Epstein-Barr Virus Associated Ulcers in a Patient with Ulcerative Colitis Taking Vedolizumab: A Case Report

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Introduction

Wiskott-Aldrich Syndrome (WAS) is an X-linked recessive disorder characterized by immunodeficiency, eczema and microthrombocytopenia. The syndrome is caused by compromised function of the WASp protein resulting in a decreased ability of cells to polymerize and reorganize actin, which is crucial for various cellular processes such as cellular division, immune synapse formation, phagocytosis, antigen presentation, and immune surveillance and regulation.

The immune dysfunction in WAS can lead to the development of other autoimmune diseases and non-Hodgkin lymphoma. Additionally, individuals with WAS have an elevated risk of developing chronic inflammatory bowel disease. Here, we discuss a patient with WAS and ulcerative colitis (UC) receiving vedolizumab (VDZ) who developed an Epstein Barr virus (EBV) associated mucocutaneous rectal ulcer.

High Yield Information

- 32 y/o Male with recurrent episodes of bright red rectal bleeding
- PMH: WAS, ITP
- PSH(x): Splenectomy
- Meds: Prednisone tapers for flares + antibiotic prophylaxis.

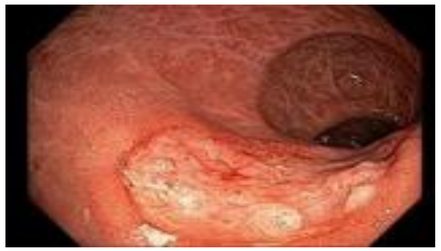
Case

A 32-year-old male with a medical history of congenital WAS, with resulting splenectomy, and recurrent bacterial infections presented with bloody diarrhea and lower abdominal pain. After infectious causes were excluded, a colonoscopy revealed findings and pathology consistent with ulcerative colitis. Despite initial treatment with prednisone and sulfasalazine, he remained symptomatic. The patient was then started on VDZ, which resulted in a resolution of his symptoms and follow up colonoscopy showed normal mucosal and no further inflammation on pathology except for the presence of a new 15-cm rectal ulcer that tested positive for EBV; there was no dysplasia, granulomas, cytomegalovirus (CMV), human herpesvirus 8 (HHV-8), or lymphoma. The patient continued with VDZ treatment, and 6-month follow-up colonoscopy revealed a stable mucocutaneous ulcer (Figure 1). Rectal steroids were initiated and VDZ was discontinued, resulting in symptom resolution. The patient's condition has remained asymptomatic, and further evaluation is pending with a repeat colonoscopy.

Patient Timeline

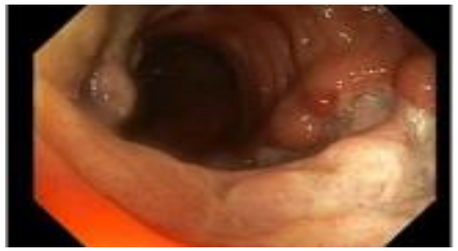
- 07/2019: Presents to ER with bright red bloody stools; Hg 7.7.
- Colonoscopy: Significant circumferential inflammation involving distal third colon. Differential= UC vs. CMV Colitis.
- Infectious w/u: Negative for CMV, C. diff.
- Symptomatic for 8 months. Opted for Vedolizumab (03/2020).
- 04/2021: Repeat C/S shows 15cm ulcer in Cecum. B(x) shows +EBV mucocutaneous ulcer. No dysplasia. Inflammatory & hyperplastic polyps.
- 01/2022: C/S & b(x) shows an unchanged +EBV mucocutaneous ulcer.
- 12/2022: C/S & b(x) show moderately larger and progressive ulcer
- 07/2023: Pathology consult concludes findings consistent with +EBV mucocutaneous ulcer/EBV positive/CD30 positive lymphoproliferative disorder
- 08/2023: Ulcer is larger and more diffuse with no new b(x) findings.

Figure 1: Colonoscopy Identification of Ulcers in the Rectum



Colonoscopy on 4/13/21

Colonoscopy on 12/13/22



Colonoscopy on 8/21/23

Discussion/Conclusion

The presence of EBV-associated mucosal ulcers in immunosuppressed patient on VDZ raises several important questions regarding VDZ use in patients with WAS and UC. EBV-related mucosal ulcerations (EBVMCU) are commonly misdiagnosed under the lymphoma family. However, they are distinctly different and importantly share different prognoses. The precise role of VDZ in developing EBV mucosal ulcers remains uncertain and future evaluation is warranted to understand the relationship between WAS, UC, and treatment with VDZ to guide and optimize management decisions.

References

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