A Case of Significant Gastrointestinal Involvement in Granulomatosis with Polyangiitis

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Granulomatosis with polyangiitis (GPA) is a rare, autoimmune condition usually presenting with pulmonary and renal involvement. There are not many reports of gastrointestinal involvement and even fewer cases of disseminated disease. We present a case of biopsy-proven, active gastrointestinal GPA throughout the stomach, small intestine and colon. Patients may present with diffuse abdominal pain, making the diagnosis difficult. We suggest that unidentified or inadequately managed gastrointestinal involvement in GPA is a potentially serious condition. This disease manifestation must be maintained in the differential in at-risk patients with gastrointestinal symptoms, and appropriate imaging should be considered when clinical suspicion warrants.

INTRODUCTION

Granulomatosis with polyangiitis (GPA) is a rare autoimmune condition usually presenting with pulmonary and renal involvement. The prevalence of this condition is 3 in 100,000 people in the United States. There are numerous studies documenting cerebral, cutaneous and cardiac manifestations, however, few reports of gastrointestinal involvement exist. Even in patients with known GPA, abdominal symptoms are significantly more likely to represent common underlying etiologies rather than the vasculitis itself. GPA has been included in a group of systemic vasculitides that have been described as “great masqueraders” due to the ability of this condition to manifest itself in ways that differ widely from their more typical presentation. For this reason, it is important to consider GPA when evaluating patients with a history of GPA who have abdominal complaints after more common etiologies have been ruled out.

CASE

A 38-year-old woman presented with several months of postprandial periumbilical abdominal pain. Accompanying symptoms included anal pain upon defecation, fatigue, malaise, night sweats, weight loss, arthralgias and decreased oral intake. Her medical history was significant for granulomatosis with polyangiitis, manifested by pulmonary, renal, sinus and ocular involvement, in remission for three years and off of therapy. Her exam was notable for abdominal tenderness, supraclavicular lymphadenopathy, and a perianal ulceration. Initial workup was concerning for new onset anemia and a positive fecal occult blood test. Upon admission, upper endoscopic and colonoscopic evaluations were performed. Patchy erythematous mucosa, with erosions, was identified in the gastric body, antrum, duodenum, terminal ileum and descending colon; a large ulceration was seen in the splenic flexure.

(continued on page 52)
With the exception of the ileal biopsies, neutrophilic infiltration and reactive capillary endotheliitis without granulomata, consistent with active gastrointestinal GPA, was noted pathologically. Additionally, left anterior cervical lymph node biopsy revealed low-grade follicular lymphoma, which was also noted in the terminal ileum. She was treated with high-dose glucocorticoids with pending transition to rituximab.

DISCUSSION

This is a rare case of gastrointestinal GPA in a young female with multiple organ system involvement. GPA is an ANCA-associated vasculitis, and most commonly presents with upper respiratory, pulmonary and renal manifestations, as seen in our patient.\(^1,4\) Cutaneous, ocular and nervous system manifestations are less common but still described regularly.\(^1,2\) Gastrointestinal involvement is not commonly seen.\(^1,6\) Often, these cases are discovered after an extensive evaluation culminating in endoscopic evaluation with confirmatory pathology. Most of the described cases of intestinal involvement have been noted intraoperatively during bowel perforation repair. Given the potential severity of unidentified or inadequately managed gastrointestinal involvement in GPA, this disease manifestation must be considered in at-risk patients with gastrointestinal symptoms. Appropriate endoscopic workup with biopsies should be performed when clinical suspicion warrants. Missing a diagnosis of gastrointestinal GPA can have a serious impact on morbidity or mortality of the patient as life-threatening consequences, such as bowel ischemia and perforation, are potential complications.\(^3,6\) Early recognition and management are paramount for appropriate patient care.

References