

Colonic Perforation Due to Behçet's Disease: A Rare Complication and Delayed Diagnosis

Behçet Hastalığına Bağlı Kolon Perforasyonu: Ender Bir Komplikasyon ve Gecikmiş Tanı

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ÖZET

Behçet hastalığı birçok organı ilgilendiren otoimmün bir kronik vaskülit hastalığıdır. İlk kez ağız ülseri, genital ülser ve uveit üçlüsü olarak tanımlanmıştır. Ancak vücudun çeşitli sistemlerini ilgilendirebilir. En fazla görüldüğü yer tarihi "İpek Yolu" olarak bilinen ve Uzak Doğu'dan Akdeniz'e kadar uzanan bölgedir. Dominant olarak etkilenen sistemler ve ortaya çıkan belirtiler bu coğrafik dağılımda farklılıklar gösterir. Uzak Doğu'da barsak tutulumu sıktır ve ileoçekal bölge en fazla etkilenmektedir. Türkiye'nin içinde bulunduğu coğrafik konumda barsak belirtileri oldukça ender olmasına karşın; barsak tutulumuna bağlı, bir çıkan kolon perforasyon olgusunu sunmayı amaçladık.

Anahtar Kelimeler: Behçet Hastalığı, Kolon Perforasyonu

ABSTRACT

Behçet's disease is an autoimmune chronic vasculitic multisystem disorder. It was first described as a triad of oral ulceration, genital ulceration and uveitis. But it may invade various systems in the body. It is prevalent along ancient "silk road" from Far East to Mediterranean. Predominance of affected systems and manifestations are different in this geographical distribution. Intestinal involvement is common in the Far East and the ileocaecal region is the most commonly affected site. Although intestinal manifestations are rare in the Turkey's geographical area, we aim at presenting a case of colonic perforation based on bowel involvement.

Key words: Behçet's Disease, Colonic Perforation

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Introduction

Behçet's Disease (BD) is a vasculitic multisystem disorder. It was first described by the Turkish dermatologist, Hulusi Behçet in 1937 as a triad of oral ulceration, genital ulceration and ocular inflammation.¹ Recently, the Internationally Study Group (ISG) criteria and the International Criteria for Behçet's Disease (ICBD) were created in 1990 and 2006 respectively.^{2,3} For ISG, oral aphthosis is mandatory. The presence of any two of the following (genital aphthosis, skin lesions, eye lesions, and positive pathergy test) will diagnose/classify the patient as BD. For ICBD, vascular lesions were added, while oral aphthosis was not mandatory anymore. Getting 3 or more points diagnose/classify the patient as BD (genital aphthosis 2 points, eye lesions 2 points, and the remaining each one point). Although the etiology is not clearly defined, it is likely to be due to an autoimmune vasculitis in genetically predisposed individuals triggered by an infectious agent or other antigens.⁴ The prevalence rate is 0.3/100,00 in the United States while it is 1/10,000 in Japan.⁵ BD is prevalent along ancient "silk road" from Far East to Mediterranean. Predominance of affected systems and manifestations are different in this geographical distribution. Intestinal involvement is common in the Far East and the ileocaecal region is the most commonly affected site. There are deep mucosal ulcers and they may perforate.^{6,7}

Case Presentation

A 41-year-old woman was admitted to the emergency department for abdominal pain and nausea. Her blood pressure, pulse rate, respiratory rate and body temperature were normal. Physical examination revealed typical abdominal signs for acute appendicitis. She had abdominal tenderness and rebound in the right lower quadrant and also abdominal scars due to previous operations. We learned that she had laparoscopic cholecystectomy and umbilical hernia repair operations 3 and 5 years ago respectively. She and her relatives did not give exact and healthy information. We learnt that she was diagnosed with BD 10 years ago. She had recurrent, painful oral and genital ulcers and positive pathergy test but no ocular signs. She had been treated with oral colchicum (0,5 mg / day) and dexamethasone (5 mg / day) in an outpatient clinic. She had no family history of BD. The results of laboratory tests showed

15,400/ μ l and neutrophil rich leucocytosis, other biochemical analyzes and coagulation parameters were normal. There was no pathologic sign on plain thorax and abdominal x-ray films. There was no free fluid in peritoneal cavity and there was no positive sign for acute appendicitis in abdominal ultrasonography. Because she had acute abdomen signs, we decided exploratory laparotomy. Under intratracheal general anaesthesia, abdomen was opened with midline incision. There was 3cm x 2cm dimensions fascial defect and prolene mesh adapted on lay. The mesh was excised. Abdominal exploration revealed acute appendicitis and there was no other pathologic sign for acute abdomen. Appendectomy procedure was performed and abdomen was closed without drain. Dermatology, rheumatology and ophthalmology consultations were done. There was neither ocular nor active skin lesion. She was discharged on postoperative second day without complication. Continuation of medical treatment was advised on the results of consultations. There was acute appendicitis and lymphoid hyperplasia on histopathology report. Three days after discharging, she was admitted to emergency department again for acute abdominal signs. Her vital signs were normal. This time, her laboratory tests showed 21,700/ μ l WBC, mild elevation of BUN and creatinin level. Abdominal ultrasound revealed free liquid in the peritoneal cavity including whole recesses which was measured 75 mm thickness in the deeper area. She was re-operated. About 500 ml purulent fluid aspirated from abdominal cavity. Appendectomy stump was normal. Fluid sample was taken for bacteriologic

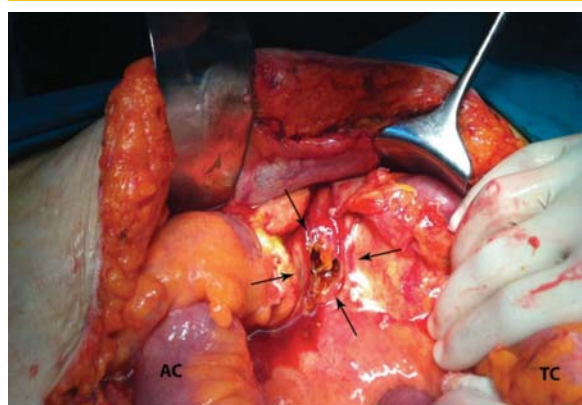


Figure 1. Multiple punctuated perforations in an area of ascending colon, near the hepatic flexure shown by arrows. AC: ascending colon, TC: transverse colon.

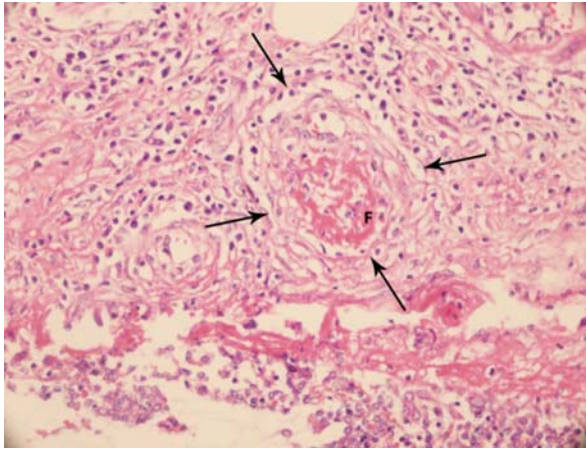


Figure 2. Fibrin deposition and inflammatory cell infiltration on the vessel wall. Arrows mark the inflammatory cells. F: Fibrin. HE X 400.

culture. During the continuation of exploration, intestinal and colonic adhesions between the loops were seen. We found multiple punctuated perforations in an area having 5 cm diameter of ascending colon near the hepatic flexure (figure 1). Abdominal cavity was irrigated with 6 litres of warm serum physiologically. Right hemicolectomy and end to side ileotransversostomy was performed. Two abdominal drains were inserted to right paracolic and retrovesicale regions and one to subcutaneous tissue, and then abdomen was closed. Histopathologic examination revealed acute and chronic inflammatory vasculitic changes (figure 2) and mucosal aphthous ulcerative areas (figure 3) on the specimen. On the postoperative seventh day (in the second hospitalisation period), we decided on abdominal

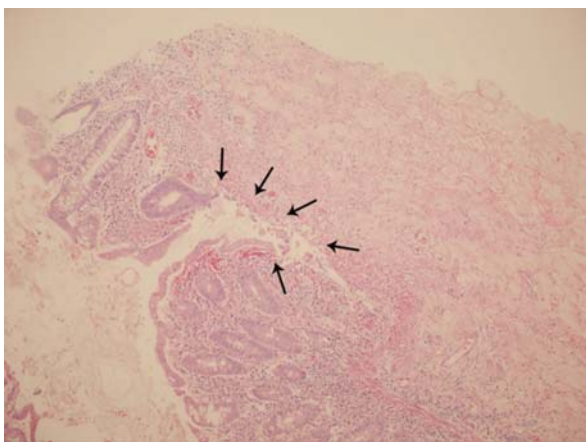


Figure 3. Aphthous ulcerative area on the colonic mucosa shown by arrows. HE X 100.

exploration; because there was intestinal content in the abdominal drainage material and ultrasonography revealed free fluid in peritoneal cavity. On operation, we found 500 ml faecal fluid in peritoneal cavity. After aspiration and irrigation, dehiscence and leakage from anastomosis were revealed. After debridement of necrotic tissues, colonic closure and terminal ileostomy were done. Abdominal closure was postponed; open abdomen procedure was applied with Bogota Bag. Peritoneal cavity was irrigated, cleaned three times in different days. After determination of negative peritoneal culture and it was proven that no residual material in the peritoneal cavity by computed tomography; abdominal closure was finished on postoperative 17th day with skin sutures only. In the second hospitalization period, the patient stayed in the intensive care unit for 7 days and then she was transferred to ward and discharged on postoperative 44th day. In this period, she needed 6 units of fresh frozen plasma and 7 units of erythrocyte suspension. These blood products were given in postoperative 48 hours period after colonic resection and ileotransversostomy procedure. According to wound and blood culture results, antibiotic administrations were begun and continued in consideration of infection specialist's advise. In this period, the patient was treated with imipenem (26 days), imipenem + colistin + flucanazole (11 days) + beta lactamase (7 days). Colchicum (0.5 mg tablet three times a day per oral) and dexamethasone administration (20 mg/day intravenously for 5 days and soon 10 mg tablet twice a day per oral) was begun at the starting time of oral intake on 28th day of second hospitalization.

Discussion

The disease mainly affects commonly young adults between the second and fourth decades of life.^{3,5} In 1990, the ISG defined the diagnostic criteria based on a survey of 913 patients recruited from 12 centres in 7 countries.² In 2006, the ICBID were presented to the International Conference of Behçet's Disease in Lisbon (Portugal).³ According to these, there are major and minor manifestations. Also, a specific organ classification that includes mucocutaneous, arthritic, neurological and ocular types has been developed. Intestinal involvement may be a feature of any of these types.⁸ If intestinal ulcers are confirmed by radiography, endoscopy or

surgery, the disease is referred to as “intestinal” BD.⁹ The frequency of intestinal involvement in patients with BD is known to be 0-60%, with geographic differences. While symptomatic intestinal involvement is rare in Mediterranean patients, it is more common in East Asian patients. Gastrointestinal involvement is uncommon in Turkish patients with an incidence ranging from 0 to 5% [10]. In contrast, this ratio is higher in Chinese (15%) and Japanese (50-60%).^{11,12} Our presented case was a 41-year-old woman. She had BD for ten years and she had perforated ascending colonic area in an acute abdomen picture.

Symptomatic gastrointestinal involvement can result in abdominal pain, diarrhea, bleeding and sometimes perforation. Gastrointestinal BD shows a wide range of sites of involvement and types of lesions.^{6,13} Inflammatory bowel diseases should be kept in mind in the differential diagnosis of intestinal BD. Although International Study Group criteria for BD accurately distinguishes between BD and Crohn's Disease, there are some common features.^{2,9,14} Extraintestinal manifestations are similar in both diseases such as arthritis, uveitis. Also, rectal sparing is common for both. Intestinal ulcers of BD are usually round or oval with a “punched out” appearance mainly located in the ileocecal region and deeper than those in ulcerative colitis. They may rarely display granulomatous features and longitudinal appearance. The intestinal lesions of BD are manifestations, mucosal inflammation or ischemia/infarction, small and large vessel involvement, respectively.¹⁵ Our patient showed complex clinical features. First, she had acute appendicitis. Second, she had acute abdomen syndrome due to perforated colon in the recovery period of appendectomy. Most likely, perforations occurred due to mucosal infarctions secondary to vessel involvement of BD. There are two types of intestinal ulcers in BD: localized and diffuse. According to literature review of Turan et al. , the localized types predominantly located in ileocecal region with 76% ratio.⁹ The other parts of gastrointestinal tracts were reported as cases in literature.^{4,8,9,12,15,16,17} Some of them contained simultaneous perforations from different parts of the tract or from entire intestine/colon.^{11,12,18,19} Adorian et al. reported a case

of Behçet's disease complicated by severe toxic megacolon.¹³ The presented case had perforated appendix and soon multiple punctuated perforations in an area of 5 cm diameter of ascending colon near the hepatic flexure in a week.

Medical treatment of BD is largely empirical, and a multidisciplinary approach is preferred. This multidisciplinary team should involve specialists in dermatology, rheumatology, immunology, ophthalmology, gastroenterology and others. Medical treatment for intestinal BD remains empiric and palliative. Corticosteroids, colchicum, sulfasalazine, azathioprine, thalidomide, pentoxifylline and antitumor necrosis factor monoclonal antibody (infliximab), have all been used to treat BD.^{4,9} However, Demirel et al. reported a case that was treated successfully with conservative medical therapy after descending colon perforation.¹⁵ Our patient was under colchicum and corticosteroid medical treatment for ten years. This regimen continued after discharging from hospital.

In the case of surgery for perforated intestinal BD, the length of normal bowel adjacent to ulcerated segment, which should be resected, is controversial. Some authors recommend removal of intestine as much as 60 cm or more, while others advocate a more conservative approach with removal of only the grossly involved area.^{9,14} On the other hand, the postoperative recurrence rate for intestinal complications in BD was found very high (40-87.5%) in Kasahara's and Işık's studies, the rates of patients requiring reoperation were 37.5-47%.^{20,21} In the light of these data, conservative resections may be enough. So, we did the conservative resections in margins allowed by surgical technique.

Conclusions

In conclusion, colon perforation is a rare complication of vasculitis secondary to BD. Unfortunately, our patient had complex clinical manifestations. However, peritoneal cavity was explored carefully through the big midline incision probable minor/microscopic ascending colon perforations might be hidden behind the acute appendicitis manifestations. So, it was possible to diagnose only in faecal contamination clinic by delaying.

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