Diagnosing Visceral Kaposi Sarcoma: The necessity to avoid anchoring on the diagnosis of a Crohn's flare

Emily S. Seltzer[1], Shabari Mangalore Shenoy[1], Bo Hyung Yoon[1], Frederick Rozenshteyn[1], Kevin Mijares[1], Kimberly Cavaliere[1]
[1]Mount Sinai Morningside-West Hospitals, Icahn School of Medicine at Mount Sinai, New York, NY, United States

LEARNING OBJECTIVES:

1. Learn to broaden the diagnostic arsenal where the picture of inflammatory bowel disease (IBD) is not clear cut
2. Understand when it is not appropriate to start steroids for a possible Crohn's flare

INTRODUCTION:

45-year-old male with reported history of Crohn's disease (off therapy) presenting with six months of worsening weakness, abdominal pain, bloody diarrhea (>10 episodes most days), dysphagia to solids, and a 27 lbs. weight loss.

Evaluation:

- Multiple violaceous plaques on his left leg
- Left lower quadrant abdominal tenderness
- Normocytic Anemia
- Elevated inflammatory serum and stool markers
- CT consistent with possible inflammatory process
- Negative stool infectious work-up
- Days into the admission, became septic and found to be newly HIV+

Initial Management:

Avoided starting steroids for possible Crohn's flare given unclear infectious etiology. Upper endoscopy and colonoscopy were preformed showing numerous violaceous nodules in the upper and lower GI tract consistent with Kaposi sarcoma (KS) (confirmed on immunohistochemistry).

Final treatment:

KS was staged as Tumor 1 (visceral disease), Immune system 1 (CD4 <200), Systemic illness 1 (B symptoms and opportunistic infections). Paclitaxel chemotherapy was started, but anti-retroviral therapy was held due to risk of immune reconstitution inflammatory syndrome

INPATIENT IMAGING/ENDOSCOPY:

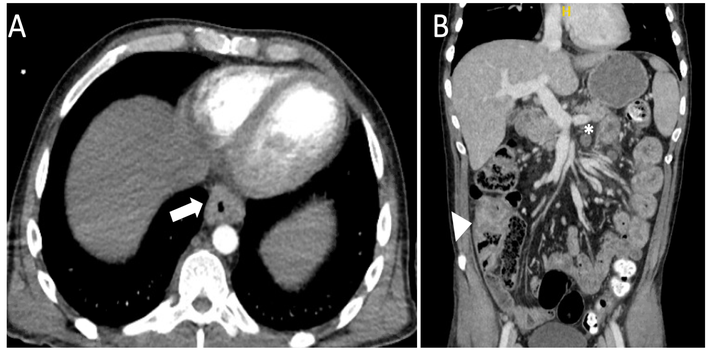

Figure 1 Computed Tomography Angiography with (A) mural thickening in the mid and distal esophagus (arrow) and (B) new mesenteric adenopathy (asterisk) and localized wall thickening of ascending colon (arrowhead).

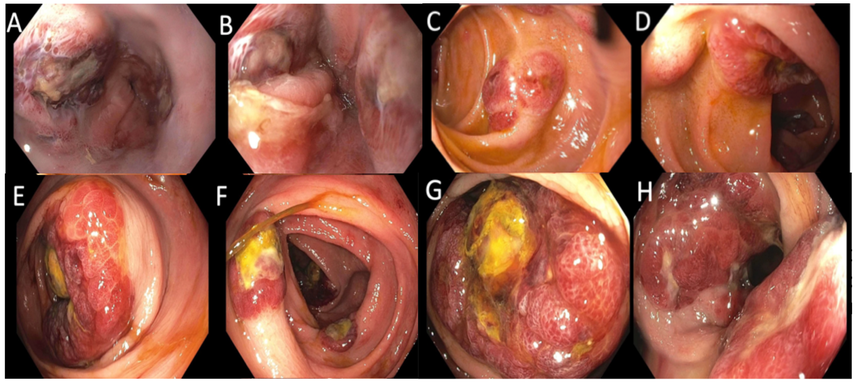

Figure 2. Endoscopy/Colonoscopy of multiple 10 to 50 mm violaceous nodules throughout the (A) distal esophagus, (B) gastroesophageal junction, (C, D) second portion of the duodenum (E) ileocecal valve, (F) ascending colon, (G) hepatic flexure and (H) rectum concerning for KS.

HISTOLOGY:

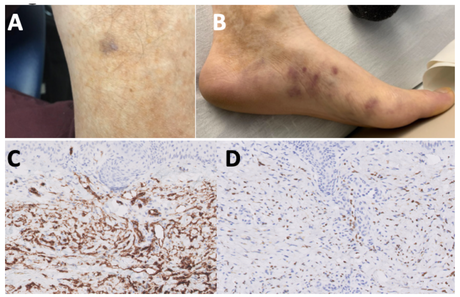

Figure 3. KS skin lesions on the left (A) leg and (B) foot, which were biopsied displaying positive (C) CD34, highlighting the vascular spaces, and (D) Human Herpes Virus 8, highlighting endothelial cell nuclei

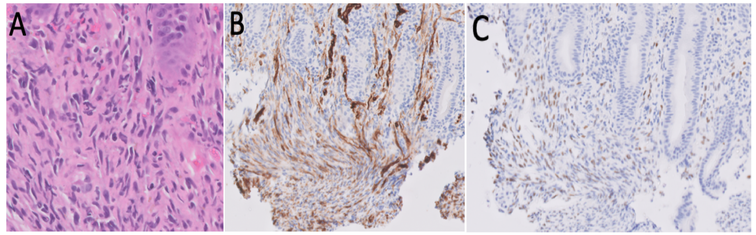

Figure 4. Ileocecal valve lesion biopsy: (A) lamina propria with atypical spindle cell proliferation with minimal pleomorphism and few extravasated red blood cells, with the atypical spindle cells positive for (B) CD34 and (c) HHV8 supporting the diagnosis of KS.

DISCUSSION:

Unique features of this case:

1. Kaposi Sarcoma (especially visceral KS) in patients with HIV and IBD is extremely rare.
2. Based on inflammatory markers and stool pathogens alone, it would have been reasonable to start high-dose steroids, however likely detrimental.

Take home points: Maintain a high index of suspicion for infectious etiology before starting high-dose steroids for an IBD flare. Further studies are required to understand the safety profile in treating IBD relapses in patients with uncontrolled HIV