Multiple Intestinal Perforations in A 34 Years Old Male with Behçet Disease: A Case Report

Taher Y. Somili, Jamal S. Matar, Mohammed A. Albasiouny, Raed H. Rayani, Naif A. Hakami, Ohud E. Alakhrash, Fazal H. Shah

Abstract— 34 years old male patient patient, a known case of Behçet disease on high dose of daily cortisone for seven years, presented with generalized abdominal pain, blood-streak stool and multiple mouth and scrotal ulcers with pneumoperitonium confirmed by erect abdominal and chest x-ray. Diagnosed as a case of perforated intestine with peritonitis. His condition improved after exploratory laparotomy and resection of distal ileum with limited right hemicolectomy.

Index Terms— Behçet syndrome, Behçet disease, Inflammatory bowel disease, Gastrointestinal ulcers, Intestinal perforation, Exploratory laparotomy.

INTRODUCTION

Behçet disease (BD) is an idiopathic, chronic, relapsing, multi-systemic vasculitis, so it will be affecting multiple organ systems (1,2). We are presenting this case of young male patient who is a known case of Behçet disease for seven years on cortisone therapy with a rare complication of the disease as the first reported case in Jazan region-KSA to increase the awareness about the disease and its complications.

CASE REPORT

34 years old heavy smoker Saudi male, a known case of Behçet disease for seven years. He was doing well till seven years prior to admission when he started to notice multiple mouth ulcers, he neglected that, but after one month he developed scrotal ulcers. Then he went to private hospital, seen by doctors there, and after workup they diagnosed him as Behcet disease. The patient discharged on oral cortisone but he was not compliance on treatment. One year later, he developed sudden loss of vision in his left eye, went to ophthalmologist and diagnosed as retinal hemorrhage and referred to higher center for management, after that surgery was done and condition improved and discharged on daily cortisone (10mg Orally). The patient after that was on usual status of health for five years until two weeks prior to presentation when he started to have multiple mouth ulcers, blood-streak stool and on-off abdominal pain at lower abdominal area, he took analgesics and the pain relieved. One day prior admission at 9:00 p.m. he developed generalized abdominal pain, progressively increased during night till 4:00 a.m. when the pain was severe enough to make the patient restricted to his bed. The family took him to the nearest hospital, investigations were done including erect abdominal x-ray which confirmed the presence of air-under-diaphragm and the patient advised for operation but he refused and leave against medical advice. After that he came to our hospital ER at 1:00 p.m. with abdominal pain but less severe because he took analgesic before he came. At presentation, the patient was conscious, alert and oriented, Bp=90/60, temp=36.8oC, HR=110, RR=25 and sPo2=98%. By examination, there was generalized abdominal tenderness and guarding, multiple mouth ulcers [Fig.1] and scrotal ulcers, otherwise no abnormality detected in other systems. Patient underwent abdominal ultrasound, erect abdominal x-ray and other workup. The x-ray confirmed air-under-diaphragm [Fig.2]. The patient started on IVF, keep

NPO, and given analgesics and antibiotics then shifted to OR. In OR, exploratory laparotomy was done, we found approximately three liters of fluids with fecal material in peritoneal cavity, six perforations on anti-mesenteric border of the ileum with multiple hemorrhagic spots [Fig.3], and two lacerations, the proximal one was three feet from ileocecal valve while the distal one was six inches from ileocecal valve. After that resection of the four feet of distal ileum with limited right hemicolectomy done, end-to-end anastomosis by stapler established and two large bore drains was placed, and then abdomen closed by proline along with tension sutures. The postoperative course remained smooth, drains were removed in 3rd and 7th post-operative days respectively and hydrocortisone was tapered off and replaced by oral prednisolone. Patient discharged after ten days of hospital stay with stable condition. Past and family history are not significant. The patient is allergic to eggs, mango and milk products, and he is on daily cortisone.

DISCUSSION

Behçet disease (BD) is an inflammatory disorder characterized by recurrent multiple oral and genital ulcers, uveitis and skin lesions. Behçet disease patients can also present with gastrointestinal lesions, arthritis, CNS symptoms and vascular lesions (1,2,3). Gastrointestinal manifestations of Behçet disease are of particular importance as they are associated with significant mortality and morbidity, because of that reason, surgeons should be aware about complications of Behçet disease with intestinal ulcers, which tend to perforate at multiple sites (3,4). GI manifestations usually occur up to six years after the onset of oral ulcers (5). The most common symptoms of Behçet disease are abdominal pain, nausea, vomiting, diarrhea and GI

bleeding (6). Although ileocecal involvement is most commonly described, Behçet disease can involve any alimentary tract segment and various GI organs (5,7). Behçet disease can present with clinical manifestations that may mimic other diseases like Crohn's disease. The diagnosis of Bechet disease still a challenge in the medical field because there are no pathognomonic laboratory tests for it. The most widely acceptable is the International Study Group (ISG) published criteria for Behçet Disease in 1990 (4). The suspicion of intestinal perforation in our case has arisen due to high dose of cortisone along with signs and symptoms during presentation. Patient treated from Behçet disease-related intestinal perforation with laparotomy and resection of the affected segments. Then he discharged home after he became stable, on oral prednisolone after adjustment of the dose.

CONCLUSION

Gastrointestinal manifestations of Behçet disease are common and very important because of their association with significant morbidity and mortality. Ileocecal involvement is most commonly involved, because of that reason surgeons should to be aware of complication of Behçet disease. Many surgeons consider Behçet disease as a subtype of inflammatory bowel disease. Proper diagnosis and management of the disease along with regular follow-up will decrease the future serious complications and lead to good prognosis of such patients.

FIGURES

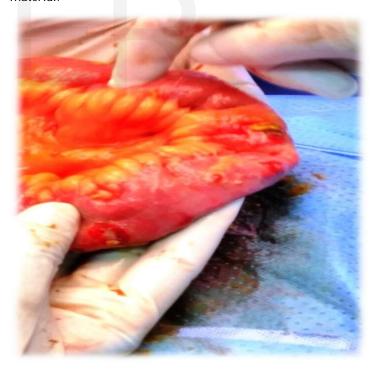
Figure 1: Photo shows a large ulcer in soft palate.



Figure 2: Erect abdominal x-ray shows air-under-diaphragm, which indicates intestinal perforation.



Figure 3: Photo shows multiple hemorrhagic spots with fecal material.



REFERENCES

- Sakane T, Takeno M, Suzuki N, Inaba G. Behçet's disease. N Engl J Med. 1999;341:1284–1291. [PubMed]
- (2) Saleh Z, Arayssi T. Update on the therapy of Behçet disease. Ther Adv Chronic Dis. 2014;5:112–134. [PMC free article] [PubMed]
- (3) Hamdan A, Mansour W, Uthman I, Masri AF, Nasr F, Arayssi T. Behçet's disease in Lebanon: clinical profile, severity and twodecade comparison. Clin Rheumatol. 2006;25:364– 367. [PubMed]
- (4) Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. Lancet. 1990;335:1078– 1080. [PubMed]
- (5) Bayraktar Y, Ozaslan E, Van Thiel DH. Gastrointestinal manifestations of Behçet 's disease. J Clin Gastroenterol. 2000;30:144– 154. [PubMed]
- (6) Grigg EL, Kane S, Katz S. Mimicry and deception in inflammatory bowel disease and intestinal behçet disease. Gastroenterol Hepatol (N Y) 2012;8:103–112. [PMC free article][PubMed]
- (7) Ebert EC. Gastrointestinal manifestations of Behçet's disease. Dig Dis Sci. 2009;54:201–207. [PubMed]

