

Surgical Treatment of Intestinal Behçet's Disease

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From 1983 to 1994, we diagnosed 37 cases of intestinal Behçet's disease and performed operations on 26 patients at the Surgical Department of Yonsei University Medical Center. Sixty percent of patients were in their teens and twenties (mean age: 36.4 years). Preoperative diagnosis of Behçet's disease was correctly made in only 8 cases (30%). In 32 cases (86.5%), the lesion was localized regardless of the number of ulcers. A solitary ulcer was observed in 22 cases (60%), while multiple ulcers were present in 15 cases. A recurrence after the initial operation was observed in 12 patients (46.1%) and reoperation was performed 19 times. Fifty percent of recurrence developed within 2 years after each operation. The type of operation, the location of lesion and the number of ulcers did not appear to be related to the recurrence. As a preoperative diagnosis is difficult and the recurrence rate is high, post-operative periodic follow-up with radiography and endoscopy are strongly recommended. At the time of operation, the entire bowel should be examined and bowel resection should include a generous normal resection margin as well as skip lesions.

Key Words: Intestinal Behçet's disease, operation, recurrence

The symptom complexes of recurrent oral ulceration, genital ulceration and ocular inflammation were first reported in 1937 by Hulshi Behçet (Behçet, 1937). Since the initial description, additional involvement of many other organs has been reported including joint, skin, blood vessels, nervous system and gastrointestinal tract (Chajek and Fainaru, 1975). In 1940, Bechguard first described the GI involvement in Behçet's disease and the first fatalities due to GI involvement were reported 4 years later (Saugman-Jensen 1944). In the case of gastrointestinal tract involvement, many patients complained of non-specific symptoms such as nausea, vomiting, and abdominal pain. As the incidence of

ulcerative perforation or hemorrhage is relatively high, the operation should be performed as an emergency without preoperative diagnosis of Behçet's disease. For this reason the resection is restricted, and therefore the incidence of recurrence and complication after surgery is relatively high.

In this study, we reviewed 26 patients with intestinal Behçet's disease who underwent a total of 45 operations and 11 non-operated patients. We attempted to ascertain the postoperative course of intestinal Behçet's disease: the incidence of recurrence, complication and its relation to the types of operation and the clinicopathologic parameters.

MATERIALS AND METHODS

Between November, 1983 and December, 1994, 37 patients were diagnosed with intestinal Behçet's

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disease at Yonsei University College of Medicine. Twenty six of those patients underwent a total of 45 operations, and were followed-up with double-contrast radiography and/or endoscopy of the intestine. We reviewed the clinical records of these patients, including the clinicopathologic parameters, types of surgery, incidence and pattern of recurrence, as well as the incidence and the causes of reoperation.

RESULTS

Patient characteristics

The average age of the patients was 36.4 years, ranging from 9 to 59 years old; 22 patients (59.6%) were in their teens and twenties, 7 patients in their thirties and 5 patients in their forties (Table 1). There was no difference in the ratio of the male (19) to female (18) patients.

Table 1. Age distribution

	Cases No.	%
~10	1	2.7
11~20	2	5.4
21~30	10	27.2
31~40	12	32.4
41~50	7	18.8
51~	5	13.5
Total	37	100.0

Male: Female=19:18

Mean Age: 36.4

Table 2. Clinical manifestation

	Cases No.	%
Abdominal pain	35	94.6
Gastrointestinal bleeding	12	30.6
Diarrhea	9	24.3
Nausea/vomiting	6	16.2
Abdominal mass	6	16.2
Acute abdomen	4	8.1
Fistula formation	2	5.4

Clinical manifestation

The chief complaints of patients were abdominal pain (94.6%), upper gastrointestinal bleeding (30.6%), diarrhea (24.3%), nausea and vomiting (16.2%), abdominal mass (16.2%), acute abdomen (8.1%) and 2 cases of enterocutaneous fistula which were operated at other hospitals (Table 2).

Preoperative diagnosis

The diagnosis was made by barium enema in 10 cases, colonoscopy in 8 cases, and a combination of barium enema and colonoscopy in 9 cases before operation, while 9 cases (24.3%) were diagnosed after emergency operation without preoperative diagnosis (Table 3). The preoperative diagnosis of 26 patients who underwent initial surgery, were 8 with intestinal Behçet's disease (30.8%), 6 with acute appendicitis (23.1%), 4 with peritonitis (15.4%), and one each with intestinal ulcer, malignant neoplasm, inflammatory mass, intestinal obstruction and periappendiceal abscess (Table 4).

Table 3. Diagnosis tools

	Cases No.	%
Barium enema	10	27.1
Colonoscopy	8	21.6
Barium enema + colonoscopy	9	24.3
Colonoscopy + small bowel series	1	2.7
Explo-Iapa	9	24.3

Table 4. Preoperative diagnosis

	Cases No.	%
Behçet's disease	8	30.8
Acute appendicitis	6	23.1
Acute peritonitis	4	15.4
Intestinal ulcer	2	8.2
Inflammatory mass	2	8.2
Malignant neoplasm	2	8.2
Intestinal obstruction	1	4.1
Periappendiceal abscess	1	4.1

Table 5. Types & location of intestinal ulcer

Localized type	32(86.5%)
Terminal ileum	11
Ileocecal area	15
Cecum	2
Ascending colon	4
Diffuse type	5(13.5%)
Terminal Ileum-entire colon	1
Cecum-transverse colon	1
Ascending & transverse colon	1
Descending & proximal sigmoid Colon	1
Terminal Ileum & descending colon	1

Table 6. Types of the first operation

	Cases No.	%
Rt. hemicolectomy	18	69.3
Resection of ileocecal area	3	11.5
Segmental resection of ileum	2	7.7
Ileostomy	1	3.8
Subtotal colectomy	1	3.8
+ileosigmoid colostomy	1	3.8
Segmental resection of terminal ileum	1	3.8
+Lt. hemicolectomy	1	3.8

Types of intestinal ulceration

There were 2 types of ulcer formation; localized and diffuse in intestinal Behçet's disease. In 32 cases (86.5%) of localized type, terminal ileum was involved in 11 cases, ileocecal region in 15, ascending colon in 4 and cecum in 2. In 5 cases (13.5%) of the diffuse type, lesions were noticed in the small bowel and the entire colon diffusely (Table 5). A solitary ulcer was observed in 22 cases (59.5%) and multiple ulcers in 15 cases (40.5%).

Types of operation

Of 37 patients with intestinal Behçet's disease, 26 patients underwent a total of 45 operations. Of these, 12 patients received reoperation more than twice: 6 patients twice, 5 patients three times, and 1 patient four times. The first operative procedures for intestinal Behçet's disease were right hemicolectomy in 18 cases, resection of ileocecal region in 3 cases,

Table 7. Postoperative recurrence

Site	Cases No.
Anastomosis site	14
Ileum	7
Jejunum	2
Colon & ileum	1
Total	24(53.3%)

Table 8. Time interval of recurrence

Interval(yr)	Cases	%
<1	5	20.8
1~2	7	29.1
2~3	3	12.5
3~4	5	20.8
>4	4	16.7

Mean: 27.7 months(7~264)

segmental resection of ileum in 2 cases, and one each with bypass surgery, subtotal colectomy & ileosigmoidcolostomy, segmental resection of ileum and left hemicolectomy (Table 6).

Postoperative recurrence

Postoperative primary recurrence of intestinal Behçet's disease was observed in 12 (46.1%) of 26 patients and subsequently there were 24 recurrences. The most common site of recurrence was the anastomotic site (14 cases), followed by ileum (7 cases), jejunum (2 cases) and colon & ileum (1 case)(Table 7). The period between operation and recurrence ranged from 7 months to 1 year in 5 cases, from 1 year to 2 years in 7 cases, from 2 to 3 years in 3 cases, from 3 to 4 years in 5 cases and over-4 years in 4 cases. The average period of recurrence was 27.7 months with a range of 7 to 264 months (Table 8). In 12 patients with a single lesion, 7 cases (63.6%) experienced recurrence and reoperation was performed in 5 cases; whereas 9 patients (60%) with multiple lesions experienced recurrence and 7 cases underwent reoperation (Table 9).

Table 9. Relationship between number of ulcers recurrence & reoperation

Ulcer	Cases	Recurrence	Reoperation
Solitary	11	7(63.6%)	5
Multiple	15	9(60.0%)	7
Total	26	16(61.5%)	12

Table 10. Indication for reoperation

	Cases No.	%
Panperitonitis due to multiple perforation	7	36.7%
Gastrointestinal bleeding	4	21.1%
enterocutaneous fistula	4	21.1%
Intractable pain with recurrence	4	21.1%

Reoperation

Among 24 recurrences, 19 reoperations were carried out. The causes of reoperations were peritonitis due to bowel perforation in 7 cases (36.7%), gastrointestinal bleeding in 4, enterocutaneous fistula in 4 and intractable pain in 4 cases (Table 10). The types of reoperation were segmental resection of anastomotic site in 13 cases (68.4%), right hemicolectomy in 3, segmental resection of jejunum in 2 and total colectomy in 1 (Table 11).

Postoperative complications

The incidence of complication was 35.6% (16/45); wound infection in 4 cases, enterocutaneous fistula in 4, bleeding in 4 and one each with multiple organ failure, leakage, and short bowel syndrome. Mortality occurred in 3 cases due to sepsis, multiple organ failure and malnutrition.

DISCUSSION

Behçet's disease is a multisystemic disorder characterized by recurrent oral and genital ulceration as

Table 11. Types of reoperation

	Cases No.	%
Segmental resection of anastomosis site	13	68.4
Rt. hemicolectomy	3	15.8
Segmental resection of jejunum	2	10.5
Total colectomy	1	5.3

Table 12. Postoperative complication

	Cases No.	%
Wound infection	4	25
Enterocutaneous fistula	4	25
Postoperative bleeding	4	25
Rectovaginal fistula	1	6.3
Multiple organ failure	1	6.3
Anastomotic leakage	1	6.3
Short bowel syndrome	1	6.3
Total	16	

well as ocular involvement, most likely occurring due to underlying vasculitis. The etiology and pathogenesis of this disease remain obscure. It is considered as an autoimmune disease since vasculitis is the main pathologic lesion and circulating autoantibodies to oral human mucous membrane were found in approximately 50% of the cases (Haralampos and Moutsopoulos, 1997). Recently, diagnostic criteria have been proposed by the International Study Group for Behçet's disease (1990): recurrent oral ulceration plus 2 of the following; recurrent genital ulceration, eye lesion, skin lesion and pathology test. Even though Behçet's disease is widespread throughout the world, there are different incidences and involvement of different organs in patients depending on their country of origin. The prevalence of Behçet's disease ranges from 1:10,000 in Japan to 1:150,000 in North America and Europe. Ocular and central nervous system involvement is extremely common in Turkish patients but much less common in Americans. Similarly, gastrointestinal symptoms are common in Japan but rare in Israel (Chajek and Fainaru, 1975).

Intestinal Behçet's disease is characterized by

deep ulcers, which are more commonly located in the terminal ileum or the ileocecal region and which tend to perforate or penetrate the intestinal wall. The incidence is highest in patients in their twenties, while that of intestinal ulcers requiring surgery is highest in patients in their thirties (Kasahara *et al.* 1981), with more severe cases in males than females. The symptoms of Behçet's disease have been classified into 4 types: complete, incomplete, suspicious, and possible. Intestinal Behçet's disease may occur in any of these types (Baba *et al.* 1976). The ratio of incidence of complete type to other types is 1:1.7 in Japan (Kasahara *et al.* 1981) and it is said that it account for 40% to 78% of all cases of Behçet's disease (Shimizu, 1977). But in our study, there were no cases with complete type, while incomplete type was found in 13 cases, suspicious type in 14 cases and possible type in 10 cases.

Gastrointestinal disease is common in Behçet's disease, occurring in more than 90% of patients in an extensive international project (International Study Group for Behçet's disease, 1990). Many patients complained of abdominal symptoms such as nausea, vomiting and abdominal pain, but ulcerative changes in the intestine were found in 1% or less of all patients with Behçet's disease (Baba *et al.* 1976; Kasahara *et al.* 1981). About 22% of the patients with intestinal Behçet's disease developed symptoms mimicking appendicitis during the clinical course (Kodama, 1977). Since the changes were mainly in the ileocecal region, it was thought to be inevitable that the symptoms are due to appendicitis. In our study, 6 patients (23.1%) were diagnosed with appendicitis before operation.

Intestinal ulcers in Behçet's disease can be confirmed by double-contrast small bowel series or double-contrast barium enema, fiberoptic colonoscopy and biopsy. Intestinal ulcers are characterized by penetration to the serous membrane or the fascia with a marked tendency to undermine such tissues irregularly, and with edema-like swelling and crater-shaped formation around the ulcer margin. The clinical manifestations of Behçet's colitis are easily confused with those of ulcerative and granulomatous colitis. Compared with Crohn's disease, there is a reduced inflammatory process surrounding the ulcer (e.g. less thickening of the intestinal wall and no stenosis of the area). Occasionally, fistula formation

was reported, and ulcers of Behçet's enteritis are multiple, scattered, and tend to perforate easily compared to those of Crohn's disease. Unfortunately, these characteristics of easy bleeding and perforation of intestinal Behçet's disease make diagnosis difficult before operation. However, as patients can usually describe other findings specific to Behçet's disease, careful evaluation of personal history and thorough physical examination are very important in order to obtain an exact diagnosis preoperatively. There were no cases of inflammatory bowel disease before or after operation in this study.

The most common sites of intestinal Behçet's disease were terminal ileum in 44%, ileocecal region in 34% and the cecum in 12%. Multiple ulcers are usually found along the terminal ileum to the cecum, but they may be present at any site throughout the digestive system (Kasahara *et al.* 1981). In this study, the patients with localized ulcers were 32 (86.5%), which involved terminal ileum in 11 cases, ileocecal region in 15, ascending colon in 4, and cecum in 5. Diffuse ulcers were found in 5 cases.

The ulcers in Behçet's disease tend to perforate at multiple sites, so it is necessary to confirm the presence or absence of skip lesions at the time of operation as well as during the preoperative examination (Kasahara *et al.* 1981). But macroscopic findings during operation may not be accurate enough for evaluating the full extent of involvement because this disease is often accompanied by small shallow ulcers or aphthous ulcers (Iida *et al.* 1994). For this reason, Iida *et al.* proposed the use of intraoperative endoscopy. The incidence of postoperative recurrence in 6 cases with intraoperative endoscopy (50%) was lower than in 9 cases without endoscopy (100%).

Although operative treatment is regarded as the first choice due to the high rate of ulcerative perforation, the recommended length of the normal bowel adjacent to the segment which should be resected is controversial. Some have recommended the removal of as much as 60cm or more of the ileum (Baba, 1979), while in another series (Ketch *et al.* 1980), a more conservative approach with the removal of only the grossly-involved bowel seemed warranted. It is said that after resection of 50cm, the risk of recurrence is 10% (Baba *et al.* 1976). A review of surgical cases from Japanese articles

(Kasahara *et al.* 1981) revealed that postoperative recurrence was seen in 38 of 95 cases (40%), whereas in our series, primary recurrence occurred in 12 of 26 patients (46.1%) and these 12 patients received reoperations more than twice. The most common site of recurrence was the anastomotic site, followed by ileum, jejunum, and colon & ileum. The average period between operation and recurrence was 27.7 months, ranging from 7 to 264 months and 50% recurred before 2 years. The types of the first operation, the site of the disease and the numbers of ulcers didn't seem to be related to the recurrence. As in other reports, the most common site of recurrence was the anastomotic site within two years after surgery. Therefore, we suggest a follow-up every 6 months, including radiography and endoscopy, for two years after surgery.

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