonic manifestations may have been suffering from one of at least three different disorders, and it is possible that the colonic lesions in some reported cases are due to

some concomitant disease rather than being manifestations of Behçet's syndrome.

Acknowledgments

We wish to thank Professor J. H. Peacock and Dr M. I. V. Jayson for permission to write this account of a patient under their care. K. H. N. holds a Rose Hellaby Medical Scholarship (New Zealand).

References

- BØE, J., DALGAARD, J.B. & SCOTT, D. (1958) Mucocutaneous-ocular syndrome with intestinal involvement. *American Journal of Medicine*, **25**, 857.
- CHAJEK, T. & FAINARU, M. (1975) Behçet's disease. *Medicine*. Baltimore, **54**, 179.
- EMPEY, D.W. (1972) Rectal and colonic ulceration in Behçet's disease. *British Journal of Surgery*, **59**, 173.
- FADLI, M.E. & YOUSSEF, M.M. (1973) Neuro-Behçet's syndrome in the United Arab Republic. *European Neurology*, **9**, 76.
- LEHNER, T. (1969) Pathology of recurrent oral ulceration and oral ulceration in Behçet's syndrome. *Journal of Pathology*, **97**, 481.
- MASON, R.M. & BARNES, C.G. (1969) Behçet's syndrome with arthritis. *Annals of the Rheumatic Diseases*, **28**, 95.
- SAUGMANN JENSEN, J. (1944) Ulcerøs hæmorrhagisk colitis kombineret med Behçet's syndrom. *Ugeskrift for læger*, **106**, 176.
- SMITH, G.E., KIME, L.R. & PITCHER, J.L. (1973) The colitis of Behçet's disease; a separate entity. *Digestive Diseases*, **18**, 987.
- WATSON, P.G. (1975) Behçet's syndrome. In: *Recent Advances in Ophthalmology*, No. 5 (Ed. by P. D. Trevor Roper), p. 249. Churchill Livingstone, London.