Symptom clusters in Behçet's disease: A case series

Young Joon Park[1], Heera Lee[2], Eun-So Lee[1]

[1]Department of Dermatology, Ajou University School of Medicine, Suwon, Korea

[2]Department of Dermatology, Ewha Womans University College of Medicine, Seoul, Korea

INTRODUCTION

- Cutaneous manifestations are not only the most common findings, but are crucial to understanding Behçet's disease (BD)
- However, clinicians who encounter BD initially, should also be aware of.the extracutaneous manifestations, as these may help predict disease severity of BD.
- Unfortunately, no validated biomarkers currently exist to reliably assess the disease severity in BD.

Case 1

- A 44-year-old man was diagnosed with BD (incomplete type according to Japanese criteria; oral ulcers, genital ulcers, and skin lesions) in 1998 at the age of 19. **Intestinal involvement** was first confirmed through colonoscopy in 2005, and subsequently he was administered Humira (adalimumab). However, he underwent right colectomy including ileal resection in 2020, as the disease was uncontrollable. Later, in 2023, the patient complained of headaches accompanied by slurred speech, fecal and urinary incontinence, and even suffered seizure. Brain magnetic resonance imaging (MRI) revealed 5.5 x 4 cm-sized **brain abscess** in the Lt. frontal lobe, which was drained by catheter insertion. The abscess was sterile.

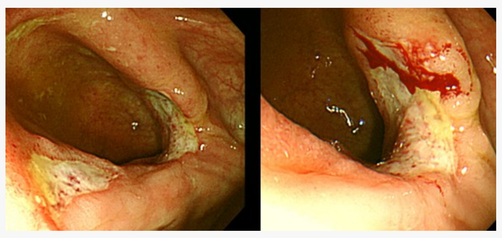

Fig 1.
ulcerative lesion covered with exudate on IC valve
(colonoscopy in 2012)

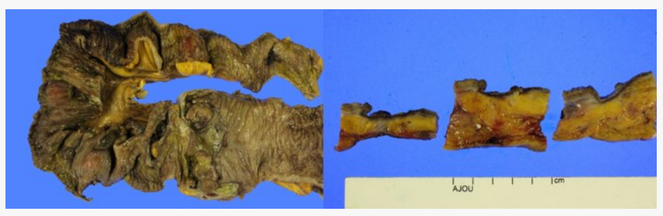

Fig 2.
colon and small intestine, after Rt. hemicolectomy with ileal resection (2020)

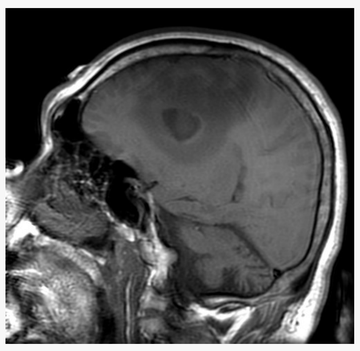

Fig 3.
5.5 x 4cm sized peripheral enhancing lesion at Lt. frontal lobe showing internal diffusion restriction content, with peripheral low SI margin causing midline shifting and unchal herniation
(brain MRI in 2023)

Case 2

- A 40-year-old man was diagnosed with BD (incomplete type according to Japanese criteria; oral ulcers, genital ulcers, and skin lesions) in 2009 at the age of 26. He underwent right hemicolectomy in 2015 due to the aggravation since **intestinal involvement** confirmation in 2009. Then in 2016, **aortic aneurysm** had been found, and graft interposition was performed. Soon after, he started psychiatric consultation because of the adjustment disorder with neurotic depression.

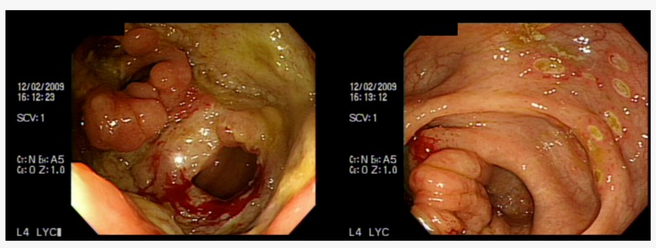

Fig 1.
deep large ulceration on terminal ileum
(colonoscopy in 2009)

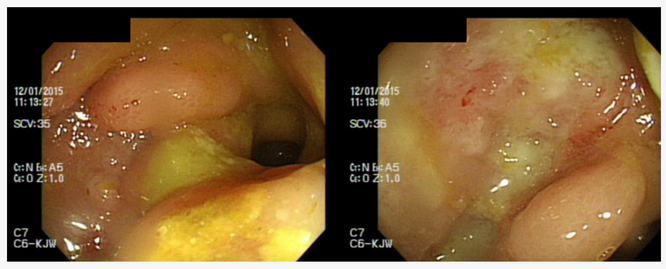

Fig 2.
retractable deep large active ulceration on terminal ileum
(colonoscopy in 2015)

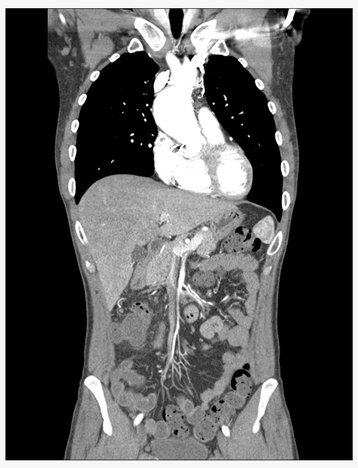

Fig 3.
aneurysmal dilatation of ascending aorta measuring 5.2 cm in dimension (CT in 2016)

DISCUSSION

- Previously, symptoms of BD were considered independent. However, there have been some reports that demographic and genetic features may be associated to different clinical characteristics. Recently, multiple studies have been conducted grouping the symptoms of BD and approach them as a cluster. Mizuki group (2021) proposed five independent clusters of mucocutaneous, mucocutaneous with arthritis, neuro, gastrointestinal, and eye. Guan group (2021) suggested five subgroups with skin mucosa, joint, gastrointestinal, uveitis, and cardiovascular and CNS clusters representing commonly involved organs.

CONCLUSION

- As our cases show, BD patients might show a cluster of symptoms, and symptom clustering may lead to insights into the BD pathogenesis.