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Behçet's syndrome: severe proctitis with rectovaginal fistula formation

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SUMMARY The development of severe proctitis and a rectovaginal fistula two months after the onset of Behçet's syndrome in a 41 year old woman is reported.

Mild gastrointestinal symptoms in patients with Behçet's syndrome are common, but severe colitis especially in the early stages of the disease is rare.[1–3] We report a case of severe inflammatory disease of the rectum, which developed two months after the initial symptoms of Behçet's syndrome.

Case report

A 41 year old white woman presented with a two month history of recurrent mouth and vulval ulcers, a pustular skin rash, arthralgia, and an offensive vaginal discharge. On systemic inquiry she admitted to weight loss of two stones, increasingly frequent bowel motions, and pain on defecation with blood staining of the stool.

When first seen there were numerous large, painful ulcers on the tongue and pharynx and a pustular, follicular rash on the arms, legs, axillae, and groins. Her knees were tender with evidence of active synovitis. Abdominal examination was unremarkable, but rectal examination was painful. There was a large ulcer present on the vulva, which on biopsy showed non-specific inflammatory changes with no evidence of granuloma formation, vasculitis, or neoplasia. Symptomatic treatment with ibuprofen (200 mg three times a day) relieved her joint pain, but at review two months later the vaginal discharge and bowel symptoms had persisted.

Investigations were as follows: erythrocyte sedimentation rate 120 mm/h, C reactive protein 124 mg/l (normal <10 mg/l), haemoglobin 101 g/l, white cell count $4{\cdot}3\times10^9$/l, platelets 620×10^9/l. An autoantibody screen was negative. The urea and electrolytes, serum iron, vitamin B_{12}, and folate were all normal. Swabs from the vagina, mouth, and skin pustules showed normal flora. A barium enema showed that the rectal mucosa was abnormal, the rectum was displaced anteriorly, and a rectovaginal fistula was present (Fig. 1). The remainder of the

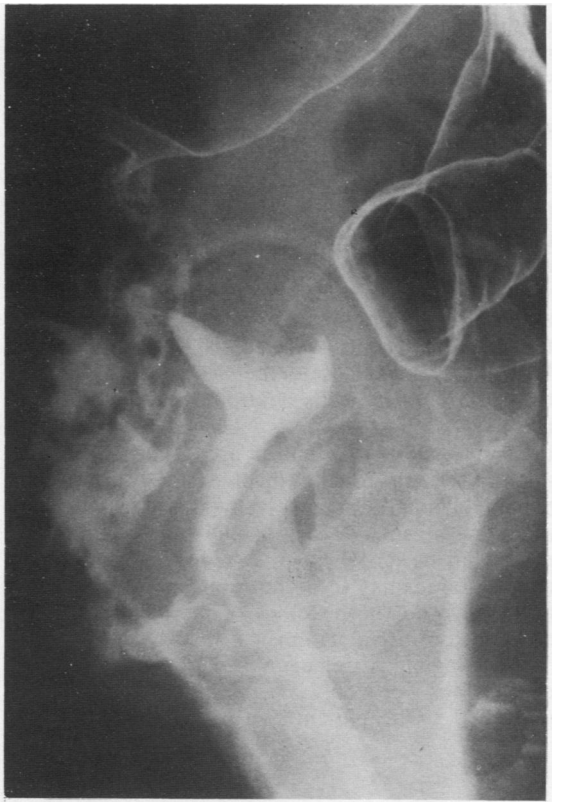

Fig. 1 *Barium enema showing a rectovaginal fistula.*

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colon was normal. Barium meal and follow through showed no abnormality in the stomach, jejunum, or ileum. Sigmoidoscopy performed under general anaesthesia showed extensive deep ulceration of the anal canal. Approximately 70% of the circumference of the anal canal was ulcerated with only two narrow strips of epithelium visible at the 5 and 7 o'clock positions. The lower rectum immediately proximal to the anal canal was also deeply ulcerated with nodular pseudopolyps. Anteriorly there was a deep cavitating ulcer in the rectovaginal septum and similar ulceration in the posterior vaginal wall, leading to free communication between the rectum and vagina. Biopsies of the rectal, anal, and perianal ulcers were performed and all showed non-specific inflammatory changes with no evidence of granuloma formation, vasculitis, or neoplasia.

Four days after starting treatment with prednisolone (20 mg three times a day) and broad spectrum antibiotics (metronidazole 400 mg three times a day and cephradine 500 mg four times a day) the mouth ulcers had disappeared and the vaginal discharge had diminished. At follow up, two months later, the vaginal discharge had settled and all the perianal, rectal, and vaginal ulceration had healed, leaving a minimally symptomatic rectovaginal fistula. These local changes remained inactive after reduction of steroids to maintenance doses.

Discussion

This woman satisfied the clinical criteria necessary for the diagnosis of Behçet's syndrome.[4] Gastrointestinal symptoms, which have been reported to occur in about 50% of patients with Behçet's syndrome, are mostly minor and include vomiting, abdominal pain, flatulence, diarrhoea, and constipation. Most patients with colitis have established Behçet's syndrome, and the average interval between onset of Behçet's syndrome and the development of colitis is six years.[5] Ulcerative changes in the small and large bowel are found in 1% of patients[3 6] and usually involve the caecum and terminal ileum.[6] Anal involvement is rare and when it occurs is usually accompanied by colitis.[7 8] The ulcers may be superficial or deep and are sometimes transmural leading to perforations or fistula formation, or both. The histology of the ulcer is that of non-specific inflammation.

Rectovaginal fistulae, however, are rare and descriptions of only four cases have been published.[5] One patient had a 15 year history of recurrent iritis/conjunctivitis, oral ulcers, and skin lesions and a six year history of vulval and anorectal ulcers before the development of diarrhoea and a rectovaginal fistula. In the second patient a rectovaginal fistula developed 15 years after the patient presented with central nervous system symptomatology, followed by arthralgias and oral and vulval ulcers. In two further cases one had an 18 month history of relapsing oral and vulval ulcers and conjunctivitis before developing a rectovaginal fistula and the other patient presented with diarrhoea and a rectovaginal fistula two years after developing arthritis, phlebitis, and oral ulcers.

Although there are similarities in the clinical manifestations, there are sufficient differences between the histological appearances to enable Behçet's colitis to be differentiated from Crohn's and ulcerative colitis. The rarity of granuloma formation, lymphoid aggregates, and submucosal fibrosis in Behçet's colitis helps to distinguish it from Crohn's disease,[7] whereas in ulcerative colitis the ulcers are confined to the mucosa and fistula formation is rare.

This patient is unusual because of the early development, the site, and the severity of the inflammatory bowel disease. It is evident from her history that symptoms of proctitis and fistula formation developed within two months of the onset of Behçet's syndrome. Anal involvement is rare as is severe localised proctitis in the absence of involvement of other parts of the gastrointestinal tract.

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References

1 Bechguard P. Case of recurrent aphthous stomatitis accompanied by conjunctivitis and ulceration of the genitalia and skin. *Ugeskr Laeger* 1940; **102:** 1019–23.
2 Saugman, Jensen J. Ulcers haemorrhagish colitis kombineset med Behçet's syndrome. *Ugeskr Laeger* 1944; **105:** 176–9.
3 Shimizu T, Ehrlich G E, Inaba G, Hayashi K. Behçet's disease. *Semin Arthritis Rheum* 1979; **8:** 223–60.
4 Mason R M, Barnes C G. Behçet's syndrome with arthritis. *Ann Rheum Dis* 1969; **28:** 95–103.
5 Smith G E, Kime L R, Pitcher J L. The colitis of Behçet's disease: a separate entity. Colonoscopic finding and literature review. *Am J Dig Dis* 1973; **18:** 987–1000.
6 Kasahara Y, Tanaka S, Nishino M, Umemura H, Shiraha S, Kuyama T. Intestinal involvement in Behçet's disease: review of 136 surgical cases in the Japanese literature. *Dis Colon Rectum* 1981; **24:** 103–6.
7 Randall G L. The colitis of Behçet's syndrome. *Am J Surg Pathol* 1986; **10:** 888–93.
8 Iwama T, Utzunomiya J. Anal complications in Behçet's syndrome. *Jpn J Surg* 1977; **7:** 114–7.