Journal of Rheumatic Diseases
Vol. 21, No. 6, December, 2014
http://dx.doi.org/10.4078/jrd.2014.21.6.311

□ Case Report □

A Case of Behçet's Disease with Both Esophageal and Pharyngeal Ulcers Successfully treated with Steroid and Sulfasalazine

Ha Yong Yoon, In Jin Cho, Hyun Lee, Yun Jung Choi, Won Seok Lee, Wan-Hee Yoo

Division of Rheumatology, Department of Internal Medicine, Chonbuk National University Medical School and Research Institute of Clinical Medicine of Chonbuk National University Hospital-Chonbuk National University, Jeonju, Korea

Behçet's disease is a heterogeneous disease that involves variable organ systems. Gastrointestinal Behçet's disease is rare and it generally affects the terminal ileum with propagation toward the cecum and ascending colon. Therefore, esophageal ulcer associated with Behçet's disease has not been reported frequently. We report an unusual case of Behçet's disease associated with both esophageal and pharyngeal ulcers. A 64-year-old man was admitted for an evaluation of pharyngeal and substernal discomfort sustained for 3 months. He had no underlying chronic disease; however, he suffered from recurrent oral and genital ulcers for 20 years and had folliculitis-like skin lesions on the face, scalp and trunk. He was diagnosed with Behçet's disease and gastroesophageal fiberscopy revealed deep ulcers on both the pharynx and upper esophagus. Esophagopharyngeal ulcers were successfully treated with prednisolone, colchicines, dapsone, and sulfasalazine.

Key Words. **Esophageal ulcer, Behçet's disease, Steroid, Sulfasalazine**

Introduction

Behçet's disease (BD) is a heterogeneous disorder with variable involvement of many organ systems, mainly including mucocutaneous, articular, vascular, ophthalmological and gastrointestinal systems. Gastrointestinal BD has been reported to be rare and mainly affects the terminal ileum, cecum, and ascending colon. Esophageal involvement of BD is much rarer and mainly reported in Japan (1). Among cases of gastrointestinal BD reported in Korea, only 8.7% showed esophageal ulcers (2).

Gastrointestinal BD has been reported to be difficult to treat and resistant to conventional medication against BD, including 5-aminosalicylic acid (5-ASA), systemic corticosteroids, and immunosuppressive agents (3). Serious complications such as strictures, bleeding, perforation, or fistulas have been reported especially in esophageal ulcers (2). In some reports, infliximab, anti-tumor necrosis factor alpha monoclonal antibody, wsa recommended for the treatment of resistant intestinal BD (3,4).

We here present an unusual case of BD associated with both esophageal and pharyngeal ulcer treated successfully with prednisolone, colchicines, dapsone, and sulfasalazine.

Case Report

A 64-year-old man was admitted for the evaluation of pharyngeal and substernal discomfort sustained for 3 months. He had no underlying chronic disease including diabetes, hypertension, hepatitis, and tuberculosis. He was non-smoker and has no history of alcohol and drug abuse. He has not been exposed to any irradiation therapy or operation. He has no familial history related to specific diseases including connective tissue disorder, vasculitis, and ulcerative disease of gastrointestinal systems. However, he has suffered from recurrent oral and genital ulcers for 20 years. At admission,

＜Received : June 15, 2013, Revised (1st : November 23, 2013, 2nd : December 12, 2013), Accepted : January 3, 2014＞
Corresponding to : Wan-Hee Yoo, Division of Rheumatology, Department of Internal Medicine, Chonbuk National University Medical School and Research Institute of Clinical Medicine of Chonbuk National University Hospital-Chonbuk National University, San 2-20 Geumam-dong, Deokjin-gu, Jeonju 561-180, Korea. E-mail : ywhim@jbnu.ac.kr

pISSN: 2093-940X, eISSN: 2233-4718

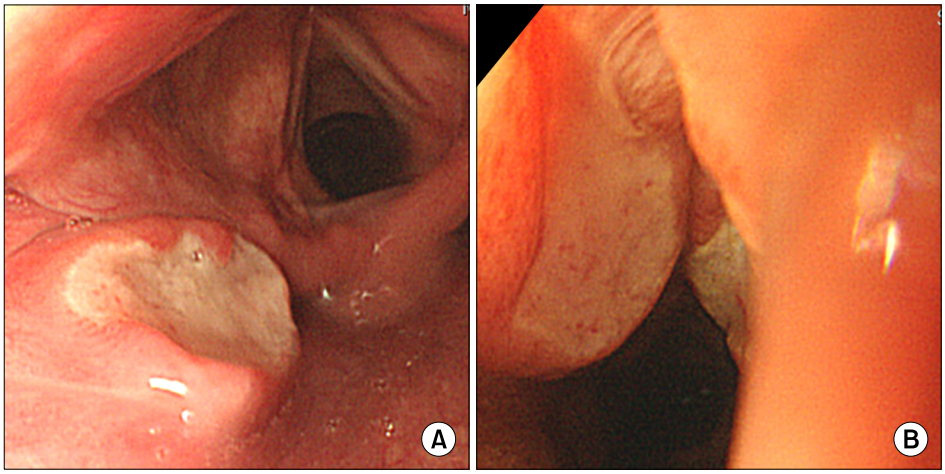

Figure 1. Pharyngeal (A) and esophageal (B) ulcers in Behcet's disease.

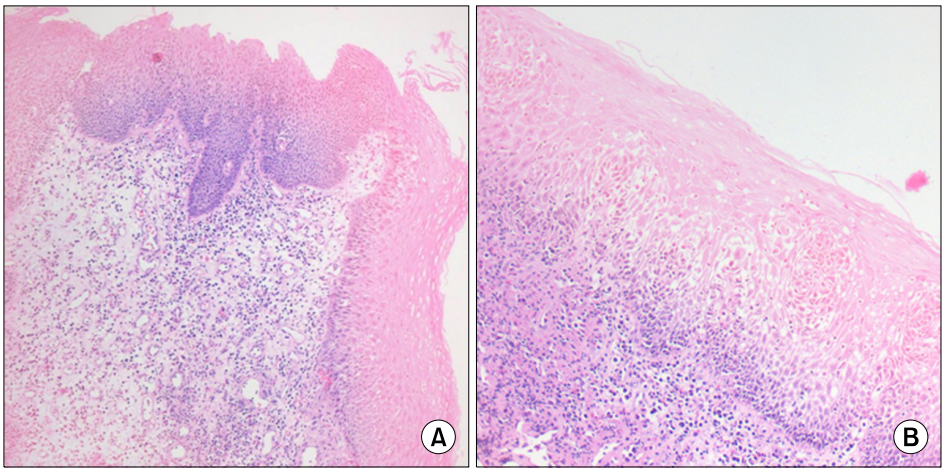

Figure 2. Histopathologic findings of ulcer lesion (A) ×200, (B) ×100.

blood pressure of 110/76 mmHg, body temperature of 36.9°C, pulse rate of 72 beats/min and respiratory rate of 20 breaths/min were checked. There were multiple oral ulcers on tongue and lip with multiple folliculitis-like skin lesions on face, scalp and trunk. Laboratory test revealed white blood cell (WBC) of 6.1×10^3/mL, Hb of 13.5 g/dL and platelet of 2.7×10^5/mL. Erythrocyte sedimentation rate (ESR), CRP, and blood chemistries were within normal limit. HLA-B51, autoantibodies and tests for viral infections including CMV, EBV, HSV, and HIV were all negative. Gastroesophageal fiberscopy revealed multiple, deep ulcers with distinct border and irregular base on pharynx and upper esophagus (Figure 1A and 1B) without specific lesion on stomach except atrophic gastritis. Histopathologic finding of ulcerative lesions showed nonspecific inflammation, excluding other possible etiologies except BD (Figure 2).

There were no specific abnormal findings on entire colon at colonoscopy (not shown here). A diagnosis of BD was made with characteristic clinical symptoms, including recurrent oral, genital ulcers, folliculitis and positive Pathergy test. He had no history of recent infections (virus, bacteria and Candida albicans), radiation, ingestion of corrosive substances, certain pills, and Crohn's disease. Thus he has been treated with prednisolone (0.5 mg/kg, 30 mg), colchicines, dapsone, and sulfasalazine.

Discussion

We here present an unusual case of BD associated with esophageal and pharyngeal ulcers which were successfully treated with prednisolone, colchicines, dapsone, and sulfasalazine.

BD is a heterogeneous disorder with variable involvement of many organ systems, mainly including, mucocutaneous, articular, vascular, ophthalmological and gastrointestinal systems. Gastrointestinal BD has been reported to be rare. The terminal ileum, cecum, and ascending colon are mainly affected by gastrointestinal BD (5) and esophageal involvement covers below 10% of gastrointestinal BD (2). To our knowledge, less than 40 cases have been reported to be associated with esophageal ulcers in English literatures (6-12). Most of the cases

were reported in Japan.

Gastrointestinal BD is difficult to treat and serious complications such as strictures, bleeding, perforation, or fistulas have been reported especially in esophageal ulcers (2). Typical symptoms suggesting gastrointestinal BD includes abdominal pain, diarrhea, and melena. Therefore, early detection of esophageal ulcer through gastroesophageal fiberscopy might be delayed in BD patients with simply poor oral intake or substernal chest discomfort.

In this case, ulcers were found in both pharynx and upper esophagus (Figure 1), whereas the esophageal ulcers of BD patients locate mainly in the middle or lower esophagus (11). Gastrointestinal BD has been reported to be difficult to treat and resistant to conventional medication against BD, including 5-aminosalicylic acid (5-ASA), systemic corticosteroids, and immunosuppressive agents (3). We decided to treat esophagopharyngeal ulcers with empirical medication including prednisolone, colchicin, dapsone and sulfasalazine. Symptom was dramatically improved within a few weeks following the start of medication.

To prevent severe complications including perforation or fistula, it is critical to diagnose gastrointestinal BD earlier and treat properly. It can be successfully treated with empirical trial of both salazosulfapyridine and steroids.

Summary

Our case suggests following clinical implications. In patients not diagnosed with BD, BD should be considered as the differential diagnosis for newly detected esophageal ulcers, especially when it is recurrent despite of conventional treatment for ulcers. In case of BD patient, early gastroesophageal fiberscopy should be recommended even for atypical upper gastrointestinal symptoms or signs.

References

1. Yasuo M, Miyabayashi H, Okano T, Aoki H, Ichikawa K, Hirose Y. Successful treatment with corticosteroid in a case of Behçet's syndrome with multiple esophageal ulcerations. Intern Med 2003;42:696-9.
2. Yi SW, Cheon JH, Kim JH, Lee SK, Kim TI, Lee YC, et al. The prevalence and clinical characteristics of esophageal involvement in patients with Behçet's disease: a single center experience in Korea. J Korean Med Sci 2009;24:52-6.
3. Maruyama Y, Hisamatsu T, Matsuoka K, Naganuma M, Inoue N, Ogata H, et al. A case of intestinal Behçet's disease treated with infliximab monotherapy who successfully maintained clinical remission and complete mucosal healing for six years. Intern Med 2012;51:2125-9.
4. Almoznino G, Ben-Chetrit E. Infliximab for the treatment of resistant oral ulcers in Behçet's disease: a case report and review of the literature. Clin Exp Rheumatol 2007;25(4 Suppl 45):S99-102.
5. Sakane T, Takeno M, Suzuki N, Inaba G. Behçet's disease. N Engl J Med 1999;341:1284-91.
6. Lehmann H, Stutte HJ, Zierott G, Schlaak M. A case of Behcet's disease with multiple intestinal ulcerations (author's transl). Dtsch Med Wochenschr 1975;100:308-11.
7. Arma S, Habibulla KS, Price JJ, Collis JL. Dysphagia in Behçet's syndrome. Thorax 1971;26:155-8.
8. Mori S, Yoshihira A, Kawamura H, Takeuchi A, Hashimoto T, Inaba G. Esophageal involvement in Behcet's disease. Am J Gastroenterol 1983;78:548-53.
9. Yashiro K, Nagasako K, Hasegawa K, Maruyama M, Suzuki S, Obata H. Esophageal lesions in intestinal Behçet's disease. Endoscopy 1986;18:57-60.
10. Glosemeyer R, Deppe H, Dormann AJ, Wigginghaus B, Stolte M, Stadler R, et al. Ulcerative esophagitis and colitis as rare manifestations of Adamantiades-Behcet disease. Z Gastroenterol 2001;39:167-71.
11. Lorenzetti ME, Forbes IJ, Roberts-Thomson IC. Oesophageal and ileal ulceration in Behçet's disease. J Gastroenterol Hepatol 1990;5:714-7.
12. Ikezawa K, Kashimura H, Hassan M, Nakahara A, Yanaka A, Matsuzaki Y, et al. A case of Behçet's syndrome with esophageal involvement treated with salicylazosulfapyridine and prednisolone. Endoscopy 1998;30:S52-3.