Letter to the editor: "RE: Firdaus et al. Case Report: Intestinal Obstruction in Behçet's Disease: A Rare Presentation."

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Behçet's disease (BD) is a multi-systemic auto-immune condition characterised by ulcerations in the oral cavity, ocular lesions, skin manifestations and complications in the gastrointestinal (GI) tract [1, 2]. Patients with BD have an abnormally heightened systemic inflammatory response resulting in multi-organ clinical pathologies, including mucocutaneous and ophthalmological manifestations[3]. Although oral ulceration and ocular lesions are the most common symptoms in Behçet's disease patients, GI manifestations have also been reported.

In a recent case report by Firdaus et al.[5], the authors reported a rare case of intestinal obstruction in a BD patient. Contrary to the authors' remark, the occurrence of intestinal manifestation in BD is not rare. The occurrence of intestinal BD occurs in 3-60% of patients with BD [1-4], compared to 3-26% as commented by the author [5]. The frequency stated in this report is outdated and this is evidenced by the use of an inappropriate and old reference (a Behçet's Syndrome Society Website Factsheet from 2008). Moreover, the frequency of intestinal manifestation in BD is higher in East Asian countries than in Western or Middle Eastern countries[6]. Classically, intestinal BD manifests as ulcers in the ileocecal region with symptoms varying from abdominal pain, diarrhoea or constipation due to perforation of ulcers[3]. Intestinal BD and inflammatory bowel disease (IBD), such as Crohn's disease (CD), share a considerable number of clinical features. The case reported presented by Firdaus et al.[5] showed multiple mucosal ulcerations in a localised area in the lower intestinal region of the patient's abdomen. Furthermore, the intestinal ulcers found in this patient were described as "deep multiple serosal punctum" characteristic of round and oval shaped ulcers. The focal distribution and punctate shape of the ulcers is consistent with the colonic description in the intestines of patients with BD, not CD [7, 8]. Moreover, histological examination of the resected specimen revealed vasculitis [5]. Although vasculitis is a commonly accepted feature in inflammatory bowel diseases [9], to date there are no indicative histologic findings regarding intestinal BD [10].

The authors in the case report acknowledged the challenges in discerning between intestinal BD and IBD[5]. Although clinical and endoscopic manifestations resemble each other, independent characteristics can be found through careful clinical, biochemical and genetic evaluation. Human leukocyte antigen (HLA)-B51 allele and MHC class I related gene A (MICA) have been shown to be two susceptible loci for BD [11, 12]. This could be a potential differentiator to distinguish between BD and IBD as HLA-B51 or MICA has not been shown to be associated with IBD susceptibility. The possibility of the HLA-B51 or MICA loci to independently contribute to BD warrants further investigation. Although the evidence is preliminary, specific immunological events (e.g. the presence of serum anti-herpes simplex virus-1 antibodies and anti-endothelial cell antibody in the patients with BD) and changes in the oral microbiota flora (e.g. antibodies against Streptococcus sanguis detected in the oral mucosa and sera of patients with BD) can potentially assist clinicians to distinguish between BD and IBD in the future (See recent review by Kim and Cheon,[10]. Future efforts are needed to distinguish intestinal BD from other GI inflammatory conditions, such as CD.

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