

## Clinical Analysis of Perforated Intestinal Behcet Disease

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**Clinical pattern of perforated intestinal Behcet disease was analyzed in the five patients who underwent surgery in terms of preoperative symptoms, the condition of perforation, the extent of resection and recurrence.**

In the experienced patients, recurrences were included in four of the five patients in spite of treatment. Perforation was based on deep multiple ulcers, characteristic of the punched-out type.

It is emphasized that intestinal Behcet disease is more likely to occur as a catastrophic event of perforation which requires an urgent operation, and more extensive resection is mandatory for prevention of recurrence.

### Introduction

It is well known that Behcet disease involves the whole digestive canals from the esophagus to the rectum by ulcerative lesions. It is referred to as gastrointestinal Behcet which is characteristic of accompanying catastrophic complications of perforation and bleeding. The so-called Behcet syndrome is categorized into entero-, angio-and neuro-Behcet diseases.

We experienced seven patients with perforation by Behcet disease. Of the seven patients, five required emergency operation. In this study, significance of urgent operation for perforation by Behcet disease was clinically evaluated in terms of their prognoses.

### Patients

Table 1 shows the five patients with perforation by Behcet disease. The main syndrome of ulcer in external genitals was seen in all five patients. On the other hand, four out of five presented with aphthous ulcer, two with eye syndrome and one with skin sign, respectively. All complained of right lower abdominal pain. Four of the five gave a history of the past operation for Behcet disease, twice in one and three times in another.

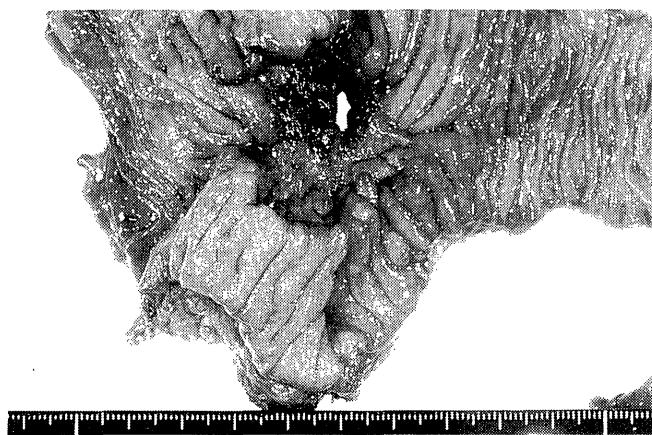
Urgent operations were indicated for clinical signs of peritonitis in four and bowel obstruction in one. The operative procedures were ileocecal resection, ranging from 10 cm to 80 cm long in three and right hemicolectomy, ranging 14 to 23 cm long in two. The resected specimens showed multiple ulcers in four with two and five perforated sites (Fig 1, 2) and in one with undetermined perforated site covered with pus mass accompanying turbid ascites. The postoperative courses were uneventful and all returned to normal social life.

### Discussion

A disease revealing four overt manifestations of buccal aphthous ulceration, eye lesion, skin lesion and genital ulceration is termed as Behcet disease and accessory

**Table 1** Patients who underwent emergency operation with perforated intestinal Behcet disease

Age	Sex	Aphthous ulcer	Genital ulcer	Skin lesion	Eye syndrome	Digestive syndrome	Reoperation	Preoperative diagnosis	Time interval and treatment	The first op resected length	Operative finding (number of perforated sites)
36	M	—	+	—	+	abdominal pain	non	perforation	(—)	ileocecum resection (80cm)	multiple-ulcers (12)
38	F	+	+	—	—	lower abd pain	(+)once	perforation	1y10m steroid	ileocecum resection (10cm)	multiples (9) (2)
40	M	+	+	+	—	lower abd pain	(+)three times	Strangulation	2y4m steroid imuran	r-hemicolectomy (14cm)	turbid ascites (unknown)
55	M	+	+	—	—	lower abd pain with vomiting	(+)twice	Fistul d abcess formation	1y2m (—)	r-hemicolectomy (23cm)	fistula between skin and anastomosis multiple ulcers (5)
55	M	+	+	—	—	lower abd pain	(+)once	ileus	8y8m	ileocecum resection (20cm)	multiple (5) with lineal at anastomosis straugerlation ileus



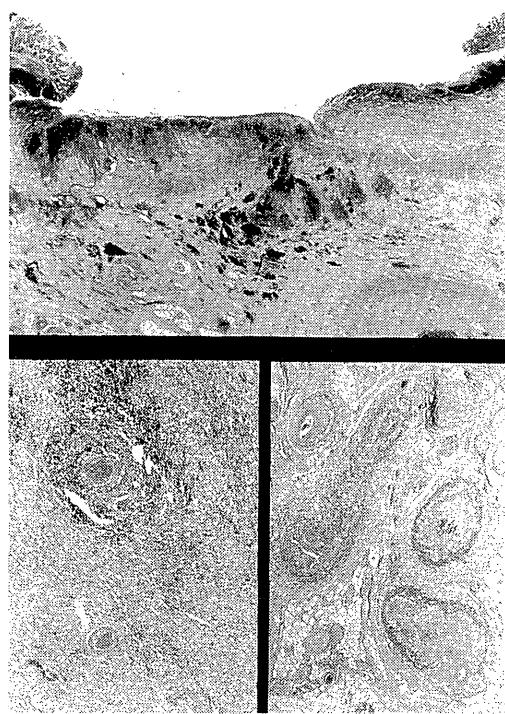
**Fig. 1** Perforated site with punched-out ulcer

syndrome may appear neuro-, vascular-, digestive and strictral-syndromes. It is reported by Japanese Welfare investigation group<sup>1)</sup> that intestinal Behcet disease corresponds to 542 out of a total of 2520 of Behcet disease cases (21.5%). It is characteristic of intestinal ulcer with the site of predilection in ileocecal region as seen in 74.2%<sup>2)</sup> and 80.2%<sup>3)</sup>. Furthermore, one more primary characteristics of Behcet disease is the high risk of perforation as reported by Nakano<sup>4)</sup>. According to Japanese literature, the ileal type is more likely to perforate than the ileocolic type.

It is well known that the intestinal Behcet disease is manifested by pain in ileocolic region as a clinical sign<sup>3)5)</sup>. Surgeons should pay attention to the necessity of urgent operation for Behcet disease suspecting acute appendicitis and acute peritonitis. In addition, the preoperative steroid therapy is a little more cumbersome to postoperatively.

Furthermore, a great concern about the treatment of intestinal Behcet disease is recurrence, as frequent as in 30.2%<sup>3)</sup> to 33.3% of cases<sup>2)</sup>. It is dubious as to whether recurrence includes remaining multiple lesions as well as a deteriorated stage at the first operation<sup>6)</sup> or stimulation by suture material<sup>7)</sup> enough to cause recurrence. Some reports emphasized that environmental contamination, viral infection and genetic background play an important role in the genesis of Behcet disease. The etiology of ulceration is explained by changes in the vascular system due to vascular obstruction and phlebitis in association with autoimmune disease.

Needless to say, the lesions should be identified by endoscopic examination during performing operation. A two stage operation is of great value to assess the lesions



**Fig. 2** Histlogic finding  
upper : punched-out ulcer  
left lower : vasculitis  
right lower : perivasculitis

accurately after eliminating inflammatory changes. There are certain problems with the extent of involving gut to be resected. Baba<sup>3)</sup> warned even a 50 cm resection apart from the lesion gave rise to 17.1% of recurrence.

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