A case of Behçet's disease presenting with massive lower gastrointestinal bleeding

Yoğun alt gastrointestinal kanama ile başvuran bir Behçet olgusu

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Behçet's disease, as initially described, is a triad of recurrent oral and genital ulcers and relapsing uveitis. Classified as a systemic vasculitis, it can involve both the arteries and veins of almost any organ. Intestinal Behçet's disease is characterized by deep ulcers, most commonly located in the ileocecal region, with tendency to bleeding and perforation at multiple sites. Here, we report a case of Behçet's disease presenting with lower gastrointestinal bleeding, mesenteric arterial thrombosis and duodenal perforation.

Key words: Behçet's disease, bowel involvement, lower gastrointestinal bleeding

Behçet Hastalığı tekrarlayan oral, genital ülser ve üveit atakları olarak ilk olarak tanımlanmıştır. Herhangi bir organdaki hem arter hem de venleri tutabilen sistemik bir vaskülit olarak sınıflandırılmaktadır. Intestinal Behçet hastalığı en çok ileoçekal bölgeyi tutan, kanamaya ve bir çok yerden perforasyona eğilimi olan derin ülserler ile karakterizedir. Behçet hastalığına bağlı alt gastrointestinal kanama, mezenterik arteriyel tromboz ve duodenal perforasyon ile seyreden bir olguyu sunacağız.

Anahtar kelimeler: Behçet hastalığı, barsak tutulumu, alt gastrointestinal kanama

INTRODUCTION

Behçet disease (BD) was first described in 1937 by a Turkish dermatologist from İstanbul, Dr. Hulusi Behçet, as a triad of symptoms consisting of oral aphthae, genital ulcers and hypopyon uveitis. Classified as a systemic vasculitis, it can involve both the arteries and veins of almost any organ (1, 2). Small-vessel vasculitis accounts for much of the pathological process of the disease, and clinical manifestations of large vessel involvement occur in between 7% and 49% of the patients (6, 8). In Turkey, depending on the geographical differences, the prevalence of BD varies between 2 and 42 cases per 10,000 (3-5). The involvement of the gastrointestinal tract is widely variable in different populations, being more common in Japan (50%–60%) and less common in the Mediterranean basin, including Turkey (0%–5%) (6, 7, 9-11).

Here, we report a case of BD presenting with lower gastrointestinal bleeding, mesenteric arterial thrombosis and duodenal perforation.

CASE REPORT

A 27-year-old man presented to the Emergency Department of our hospital with rectal bleeding and abdominal pain. According to his history, the patient was diagnosed with BD in 1997 and received colchicine 0.5 mg three times daily. One week before his presentation, an ulcer appeared on the right scrotum. Six to seven hours before his presentation, a severe colicky pain on the right upper quadrant of the abdomen started and he subsequently excreted bloody feces; two hours later, he had rectal bleeding (about 500 ml of blood). In addition, the patient had a history of smoking one-half packet of cigarettes per day for four years, but he stopped smoking 6 years ago. He underwent appendectomy in 2002. One year ago, he underwent colonoscopy to determine the causes of anemia and rectal bleeding, but it showed no abnormality.

On physical examination, he had a body temperature of 36.5°C, heart rate of 102/min and arterial

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Manuscript received: 06.08.2007 **Accepted:** 14.11.2007

blood pressure of 90/40 mmHg and he had malaise. He was also pale and his oral mucosa was dry.

There were increased bowel sounds below the navel and tenderness, slight defense and rebound phenomenon on the right quadrant. On digital rectal examination, the glove used for the examination had fresh blood on it on withdrawal. There were tender, crusted ulcers on the outer surface of the scrotum (1x1.5 cm and 0.5x1 cm), surrounded by hyperemia.

On admission, hemoglobin was 4.52 g/dl and hematocrit was 15.12%; thus, blood transfusion and fluid replacement were performed. Plain X-ray of the abdomen taken when the patient was upright was normal. On abdominal ultrasonography, the wall of the terminal ileum was slightly thick and dilated, the wall of the cecum was also slightly thick and there was little fluid surrounding the intestines in the ileo-cecal region. On colonoscopy, the patient had severe pain and there was blood in the lumen. We thus examined the intestines only up to the right hepatic flexura. The examination showed no ulcers or bleeding lesions. To investigate persistent bleeding, the patient was exposed to superior mesenteric angiography. Angiography showed thrombosis in the colic segment of the ileo-colic branch, but not an active bleeding site (Figure 1). The patient underwent right hemilectomy and ileo-colic anastomosis. On operation, the wall of the cecum was thick and the lumen between the ileum and colon was filled with blood. Macroscopic examination of the resected material showed occasional discoloration in the serosa, edema of the mucosa, an ulcer (4x4 cm) and occasional necrosis in a segment 11 cm in length involving the ileo-cecal region. On microscopic examination of the ulcer involving the serosa, there was mixed-typed purulent cell infiltration rich in neutrophils, congestion and capillary proliferation. There was considerable thickening in some arterioles and venules, lymphocyte infiltration in and around the vessel wall and thrombus and recanalization in some vessels on the base of the ulcer (Figures 2, 3). On postoperative day seven, there were intestinal constituents in the drainage, which showed a leakage in the anastomosis, which was repaired. Two days after the repair, the patient had more severe abdominal pain and underwent exploratory operation, which revealed duodenal perforation. Primary repair was made and the perforation was closed with omentoplasty. The patient died from sepsis nine days after omentoplasty.

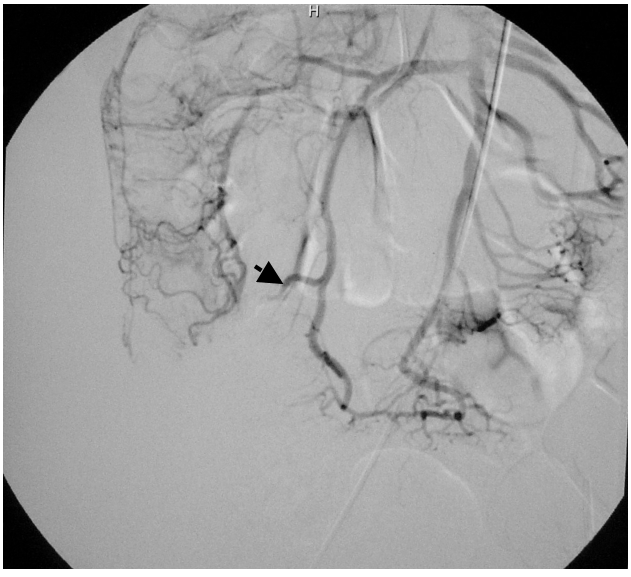

Figure 1. Thrombosis in the colic branch of the ileocolic artery (arrow head) (mesenteric angiography).

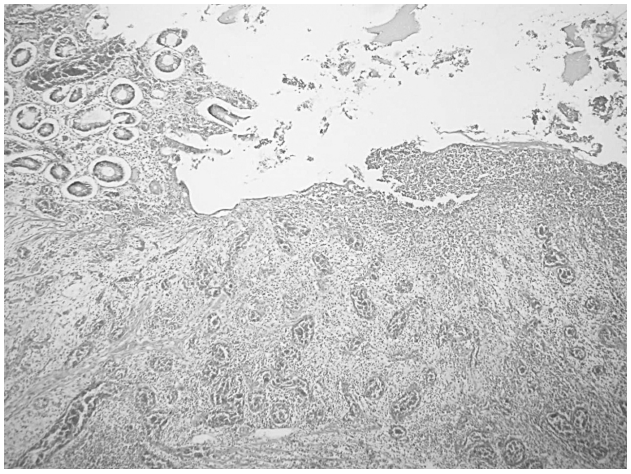

Figure 2. Deep punched-out ulcer in the intestinal wall and infiltration of the ulcer bed with dense polymorphonuclear leukocytes are shown (hematoxylin and eosin x40).

DISCUSSION

Behçet's disease is characterized by relapsing oral and genital ulcers and uveitis. Gastrointestinal involvement in BD was first reported in 1940 (13). The pattern of organ involvement in the disease varies according to regions. In Japan, 50-60% of the patients had gastrointestinal involvement, while it was less frequent in Turkey (7, 9-14, 32). If there is no ocular involvement, the disease is considered incomplete (12). Incomplete BD is more frequently encountered in Japan and Korea, where gastrointestinal involvement is more frequ-

ent. There have been seven cases of BD with gastrointestinal involvement which required surgery. Out of those seven cases, five were incomplete BD (27-31).

The case of BD presented here was classified as incomplete BD since there was no uveitis history.

Forty percent of the patients with BD have gastrointestinal complaints such as abdominal pain, nausea, vomiting, diarrhea with or without blood, and constipation. Intestinal symptoms appear 4.5-6 years after oral aphthous ulcer occurs (14, 18, 33-36). The disease may involve any part of the gastrointestinal tract from the mouth to the rectum (7, 15). However, typical gastrointestinal ulcers appear in about 1-2% of the cases. More than 50% of the cases of BD with gastrointestinal involvement had fistulization, severe bleeding and perforation. It has been reported that gastrointestinal involvement may cause massive gastrointestinal bleeding and perforation in the esophagus, jejunum and colon (21, 24, 37-42, 54). However, it was reported before 1972 that the patients with BD did not have perforation though they did have duodenal involvement (15, 51-53). Işık et al. (26) reported in 2005 that one out of several cases of BD associated with intestinal involvement with the resultant perforation had postoperative leakage of anastomosis followed by perforation of the jejunum.

The case presented here had BD for six years when he presented to our hospital, and he developed scrotal ulcer one week before his presentation. He presented with abdominal pain and rectal bleeding. Following ileo-cecal resection, dehiscence developed in the anastomosis site followed by duodenal perforation.

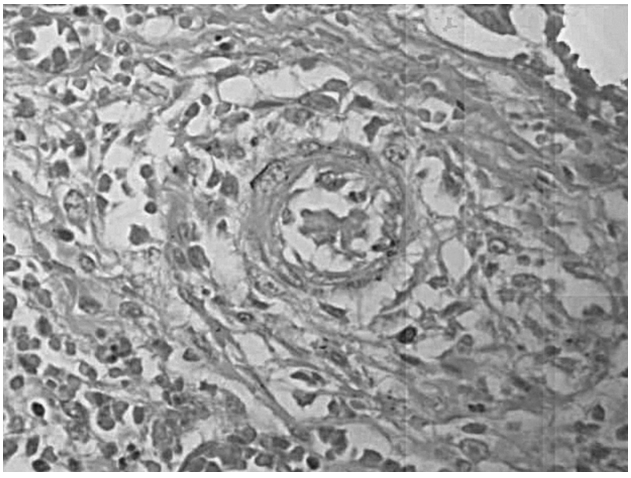

Figure 3. Necrotizing arteriolitis with lymphocyte and polymorphonuclear leukocyte infiltration of the thickened vessel wall (hematoxylin and eosin x400).

Clinical and radiological techniques may not help to differentiate intestinal involvement from Crohn's disease. BD causes large, oval, staples-like, deep ulcers, which tend to extend to the serosa. They are therefore liable to perforation. Presence of vasculitic signs such as non-specific inflammation, necrotizing arteriolitis involving small and medium vessels, lymphocytic venulitis and micro-thromboses and lack of granulomatous lesions on histopathological examinations of the ulcers help in the differential diagnosis of BD (23-26).

In the case presented here, there was a large ulcer in the cecum and pathological examination revealed signs of vasculitic and non-specific inflammation.

Ten percent of the patients with BD accompanied by intestinal involvement require surgical treatment. The complaints most frequently requiring surgical therapy are abdominal pain (92%) and melena (17%). The complications most frequently requiring surgical operations are perforation and bleeding. The most frequently performed surgical therapy is right hemilectomy and ileal resection (18, 31). The recurrence rate after surgery has been reported to be 40%-87.5% and more frequently appears on the anastomosis site. Wound infection, enterocutaneous fistulae and gastrointestinal bleeding are frequent after surgery (15, 17-22, 43). The patients who undergo re-operations for complications and recurrences die from sepsis and multiple-organ failure (20, 27, 28).

In the present case, dehiscence developed in the anastomosis site after the operation and the patient died from sepsis due to infection, which could not be kept under control after the surgical operation to repair the duodenal perforation.

Behçet's disease is a vasculitic disease, which may be characterized by both venous and arterial involvements, but the disease mostly show venous involvement. Venous involvement appears in 24% of the patients, while arterial involvement occurs in only 3%. The rate of arterial occlusion is 0.5%-1.5%. Pulmonary artery aneurysm appears in 1% of the cases in Turkey and is an important cause of mortality (44-46).

Behçet's disease may also involve the abdominal

aorta and large vessels. The mesenteric artery may also be involved, and may cause intraabdominal infarct (47-50). In the case presented here, there was thrombosis in the ileocolic branch of the mesenteric artery, which resulted in ischemic changes in the intestinal serosa.

REFERENCES

1. Behçet H. Uber rezidivierende Aphthose, durch ein Virus verursachte Geschwure am Mund, am Auge und an den Genitalien. Dermatol Wochenschr 1937; 105: 1152-7.
2. Üstün C. A famous Turkish dermatologist, Dr. Hulusi Behçet. Eur J Dermatol 2002; 12: 469-70.
3. Idil A, Gürler A, Boyvat A, et al. The prevalence of Behçet's disease above the age of 10 years. The results of a pilot study conducted at the Park Primary Health Care Center in Ankara, Turkey. Ophthalmic Epidemiol 2002; 9: 325-31.
4. Azizlerli G, Köse AA, Sarica R, et al. Prevalence of Behçet's disease in Istanbul, Turkey. Int J Dermatol 2003; 42: 803-6.
5. Çakir N, Derviş E, Benian O, et al. Prevalence of Behçet's disease in rural western Turkey: a preliminary report. Clin Exp Rheumatol 2004; 22: 53-5.
6. Sakane T, Takeno M, Suzuki N, Inaba G. Behcet's disease. New Engl J Med 1999; 341: 1284-91.
7. Yurdakul S, Tuzuner N, Yurdakul I, et al. Gastrointestinal involvement in Behçet's syndrome: a controlled study. Ann Rheum Dis 1996; 55: 208-10.
8. Kural-Seyahi E, Fresko I, Seyahi N, et al. The long-term mortality and morbidity of Behcet syndrome: a 2-decade outcome survey of 387 patients followed at a dedicated center. Medicine (Baltimore) 2003; 82: 60-76.
9. Shimizu T, Ehrlich GE, Inaba G, Hayashi K. Behçet disease (Behçet syndrome). Semin Arthritis Rheum 1979; 8: 223-60.
10. Yazici H, Tuzun Y, Pazarh H, et al. Behcet's disease as seen in Turkey. Haematologica (Pavia) 1980; 65: 381-3.
11. Dilsen N, Konice M, Ovul C, eds. Clinical evaluation of 106 cases of Behcet's disease. Behcet's Disease. Proceedings of an International Symposium on Behcet's Disease. Amsterdam: Excerpta Medica, 1979; 124-9.
12. Morita H, Kawai S, Kobori O, et al. Incomplete form of Behcet's colitis in Japan: is it a distinct entity? Am J Gastroenterol 1995; 90: 523-4.
13. Bechguard P. Et tilfaelde af recidiverende pathos stomatitis ledsaged at conjunctivitis of ulcerationer paa genitalia of hud. Ugeskr Laeger 1940; 102: 1019-23.
14. Oshima Y, Shimizu T, Yokohari R, et al. Clinical studies on Behçet's syndrome. Ann Rheum Dis 1963; 22: 36-45.
15. Kasahara Y, Tanaka S, Nihino M, et al. Intestinal involvement in Behçet's disease: review of 136 surgical cases in the Japanese literature. Dis Colon Rectum 1981; 24: 103-6.
16. Bradbury AW, Milne AA, Murie JA. Surgical aspects of Behçet's disease. Br J Surg 1994; 81: 1712-21.
17. Iida M, Kobayashi H, Matsumoto T, et al. Postoperative recurrence in patients with intestinal Behçet's disease. Dis Colon Rectum 1994; 37: 16-21.
18. Bayraktar Y, Ozaslan E, Van Thiel DH. Gastrointestinal manifestations of Behcet's disease. J Clin Gastroenterol 2000; 30: 144-54.
19. Naganuma M, Iwao Y, Inoue N, et al. Analysis of clinical course and long-term prognosis of surgical and nonsurgical patients with intestinal Behçet's disease. Am J Gastroenterol 2000; 95: 2848-51.
20. Lee KS, Kim SJ, Lee BC, et al. Surgical treatment of intestinal Behcet's diseases. Yonsei Med J 1997; 38: 455-60.
21. Smith JA, Siddiqui D. Intestinal Behçet's disease presenting as a massive acute lower gastrointestinal bleed. Dig Dis Sci 2002; 47: 517-21.
22. Pirildar T, Keser G, Tunc E, et al. An unusual presentation of Behcet's disease: intestinal perforation. Clin Rheumatol 2001; 20: 61-2.
23. Arora A, Tandon RK, Jain P, et al. Massive lower gastrointestinal bleeding as a presenting feature of Behcet's disease. Trop Gastroenterol 1989; 10: 56-61.
24. Hamza M, Eleuch M, Kchir N, Zitouna M. Ileal perforation in 3 cases of Behcet disease. Ann Med Interne (Paris) 1994; 145: 99-102.
25. Hizawa K, Kai H, Hasegawa H, et al. Ileal perforation in diffuse intestinal Behcet disease: report of a case. Surg Today 2002; 32: 167-9.
26. Isik B, Ara C, Kirimlioglu H, et al. Single or multiple perforations with varying locations as a complication of intestinal Behçet's disease: report of three cases. Scand J Gastroenterol 2005; 40: 599-603.
27. Ng FH, Cheung TC, Chow KC, et al. Repeated intestinal perforation caused by an incomplete form of Behçet's syndrome. J Gastroenterol Hepatol 2001; 16: 935-9.
28. Yildirim M, Erkan N, Bayam E, Sahin T. An uneventful complication of Behcet's disease: intestinal perforation. Chir Gastroenterol 2006; 22: 52-4.
29. Shimizu T, Saito Y. Clinical studies on Behcet's syndrome as a systemic disease. Saishin Igaku 1971; 26: 465-73.
30. Bang D, Yoon KH, Chung HG, et al. Epidemiological and clinical features of Behcet's disease in Korea. Yonsei Med J 1997; 38: 428-36.
31. Sayek I, Aran O, Uzunalimoglu B, Hersek E. Intestinal Behcet's disease: surgical experience in seven cases. Hepatogastroenterology 1991; 38: 81-3.
32. Chung YM, Yeh TS, Sheu MM, et al. Behcet's disease with ocular involvement in Taiwan: a joint survey of six major ophthalmological departments. J Formos Med Assoc 1990; 89: 413-7.
33. Bradbury AW, Milne AA, Muric JA. Surgical aspects of Behcet's disease. Br J Surg 1994; 81: 1712-21.
34. Roge J. Behcet's syndrome and the digestive tract. J Mal Vasc 1988; 13: 235-9.
35. Zhang L, Zhao G, Dai X, et al. Manifestations of Behcet's disease in the digestive tract. Chin Med J (Engl) 1997; 110: 195-7.
36. Roge J. Intestinal involvement in Behcet's syndrome. Presse Med 1985; 14: 537-41.
37. Lee RG. Case report: the colitis of Behçet's syndrome. Am J Surg Pathol 1986; 10: 888-93.
38. Brodie TE, Ochsner JL. Behçet's syndrome with ulcerative oesophagitis: report of the first case. Thorax 1973; 28: 637-40.
39. Toynton SC. Behçet's syndrome: an unusual cause of ileal perforation. J Clin Gastroenterol 1994; 19: 84-5.
40. Empey DW. Rectal and colonic ulceration in Behçet's disease. Br J Surg 1972; 59: 173-5.
41. Masugi J, Matsui T, Fujimori T, Maeda S. A case of Behçet's disease with multiple longitudinal ulcers all over the colon. Am J Gastroenterol 1994; 89: 778-80.

42. Hizawa K, Kai H, Hasegawa H, et al. Ileal perforation in diffuse intestinal Behçet disease: report of a case. Surg Today 2002; 32: 167-9.
43. Turan M, Sen M, Koyuncu A, et al. Sigmoid colon perforation as an unusual complication of Behçet's syndrome: report of a case. Surg Today 2003; 33: 383-6.
44. Zhang L, Zhao G, Dai X, et al. Manifestation of Behcet's disease in the digestive tract. Chin Med J 1997; 110: 195-7 (in English).
45. Müftüoğlu AU, Yurdakul S, Yazici H, et al. Vascular involvement in Behçet's disease - a review of 129 cases. In: Lehner T, Barnes CG, eds. Recent advances in Behçet's disease. London: Royal Society of Medicine Services Ltd., 1986; 256-60.
46. Koc Y, Güllü I, Akpek G, et al. Vascular involvement in BD. J Rheumatol 1992; 19: 402-10.
47. Hamuryudan V, Yurdakul S, Moral F, et al. Pulmonary arterial aneurysms in Behcet's syndrome: a report of 24 cases. Br J Rheumatol 1994; 33: 48-51.
48. Bayraktar Y, Balkanci F, Kansu E, et al. Cavernous transformation of the portal vein: a common manifestation of Behcet's disease. Am J Gastroenterol 1995; 90: 1476-9.
49. Bizmuth E, Hadenque A, Hammel P, et al. Hepatic vein thrombosis in Behcet's disease. Hepatology 1990; 11: 968.
50. Chubachi A, Saitoh K, Imai H, et al. Case report: intestinal infarction after an aneurysmal occlusion of superior mesenteric artery in a patient with Behcet's disease. Am J Med Sci 1993; 306: 376-8.
51. Bayraktar Y, Soylu AR, Balkancı F, et al. Arterial thrombosis leading to intestinal infarction in a patient with Behçet's disease associated with protein C deficiency. Am J Gastroenterol 1998; 93: 2556-8.
52. Chong VF, Pathmanathan R. Familial Behcet's syndrome with intestinal involvement - case reports and a review of the literature. Ann Acad Med 1993; 22: 807-10.
53. Ozenc A, Bayraktar Y, Baykal A. Pyloric stenosis with esophageal involvement in Behcet's syndrome. Am J Gastroenterol 1990; 85: 727-8.
54. Satake K, Yada K, Ikehara T, et al. Pyloric stenosis: an unusual complication of Behcet's disease. Am J Gastroenterol 1986; 81: 816-8.
55. Fujita H, Kiriyama M, Kawamura T, et al. Massive hemorrhage in a patient with intestinal Behçet's disease: report of a case. Surg Today 2002; 32: 378-82.