A 67-year-old Japanese man presented with syncope. He described recurrent fever for 1 year accompanied by oral ulcers, genital/scrotal ulcers, arthritis, abdominal pain and hematochezia. Physical examination revealed oral and inguinal ulcers (Figure 1A). Laboratory tests revealed elevated white blood cell (WBC) counts (15.5 x 10^9/l), macrocytic anemia (hemoglobin: 117 g/l) and fecal occult blood positivity. Although whole-body computed tomography, esophagogastroduodenoscopy and colonoscopy findings were normal, capsule endoscopy revealed multiple round or oval ulcers in the jejunum, ileum and terminal ileum (Figure 1B). Conventional chromosomal analysis on peripheral blood cultures confirmed the presence of trisomy 8 (see Supplementary Material), and the diagnosis of gastrointestinal Behcet’s disease (GIBD) with trisomy 8 was made. Colchicine and mesalamine treatment relieved the symptoms.

GIBD is a BD subtype characterized by predominant recurrent gastrointestinal manifestations and objectively identified intestinal ulcerations.1 This case satisfied the diagnostic criteria for GIBD proposed by Japan and Korea,2,3 including oral and inguinal ulcers, multiple intestinal round or oval ulcers and arthritis. To distinguish GIBD from Crohn’s disease, which is difficult, gastrointestinal endoscopy findings are essential. The round ulcerations of GIBD differ from the longitudinal, cobblestone-like ulcerations of Crohn’s disease.4 Although most cases of BD are idiopathic, the association of BD with myelodysplastic syndrome (MDS) accompanied by trisomy 8 has been reported.5 The typical presentation comprises late onset, periodic fever and gastrointestinal involvement,5,6 while the usual age of onset of GIBD is 30–40 years.1,2 This patient was not diagnosed with MDS owing to his sufficient WBC and platelet counts. However, the presence of macrocytic anemia could indicate the necessity of careful follow-up of his complete blood count.

GIBD accompanied by MDS with trisomy 8 should be considered if suggestive clues such as late onset, periodic fever and macrocytic anemia are present, as symptoms of severe treatment-resistant BD with trisomy 8 can be ameliorated by treatment for MDS.7

Supplementary material
Supplementary material is available at QJM online.

Funding
This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

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Conflict of interest: None declared.
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