Case Report: Intestinal Obstruction in Behcet's Disease: A Rare Presentation.

CAM Firdaus[1,3], R Reynu[1], NR Kosai[1], S Das[2], SA Salauddin[1]

[1]Minimally Invasive & Upper Gastro-Interstinal Surgery Unit, Department of General Surgery, Universiti Kebangsaan Malaysia Medical Centre, MALAYSIA, [2] Department of Anatomy, Universiti Kebangsaan Malaysia Medical Centre, MALAYSIA, [3]General Surgery Unit, Surgical Cluster, Faculty of Medicine, Universiti Teknologi MARA, MALAYSIA.

ABSTRACT

Behcet's disease is a rare entity with intestinal Behcet's being an even rarer manifestation. The disease affects almost every organ in the body and eventually leads to multi organ failure. Intestinal Behcet's can be presented as intestinal obstruction and lower gastrointestinal bleeding. Surgical resection followed by systemic control of disease using steroids or immune modulators may be beneficial to prevent further recurrence and progression of the disease.

Keywords: Intestinal obstruction, Autoimmune disease, Inflammatory, Steroids

Introduction

Behcet's Disease (BD) is a rare, chronic and multisystem inflammatory disease that affects the eyes, oral cavity, and genitalia. First described by Hulusi Behcet in 1937, the highest prevalence has been reported in Turkey with 80-370 cases reported per 100,000 individuals[1]. It is also somewhat prevalent among young East Asian women and Middle Eastern men between 20-40 years of age. A rare entity in the western hemisphere, it has been reported to affect 1-2 people per 1 million Americans[2,3] Intestinal BD or entero-Behcet disease is seen in 10-15% of BD patients. These subsets of BD patients seem to also share similar characteristics as those with inflammatory bowel disease (IBD). The blurred line of differentiation between the two clinical entities often poses a diagnostic challenge[4,5]. With no confirmatory laboratory test, diagnosis often rests on the shoulders of the attending physician. We highlight a case of intestinal BD in a young female who presented with symptoms of chronic intestinal obstruction and lower gastrointestinal bleeding.

Correspondence: *Dr Mohd Firdaus Che Ani*
Minimally Invasive & Upper GI Surgery Unit, Department of General Surgery, Universiti Kebangsaan Malaysia Medical Centre.
Email:firdaus.cheani@gmail.com

Case Report

A 35-year-old lady presented to the emergency department with complaints of oro-genital ulcers, erythematous popular rash and persistent fever for the past 2 weeks. She also complained about her progressive abdominal distension associated with intermittent colicky abdominal pain and bloody diarrhoea for the past 3 days. There were two episodes of non-bilious vomiting. There was otherwise no anaemic symptom or bleeding noted from elsewhere. Her oral intake was reduced following poor appetite. Clinically, she was mildly dehydrated and febrile with temperature of 38 degrees Celsius. She was normotensive with blood pressure of 110/70mmHg but tachycardic with heart rate of 110 beats per minute. There was generalised

abdominal distension with no sign of peritonism. Bowel sounds were hyperactive on auscultation and digital rectal examination was unremarkable. Abdominal radiograph in supine position demonstrated dilated loops of small bowel. Decision for emergency laparotomy was made and surgery undertaken on the same day following fluid resuscitation and consent from patient. Intraoperatively, a constricting stricture was noted at 10cm from ileo-caecal junction as well as deep multiple serosal punctum extending into mucosal ulcers along the length of ascending colon *(Fig.1)* Right hemicolectomy was performed with functional end to end anastomosis using linear staple. Post-operative recovery was unremarkable. Patient was allowed clear fluids 24 hours later and orally the following day. She was discharged well on post- operative day 5. Histopathology examination of the resected specimen revealed severe active enterocolitis with vasculitis changes.

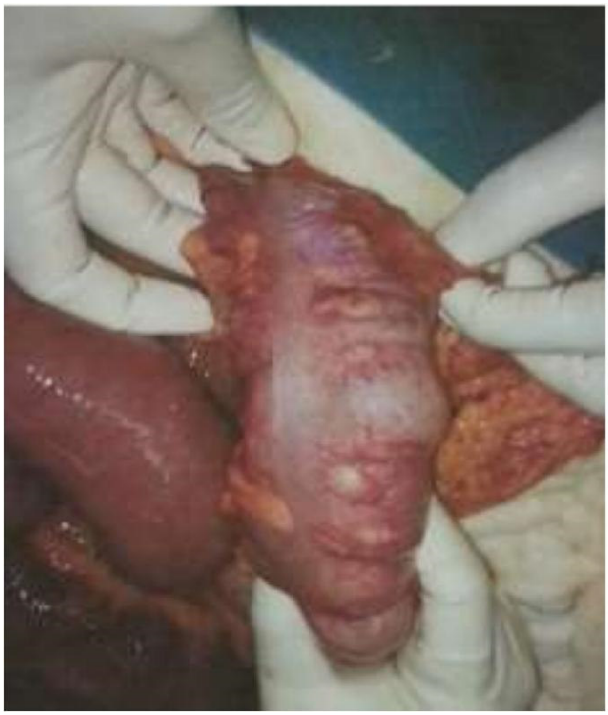

Fig.1: multiple deep multiple serosal punctum extending into mucosa forming a *volcano shaped mucosal with punched-out ulcers over terminal ileum*

Discussion

Behcets' as first described in 1935 by Halusi Behcet is a rare multi-systemic chronic and recurrent immune mediated small vessel vasculitis, often presents with mucous membrane ulceration and ocular problems[6,7]. Occasionally, it may also involves visceral organs such as the musculoskeletal, cardiovascular and neurological systems[7]. The syndrome could also be fatal as it may cause aneurysm where late presentation with ruptured aneurysm is catastrophic [8]. The prevalence of Behcets is more prominent in the eastern continent, especially in Turkey (80-370 cases per 100,000) followed by Asia and the Middle East (13.5-2- cases per 100,000)[7].

Intestinal Behcets is rare within the clinical spectrum of Behcets disease, accounting for only 3-26% of people with Behçet's[9]. It may affects any parts of the bowel from mouth to anus, but the commonest part that will cause problem when affected is at the ilio- caecal junction[9]. This condition will usually present with recurrent ulcer at about 5 year duration before the onset of intestinal problems.

Because of this rarity, this condition is still under reported worldwide. Diagnosis of Behcets Disease is mainly via clinical findings, as to date there are no standard laboratory test or imaging that could pathognomonically diagnose Behcets[6]. Active disease will show evidence of raised inflammatory index whereby, increased levels of C-reactive Proteins and erythrocyte sedimentation rate. Nonetheless, this parameters are non-specific and could also be raised in various other inflammatory or infectious conditions[6,9].

Presenting this case of intestinal obstruction in Malaysia sub-continent where suspicion of vasculitis related disease is rare. The intestinal lesion of Behcets disease usually occurs in 2 forms: mucosal inflammation or ischaemia/infarction. Clinical presentation of Behcets in general is very similar to inflammatory bowel disease,

such as Crohn's Disease, with or without extra-intestinal symptoms[5]. Behcets and Crohns share many extra-intestinal features such as oral lesions, uveitis and arthritis. Other stigmata of Behcets Disease will present at a later stage, thus ileo- colitis conditions should be followed closely looking at stigmatas of Behcets disease[5].

The ulcers in Behcet's disease tend to perforate at multiple sites, so it is necessary to examine the presence of skip lesion during operation [11]. Although surgical intervention is regarded as the preferred choice of treatment, the recommended length of the normal bowel to be resected remains controversial.

Patient with Behcet's disease who underwent surgical treatment has higher chance of recurrence with prevalence ranging from 30% to 80% compare to non-surgical patient[10]. Previous studies suggested that patients with volcano-shape ulcers has significantly higher remission rate and often require reoperation[10].

Up to this date, no well-designed studies have mentioned specific prognostic factor to predict severity of intestinal Behcet's disease despite some claiming the beneficial use C-reactive protein (CRP) levels[6].

Repeated angio-embolization of targeted bleeding artery was the preferable treatment for our patient after presented with bleeding post-surgery. Gradually, with the adjunct of systemic therapy using immunosuppressive medication, the bleeding will be suppressed. There have been anecdotal cases of intra-arterial prednisolone injection therapy given in patients that continue to bleed after surgical resection of the involved segment[12].

Conclusion

The overlap of symptoms and signs between intestinal Behçet's disease and that of inflammatory bowel disease is a definite challenge to practitioners in terms of establishing an accurate diagnosis. The rarity of intestinal Behcets makes it impossible to study this disease in a well-designed research manner. In general, if intestinal obstruction with Behcet's is encountered, the current practice is to resect the unhealthy bowel segment while leaving behind the grossly healthy portions. The decision to anastomose or not lies on the surgeon's clinical decision. Systemic control of the disease is equally as important. Use of steroids and immune modulators may also help prevent recurrence. Good cooperation between surgeons and physicians can ensure reduced morbidity and mortality in these groups of patients.

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