Ileocolonic Findings in Behçet's Disease

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Dear Sir,

I read with great interest the paper written by Köklü et al. [1] describing the frequency of gastrointestinal involvement at endoscopy in Turkish patients with Behçet's disease (BD). Of 50 asymptomatic patients with BD, 9 (18%) had ileum ulcers and the remaining 41 (82%) had a normal ileum at colonoscopy. Moreover, histologic examination of BD patients with a normal colon and terminal ileum at colonoscopy revealed 61% abnormal ileum histologies. So, they concluded that most of the patients with BD have macroscopic or microscopic ileocolonic involvement. Although they contributed an original investigation regarding ileal histologies of BD, many questions arise from their findings.

First, their description of intestinal BD needs further clarification. The objective diagnosis of intestinal BD is generally very difficult due to its rareness, the asynchronous nature of multi-systemic findings and the absence of specific macroscopic or microscopic features. Although histological vasculitis leads to a definite diagnosis of intestinal BD, it is rarely observed [2]. Biopsies often show a nonspecific inflammatory infiltrate, and therefore it plays a role mainly in excluding other intestinal diseases rather than in confirming the diagnosis of intestinal BD. There is no worldwide consensus for the diagnosis and management of intestinal BD, but we are thankful to efforts of a Korean group which proposed an algorithm based on the type of ileocolonic ulcerations and clinical manifestations [3]. Patients with <5 ulcers that were oval in shape, deep, with discrete

borders, and located in the ileocecal area were classified as having 'typical ulcers'. Ulcerations that did not fulfill all of the characteristics, such as several tiny, shallow, aphthous ulcers were regarded as atypical. The clinical characteristics were categorized as definite, probable, suspected and non-diagnostic. These criteria were validated in a group of 280 patients with ileocolonic ulcers of whom 145 were diagnosed as having intestinal BD as a final diagnosis. The overall sensitivity, specificity, positive predictive value, negative predictive value, and accuracy of the diagnostic probability of these criteria were 98.6, 83.0, 86.1, 98.2, and 91.1%, respectively. Köklü et al. [1] did not describe any features of macroscopic ileocolonic lesions apart from defining them as 'ulcers'. What about the variables, such as ulcer shape, margin and distribution of lesions? Moreover, biopsies of those macroscopic ulcers (a total of 9 cases) showed vasculitis in only 3 cases, while 5 cases had ileitis, 1 amebiasis, and 1 nonspecific ileitis. In the present series the question regarding histology should also apply for microscopic involvement of BD cases with normal endoscopy. Only 2 cases with vasculitis were found, while the remaining patients had chronic ileitis (8 cases), nonspecific ileitis (12 cases), and villous atrophy (1 case). Although the microscopic findings of BD patients were higher compared to the control group, the reported features are largely nonspecific. I wonder about the details of the macroscopic lesions and would like to have further comments regarding the microscopic features.

Second, the intestinal lesions of BD are typically resistant to medical treatment and frequently recur even with surgical treatment [4–6]. The present study did not report any symptom or complication regarding intestinal BD during follow-up (1–25 months). Which factors may explain such an unusual clinical course? If available, what were the therapy choices and results of their cases?

If we know more details about this large study, we can obtain more valuable information which will be useful in clinical practice.

References

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Reply

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Dear Sir,

Thank you for your contributions and comments regarding our study currently published in *Digestion* [1]. Further clarification regarding the description of intestinal Behçet's disease (BD) was suggested. As stated elsewhere, an accurate diagnosis of intestinal BD has great challenges. First, there are no pathognomonic, endoscopic, radiologic or histological findings for intestinal BD. Endoscopic lesions may mimic lesions of many other diseases including Crohn's disease, tuberculosis or NSAID-related lesions. Diagnosis of intestinal BD mainly depends on the exclusion of some other diseases which are not frequent in patients with BD. Moreover, not all patients with BD fulfill all the criteria of BD at the time of endoscopic evaluation. Hence, there may be an interval from several months to years between the onset of BD and intestinal involvement.

Recently, Cheon et al. [2] reported diagnostic criteria for intestinal BD. They seem comprehensive; however, as stated by the authors, further studies are required to evaluate their validity in populations other than Korean patients. The frequency of typical and atypical ulcers was similar in our series. The table in our article may help to adapt the criteria of the BD Research Committee of Japan [3].

Ozarslan regarded the histological findings of our patients as nonspecific and requested further clarification. Histological evidence of vasculitis is highly suggestive of intestinal BD; however, it is not commonly observed in those patients [4]. Also, histological examinations usually show a nonspecific inflammation near the ulcers surrounded by normal intestinal tissue. Hence, the role of histological evaluation is mainly exclusion of other intestinal diseases rather than confirming intestinal BD diagnosis [2]. Therefore, heterogeneous reports on pathological evaluation should not be found as surprising.

Finally, our study was not a follow-up study. The range (1–25 months) noted by Ozarslan was not the follow-up period, instead it defined the time to diagnosis of BD prior to endoscopic examination. Budesonide, 5-ASA and azathioprine separately or in combination were preferred in those patients with intestinal BD, according to other systemic involvements. Response to medical treatment is not as satisfactory regarding intestinal BD. However, that was beyond the aim of the present study and may be reported in the future.

References

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