Intestinal Perforation due to Miliary Tuberculosis in a Patient with Behçet's Disease

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Abstract

Gastrointestinal manifestations of both Behçet's disease and intestinal tuberculosis include the signs and symptoms of abdominal pain, weight loss, fever, vomiting, diarrhea and palpable mass in the right lower quadrant.

We report the case of a male patient with Behçet's disease who had multiple ileal perforations due to miliary tuberculosis. It was suspected that the perforations were due to intestinal manifestation of Behçet's disease, but the final pathology report and chest X-ray one week after surgery demonstrated the presence of miliary tuberculosis. To our knowledge, this is the first reported case of Behçet's disease with intestinal perforation due to miliary tuberculosis.

Key Words: Behçet's disease, intestinal perforation, miliary tuberculosis.

Introduction

Behçet's disease (BD) is a type of systemic vasculitis characterized by recurrent oral and genital ulceration and ocular inflammation. Since its original description, the additional involvement of many other organs, including joints, skin, blood vessels, nervous system, and gastrointestinal tract, has been reported (1).

Since the etiology of BD remains unknown, its treatment is usually symptomatic and palliative. Patients are usually treated with antibiotic, antiviral, corticosteroid and immunosuppressive agents according to the symptoms (2, 3). Immunosuppressive and cytotoxic agents are used for more severe involvement (3), but this causes patients to be more susceptible to opportunistic infections.

We describe a patient with Behçet's disease who presented with military tuberculosis, which most probably developed secondary to steroid use.

Case

A 34-year-old man was admitted to our hospital complaining of acute abdominal pain, fever, dyspnea and poor appetite. Physical examination revealed abdominal tenderness and rebound. Chest X-ray showed normal lungs and free air under the right diaphragm (Fig. 1). The patient had been on corticosteroid treatment for BD for 5 years, in doses ranging from 5 to 40 mg. He was given colchicine for 1 year and discontinued 2 years ago. On rare occasions, he used naproxen for his joint pains. He did not report use of aspirin.

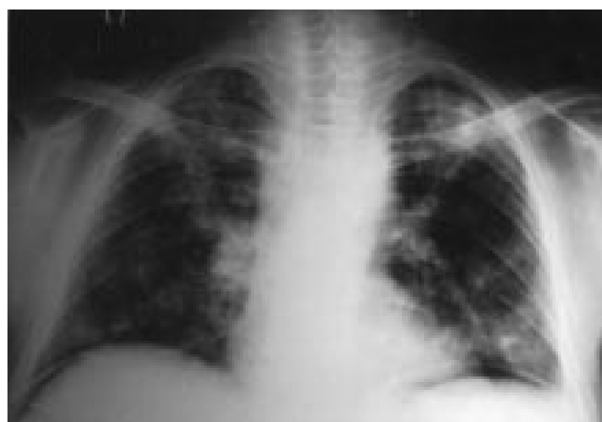

Fig. 1. Free air under the right diaphragm.

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The patient, who was operated on, had been diagnosed as having intestinal perforation due to BD. During the exploratory laparotomy, three separate perforations, in a 35 cm segment of small bowel and 40 cm proximal to the caecum, were observed (Fig. 2). The involved segment was resected and an ileoileostomy was performed. After the operation, the patient was taken to the intensive care unit. His first three postoperative days were uneventful; however, a fecal fistula developed on the fourth postoperative day. The patient was taken to the OR. During the exploratory laparotomy, a few intestinal collections around the anastomosis and a 1 cm anastomotic dehiscence were found. We did an abdominal "washout" and the anastomosis was converted to an end ileostomy. An X-ray of the chest, taken on postoperative day five, showed multinodular infiltration in both lungs. The patient died on the seventh postoperative day, due to sepsis. Microscopic examination of the specimen revealed mucosal ulceration and necrotizing granulomas in the submucosal stroma and muscularis, as well as in the Payer's patches (Fig. 3). Acid-fast bacilli were present in the area of the caseating granuloma.

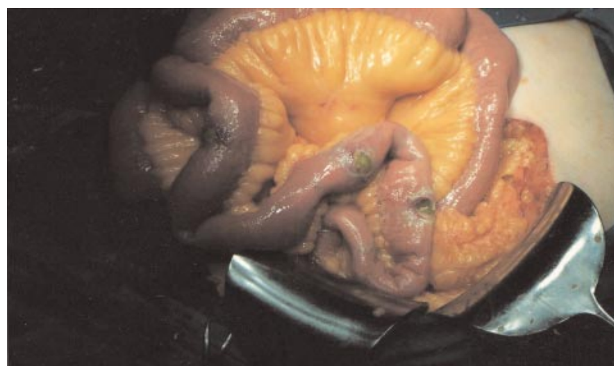

Fig. 2. Three separate perforations at the terminal ileum.

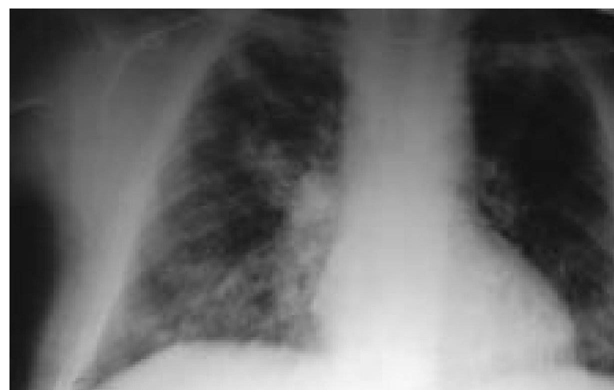

Fig. 3. Multinodular infiltration in both lungs.

Discussion

Tuberculosis of the gastrointestinal tract is usually secondary to pulmonary infection. A rare cause of gastrointestinal tuberculosis is bowel involvement during miliary tuberculosis by hematogenous spread (4, 5).

Pulmonary tuberculosis may rarely occur in patients with BD and is an important differential diagnosis, although less common than the pulmonary disease due to underlying vasculitis that is seen in BD (6, 7). There are a number of possible factors that may increase the risk of tuberculosis in patients with BD: the disease itself may produce a defect in cell-mediated immunity, which increases the individual's susceptibility to tuberculosis (6); the immunosuppressive drugs, particularly corticosteroids, used in BD, predispose these patients to developing tuberculosis (8). A common genetic background, HLA-B5, to both diseases is well known (9).

Gastrointestinal manifestations of both BD and intestinal tuberculosis have similar clinical and histological findings, and they usually affect the same segment of the intestine. The most common signs and symptoms of these two diseases are abdominal pain, weight loss, fever, fatigue, loss of appetite, vomiting, diarrhea and palpable mass in the right lower quadrant (10, 11).

The typical internal manifestation of both diseases is round or oval ulcers that can be seen throughout the alimentary tract, but most commonly in the terminal ileum and the caecum (12, 13).

Intestinal perforation due to military tuberculosis is extremely rare and only two cases have been reported (10, 11). Although any part of the intestine may perforate, the most frequent site is the distal ileum. A single perforation is present in 90% of cases. Multiple perforations are uncommon (14, 15). Free perforation is uncommon in intestinal tuberculosis due to reactive thickening of the peritoneum and the formation of adhesions to adjacent tissues (16).

Morbidity and mortality of the two diseases are high. The overall mortality rate of perforation ranges from 30–67%, even when there has been surgical treatment. The mortality is higher for patients who have had two or more perforations (14, 17, 18).

Surgical treatment of these lesions varies from simple drainage of peritoneal or pelvic abscesses to resection of the involved bowel. The most effective treatment is resection of the diseased segment with an end-to-end anastomosis, but the final decision depends upon the general condition of the patient, extent of the disease and presence or absence of strictures distal to the perforation (19).

Toxemia, wound dehiscence, adhesive obstruction and fecal fistula have been usual complications in both diseases (19).

Although intestinal perforation due to both BD and military tuberculosis is uncommon, in this patient, tuberculous perforation, perhaps due to a predisposition after steroid use, was observed.

References

1. Oshima Y, Shimizu T, Yokohari R, et al. Clinical studies on Behçet's syndrome, with special reference to 100 personal cases [Japanese]. Naika 1962; 9:701 – 714..
2. O'Duffy JD, Carney JA, Deodhar S. Behçet's disease. Report of 10 cases, 3 with new manifestations. Ann Intern Med 1971; 75(4):561 – 570.
3. Mamo JG, Azzam SA. Treatment of Behçet's disease with chlorambucil. Arch Ophthalmol 1970; 84(4):446 – 450.
4. Munt PW. Miliary tuberculosis in the chemotherapy era: with a clinical review in 69 American adults. Medicine (Baltimore) 1972; 51(2):139 – 155.
5. Hiatt GA. Miliary tuberculosis with ileocecal involvement diagnosed by colonoscopy. JAMA 1978; 240(6):561 – 562.
6. Efthimiou J, Hay PE, Spiro SG, Lane DJ. Pulmonary tuberculosis in Behçet's syndrome. Br J Dis Chest 1988; 82(3):300 – 304.
7. Lehner T, Barnes CG. Criteria for diagnosis and classification of Behçet's syndrome. In: Lehner T, Barnes CG, editors. Behcet's syndrome: clinical and immunological features. London (UK): Academic Press; 1979. pp. 4 – 5.
8. Reynolds JEF. Corticosteroids. In: Reynolds JEF, editor. Martindale: the extra pharmacopaeia. 28th ed. London (UK): The Pharmaceutical Press; 1982. pp. 447 – 448.
9. Hwang CH, Khan S, Ende N, et al. The HLA-A, -B, and -DR phenotypes and tuberculosis. Am Rev Respir Dis 1985; 132(2):382 – 385.
10. Mert A, Bilir M, Ozaras R, et al. A rare complication of miliary tuberculosis: intestinal perforation. Acta Chir Belg 2001; 101(1):42 – 45.
11. Kim JH, Langston AA, Gallis HA. Miliary tuberculosis: epidemiology, clinical manifestations, diagnosis, and outcome. Rev Infect Dis 1990; 12(4):583 – 590.
12. Yurdakul S, Tuzuner N, Yurdakul I, et al. Gastrointestinal involvement in Behçet's syndrome: a controlled study. Ann Rheum Dis 1996; 55(3):208 – 210.
13. Kasahara Y, Tanaka S, Nishino M, et al. Intestinal involvement in Behçet's disease: review of 136 surgical cases in the Japanese literature. Dis Colon Rectum 1981; 24(2):103 – 106.
14. Bhansali SK, Desai AN, Dhaboowala CB. Tuberculous perforation of the small intestine. A clinical analysis of 19 cases. J Assoc Physicians India 1968; 16(6):351 – 355.
15. Toynton SC. Behçet's syndrome: an unusual cause of ileal perforation. J Clin Gastroenterol 1994; 19(1):84 – 85.
16. Seliger G, Pinto RS. Pneumoperitoneum secondary to acute perforation of a tuberculous ulcer of the small intestine. A report of two cases. Am J Gastroenterol 1974; 62(5):430 – 434.
17. Bhansali SK, Desai AN, Dhaboowala CB. Tuberculous perforation of the small intestine. A clinical analysis of 19 cases. J Assoc Physicians India 1968; 16(6):351 – 355.
18. Mandal BK, Schofield PF. Abdominal tuberculosis in Britain. Practitioner 1976; 216(1296):683 – 689.
19. Prakash A, Rao IR. Acute tuberculous perforation of the small intestine. Int Surg 1975; 60(8):397 – 398.