

Gastrointestinal Involvement in Behçet Disease



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KEYWORDS

- Behçet syndrome • Mouth diseases • Intestinal diseases • Gastrointestinal diseases
- Epidemiology • Colonoscopy • Antirheumatic agents • TNF inhibitors

KEY POINTS

- There is a wide variation across countries in the reported frequency of gastrointestinal involvement among Behçet disease patients. Frequencies of less than or equal to 50% were reported from the Far East, whereas it is approximately 1% in Turkey.
- The scope of gastrointestinal involvement in Behçet disease includes ulcers in the ileocolonic region, which is the most commonly involved area; ulcers in other parts of the intestines; and Budd-Chiari syndrome due to hepatic vein and/or inferior vena cava thrombosis.
- The most common symptoms are abdominal pain of variable intensity, usually in the right lower quadrant; diarrhea, with or without bleeding; and fever.
- Presence of typical ulcers on colonoscopy is essential for the diagnosis. Histopathologic examination of surgical samples show neutrophilic infiltration of the vessel wall, perivascular area, and intravascular area with more frequent involvement of venules compared with arteries.
- Management consists of 5-aminosalicylate derivatives in mild cases, azathioprine in moderate to severe cases, and glucocorticoids during acute exacerbations.

INTRODUCTION

Behçet disease (BD) was first described by a Turkish dermatologist as a triple-symptom complex that consists of oral aphthous ulcers, genital ulcers, and uveitis. It is now considered a unique vasculitic condition that causes inflammation of vessels of all size with involvement of several organs and organ systems.^{1,2} Nodular lesions,

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papulopustular lesions, arterial and venous involvement, central nervous system, and gastrointestinal (GI) involvement are other manifestations. GI involvement causes both macroscopic and histopathologic inflammatory changes that resemble inflammatory bowel diseases (IBDs), creating a diagnostic challenge, as well as vasculitis-driven relapsing ischemic features that result in higher perforation and major GI bleeding rates compared with IBD.³

EPIDEMIOLOGY

Epidemiology of Behçet Disease

The prevalence of BD shows a wide geographic variation.⁴ It is known to be higher in countries along the ancient Silk Route starting from the Mediterranean and reaching the Far East.⁴ The highest prevalence was reported from Turkey, with equal to or less than 421 cases per 100,000 (95% CI 340–510) in a population-based survey.⁵ In the Far East, the prevalence has been reported as 14 per 100,000 in China and 13.5 per 100,000 in Japan.^{6,7} Differences between ethnic populations within the same country have also been reported. A study from Israel reported that the overall BD prevalence was 15.2 per 100,000 but 146.4 among Druze, 26.2 among Arabs, and 8.6 among the Jews living in Israel.⁸ The prevalence in Europe decreases from South to North and was reported as 15.9 per 100,000 in Southern Italy, 6.4 per 100,000 in Spain, 0.64 per 100,000 in the United Kingdom and 0.3 per 100,000 in Scotland; rates thus decrease as distance from the Silk Route increases.^{9–12} BD prevalence among immigrants from a high-prevalence country who move to a low-prevalence country has been estimated to belong between the 2 extremes, suggesting a role for both genetics and environment in the pathogenesis.¹³ Moreover, BD seems to run a less severe course in nonendemic countries, providing another clue for the role of environmental factors.¹⁴ Finally, a recent metaanalysis of 45 prevalence studies showed that, although there was clearly a wide variation across countries, methodologic issues (ie, registry-based studies vs direct population sampling) may be responsible for some of the variation.¹⁵

Epidemiology of Gastrointestinal Involvement of Behçet Disease

The frequency of GI involvement among patients with BD also shows a wide variation across geographies, being much more common in the Far East compared with the Middle East and Europe⁴ (**Table 1**). The frequency was as high as 50% in a Japanese cohort¹⁶ and 1% in a formal study in Turkey.¹⁷

Caution is required when interpreting the frequencies reported in different studies. Some sources of bias in reporting the frequency of GI involvement may be:

- Mode of diagnosis, such as symptom-based compared with endoscopy. Earlier symptom-based studies report a higher frequency compared with endoscopy or imaging-based reports in Japan, where BD is endemic, and other inflammatory bowel conditions, such as Crohn disease (CD), are rare. Moreover, a recent Korean study showed that only half of the subjects with upper GI symptoms did actually have esophageal involvement confirmed by endoscopy.¹⁸
- Use of highly sensitive techniques such as capsule enteroscopy or double-balloon enteroscopy. Such methods can pick up incidental nonspecific lesions even in healthy individuals and may cause an overestimation for the frequency of GI involvement.¹⁹
- Differences in criteria used for diagnosis. GI involvement is 1 of the items in Japanese criteria, whereas it does not appear in the International Study Group (ISG) criteria.^{20,21} Consequently, use of Japanese criteria may result in a higher proportion of patients with GI involvement in the total pool.

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