Subtotal colectomy for intestinal Behcet's disease: a histopathological surprise

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Received: 18 August 2008 / Accepted: 27 August 2008
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Abstract A 40-year-old male who was diagnosed to be having ileocaecal tuberculosis by colonoscopy elsewhere was put on antitubercular treatment. Despite therapy over three months patient had recurrent bouts of lower abdominal pain and was admitted repeatedly with features of subacute intestinal obstruction. CT- abdomen revealed concentric large bowel thickening at multiple sites with features of stenosing lesions. Patient underwent an exploratory laparotomy and subtotal colectomy was done for the diseased large bowel. Post operative histopathological examination revealed it to be Behcet's disease. We report a rare case of intestinal Behcet's disease requiring subtotal colectomy.

Keywords Behcet's disease · Intestinal Behcet's disease

Introduction

Behcet's disease is a chronic relapsing, multisystem disease characterised by oral ulceration, genital ulceration and ocular lesions. Gastrointestinal involvement is rare, often difficult to treat and associated with a high mortality rate [1]. It is most commonly seen in countries along the ancient Silk Road from Eastern Asia to the Mediterranean basin. Young adults between the second and fourth decades of life are mainly affected [2]. Although the causes of intestinal Behcet's disease are unknown, genetic, environmental and immunological factors have been suggested [3]. Here we report a case of a 40 year old male who underwent a subtotal colectomy for intestinal Behcet's disease.

Case report

A 40-year-old male who was diagnosed to be having ileocaecal tuberculosis elsewhere presented to us with recurrent episodes of acute lower abdominal pain. Patient had received three months of antitubercular treatment by then. The clinical picture during these admissions was that of subacute intestinal obstruction, which was managed conservatively. CT-abdomen with contrast was done, which revealed concentric large bowel thickening at multiple sites with features of stenosing lesions.

An exploratory laparotomy revealed an indurated lesion at the ileocaecal region and stenotic lesions at the proximal transverse colon and proximal descending colon. Mucosal aspect revealed multiple ulcers (Fig. 1), which appeared like tubercles to the naked eye. A subtotal colectomy was performed and the diseased colon was resected. An extramucosal single layer ileo-sigmoid anastomosis was performed.

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Post operatively patient made an uneventful recovery except for the oral ulcers, which he developed in the second week. To our surprise the histopathological evaluation of the specimen revealed intestinal Behcet's disease (Fig. 2). Retrospective interrogation revealed history of recurrent oral ulceration and skin lesions in the past. Patient was given oral prednisolone over 6 weeks, which was tapered and stopped. Patient is regularly under followup and is currently symptom free for the last nine months and has gained 15kgs of weight.

Discussion

Behcet's disease is a rare vasculitis diagnosed by the presence of recurrent oral ulcers and two of the following: genital ulcers, typical eye lesions, skin lesions and positive pathergy test. Young adults between the second and fourth decade of life are mainly affected, with abdominal pain being the most common symptom in patients of intestinal Behcet's disease

The ileocaecal region is most commonly affected with ulcerations that may penetrate or perforate [2]. Bowel

wall thickening is the most common finding on computed tomography (CT) scan [4]. Pathology shows a vasculitis mainly involving small veins or alternatively non specific inflammation. Intestinal Behcet's disease is shown to have a close relationship with pyoderma gangrenosum [5]. The tissue damage occurring in patients with Behcet's disease is believed to be caused by oxygen radicals which are promoted by proinflammatory cytokines and arachidonic acid metabolites [6].

The prevalence of Behcet's syndrome ranges from 1:10000 in Japan to 1:500000 in N.America and Europe. Males have more severe disease than females. The aetiology and pathogenesis of this syndrome remain obscure, vasculitis is the main pathologic lesion with a tendency to venous thrombus formation and circulating autoantibodies to human oral mucus membrane are found in approximately 50% of the patients. Familial occurrence has been sporadically reported. The disease appears to be linked to HLA-B5 (B51) alloantigens [7]. Superficial or deep peripheral vein thrombosis is seen in one fourth of patients. The superior venacava is obstructed occasionally and cases of Budd-chiari are reported.

Intestinal Behcet's disease frequently requires a surgical treatment and has a high recurrence rate. The patients who achieved a complete remission with medical treatment, who had no history of intestinal perforation, and who received azathioprine after operation showed better clinical courses [8]. New formulations of mesalazine (5-Aminosalicylic acid) are now available for the treatment of intestinal Behcet's disease. Mesalazine breaks the chain reaction of free radical production, thereby promoting long term healing of mucosal ulcerations of the intestine in patients with Behcet's disease [6].

Conclusion

To conclude, 5-Aminosalicylic acid, corticosteroids, immunosuppressants, enteral nutrition, parenteral nutrition and surgical therapy are considered standard therapy for intestinal Behcet's disease. Infliximab, colchicines, thalidomide, other pharmacological therapy, endoscopic therapy and leucocytapheresis are deemed experimental [9].

Conflict of interest The authors do not have any disclosable interest.

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Fig. 1 Multiple ulcers in the resected specimen

Fig. 2 Histopathologic picture of Behcet's disease

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