Purpura Fulminans in Acute Severe Ulcerative Colitis Successfully treated with Infliximab

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Background

- Most common dermatological manifestations of Ulcerative Colitis
  - Erythema nodosum
  - Pyoderma gangrenosum
- Purpura fulminans is rapidly evolving syndrome of skin microvascular thrombosis and hemorrhagic necrosis
  - Purpuric rash characterized by coagulation of the microvasculature→ purpuric lesions and skin necrosis
  - Dermatologic emergency requiring rapid diagnosis and management
  - Patients are acutely ill with fever, can have hemorrhage from multiple sites and be in shock
  - Rapidly progressive syndromes accompanied by intravascular coagulation and circulatory collapse
  - Mortality rate has been decreasing with supportive care, but still a disabling condition--> major amputations in survivors
- Few reports have shown improvement of this entity with colectomy
- Case of purpura fulminans that developed in the setting of acute severe UC (ASUC) that responded to infliximab (IFX) therapy

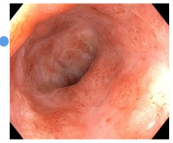
Discussion

- First case of purpura fulminans associated with severe acute ulcerative colitis managed with medical therapy
- Characterized by skin necrosis with thrombosis in dermal vessels, consumptive coagulopathy
- Derangements of the coagulation cascade may play a role
- Limb ischemia is a devastating complication
- Amputation may be necessary to reduce mortality
- Consider in patients with uncontrolled IBD who present with necrotic skin lesions as well as coagulopathy
- Trial aggressive medical therapy early on to halt progression of necrosis and save the colon
- If medical therapy fails→ colectomy

Case

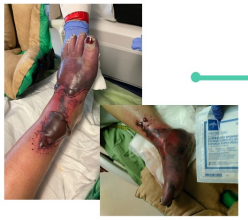
Past History: 50-year-old Female with mild pancolonic UC managed with oral mesalamine therapy for 1.5 years

1.5 years later – First presentation



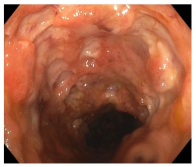
- Increased rectal bleeding and # of stools
- Colonoscopy→ mayo 2 pancolitis
- No dermatologic lesions
- Given oral steroids and started on IV vedolizumab therapy

2 months later – Current presentation



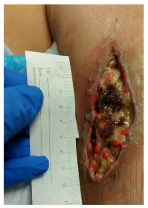
- Leg swelling
- Progressive necrotic rash of bilateral feet, left arm, and back in patches
- WBC: 35.7k, INR 1.7, CRP 227 (<7.4), Fibrinogen 129
- Factor V Leiden mutation PRESENT
- Concern for disseminated intravascular coagulation (DIC) (Initially thought Steven's Johnson's reaction to vedolizumab)
- Skin biopsy of back → microthrombi, no pus

1 day later – Colonoscopy



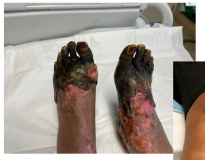
- Colonoscopy → Mayo 3 with deep ulcerations
- Biopsies = severe chronic active colitis
- 1 Cytomegalovirus (CMV) colony → course of ganciclovir
- C difficile indeterminant, negative stool aspiration → course of vancomycin
- Steroids → mild improvement in GI symptoms. not dermatologic patches
- Colectomy discussed; patient declined surgery
- Treatment: high dose of Remicade + Azathioprine

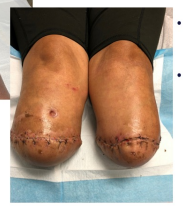
1 month later – Discharged to Skilled Nursing Facility



- In and out of hospital for skin infections
- Pyoderma gangrenosum ruled out by repeat skin biopsies

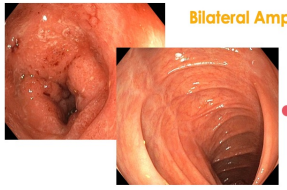
4 months later – Bilateral Amputation





- Bilateral feet → skin necrosis, dry gangrene
- No improvement → bilateral below the knee amputations

6 months later – present day



- GI symptoms under control
- CRP 6.6
- Iron indices, hgb improved
- Colonoscopy→ endoscopic remission aside from mild rectal inflammation

Contact Information

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References

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