A CASE REPORT

Granulomatosis with Polyangiitis Presenting as Crohn's Disease

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INTRODUCTION

Granulomatosis with Polyangiitis (GPA) is a systemic necrotizing vasculitis associated with antineutrophil cytoplasmic antibody (ANCA) that often presents with multiorgan involvement, frequently affecting the upper and lower respiratory tracts and kidneys.[1] While bowel involvement can occur, this is rare. Here, we describe a case of GPA with bowel involvement where the endoscopic appearance was like the "skip" lesions one may see in Crohn's disease (CD) with biopsies confirming the former diagnosis.

Case Presentation

An 11-year-old male presented to the emergency department with anorexia, a 5.48 kg weight loss over the past 5 months, persistent weakness, mouth ulcers, cough, persistent right acute otitis media (AOM), and bilateral testicular pain. His medical history included allergic rhinitis, recurrent AOM, malnutrition, and right epididymitis. Over the past two months, he exhibited a normal bowel pattern, but experienced increasing fatigue and weakness, decreased oral intake, and intermittent abdominal pain.

His physical exam revealed a cachectic adolescent with unexpected weight loss who weighed 21.82 kg (0.01 percentile); physical exam also demonstrated a purulent effusion in the right ear and a serous effusion in the left ear. Laboratory results exhibited normocytic anemia (Hgb 9.6 g/dL, MCV 85.1 fl), elevated inflammatory markers (WBC 17.7 K/mm3, ESR 62 mm/hr, CRP 10.6 mg/dL, calprotectin 1690 ug/g), elevated eosinophils (0.8 K/mm3), and a positive c-ANCA pattern with a high ANCA IFA titer (>1:1280). Nasal endoscopy revealed significant scarring and stenosis of the bilateral nasal cavities with minimal nasal patency. CT of the head and sinus revealed pansinusitis sequelae, while chest X-rays and CT demonstrated perihilar opacities and bilateral perihilar infiltrates/masses with necrotic foci, respectively. On upper endoscopy, two large esophageal ulcers were visualized adjacent to one another with biopsies showing normal surrounding mucosa. Colonoscopy revealed aphthous lesions reminiscent of "skip" lesions in the ascending

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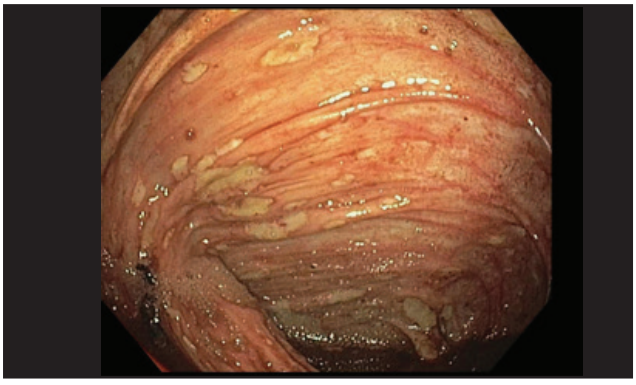

Figure 1. Colonic mucosa with skip lesion appearance and interspersed ulcerations surrounded by normal mucosa

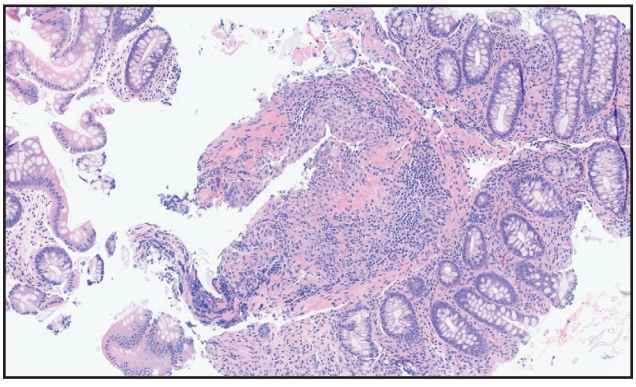

Figure 2. Colon Biopsy: Submucosal granuloma with no significant mucosal inflammation

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and transverse colon (Figure 1). Colon biopsies demonstrated submucosal vasculitis with associated poorly formed granulomas that lacked cryptitis (ie. inflammation that did not span the entire mucosal width) (Figure 2). There were also areas of fibrinopurulent exudate consistent with ulceration throughout the gastrointestinal tract and ischemic changes in the duodenum.

Based on these findings, the patient was diagnosed with granulomatosis with polyangiitis and treated with a methylprednisolone pulse (22 mg/kg on day one followed by 17.5 mg/kg after two days), three doses of rituximab (375 mg/m^2 at approximately one-week intervals), and an oral prednisolone taper (20 mg BID for seven days, weaned down to 25 mg once daily).

In subsequent follow up over a 6-month period, the patient demonstrated signs of clinical improvement, with resolution of anemia, normal inflammatory markers (i.e. ESR, CRP, and calprotectin), and a weight gain of 12.8 kg (22nd percentile); the patient continues to receive appropriate follow up care.

Discussion

This case explores a unique situation where GPA, previously Wegener's granulomatosis, manifested similarly to CD. CD is a chronic inflammatory bowel disease characterized by full-thickness inflammation of the bowel that impairs the lining of the digestive tract and can lead to abdominal pain, diarrhea, fatigue, weight loss, and malnutrition.[2] Of importance, CD may also present with various extraintestinal manifestations, including respiratory tract involvement, although this is uncommon.[3]

In our patient's case, the presentation was primarily gastrointestinal, characterized by symptoms such as decreased oral intake, early satiety, intermittent abdominal pain, and weight loss, which are more commonly associated with CD.[2] Histologically, the colon biopsies most prominently showed submucosal changes, including vasculitis and granulomas with what appeared to be secondary mucosal changes like ischemia and erosion (Figure 2). This contrasts with the transmural mucosal acute and chronic inflammation characteristic of inflammatory bowel disease. Additionally, laboratory results revealed normocytic anemia, elevated inflammatory markers, elevated eosinophils, and a positive c-ANCA pattern with a high ANCA IFA titer, indicative of GPA.[1]

While there have been reports of GPA with gastrointestinal symptoms similar to CD,[3,4] the occurrence of such symptoms is relatively low, being described in about 10-24% of GPA patients.[4] When juxtaposed with previous studies, our patient's presentation is distinct because our case involves a pediatric patient with both systemic and significant gastrointestinal symptoms,[3,4] which is highly atypical in a pediatric population, with an occurrence of GPA in this population being 1:1,000,000.[5] Furthermore, the presence of CD or GPA manifesting as a mimic of the other condition has generally only been reported in adults (two previously published case reports with patients aged 29 and 41),[6,7] potentially making this case, to the best of our knowledge, the second instance of a pediatric patient who's diagnosed GPA presents as CD without also having a concurrent diagnosis of CD.[8]

CONCLUSION

In conclusion, this case underscores the importance of considering GPA in the differential diagnosis when a patient presents with gastrointestinal symptoms akin to CD, especially when accompanied by systemic symptoms, respiratory symptoms, and a positive c-ANCA pattern.[3,4] Further research is warranted to understand the relationship between GPA and CD and to enhance the diagnosis and treatment of these conditions. ■

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