

Adult-Onset Immunoglobulin A Vasculitis

Matthew Chadwick, DO¹, Leonid Shamban, DO², and John Macksood, DO³

¹Department of Gastroenterology, Ascension Genesys Hospital, Grand Blanc, MI

²GI Medicine Associates, St Clair Shores, MI

³Flint Gastroenterology Associates, Grand Blanc, MI

ABSTRACT

Immunoglobulin A vasculitis (IgAV), formerly Henoch–Schönlein purpura vasculitis, is a vasculitis commonly seen in children and only rarely described in adult patients. IgAV can present as arthralgia, rash, discolored urine, acute kidney injury, and gastrointestinal symptoms. We present a case of a 56-year-old man who presented with 1 month of worsening abdominal pain followed by a bilateral lower extremity rash. Laboratory evaluation indicated an acute kidney injury. Computed tomography and colonoscopy revealed terminal ileitis while kidney biopsy confirmed a diagnosis of IgAV.

INTRODUCTION

Immunoglobulin A vasculitis (IgAV), formerly Henoch–Schönlein purpura, is a systemic vasculitis characterized by immunoglobulin A (IgA)-dominant immune deposits affecting small blood vessels.^{1–3} It commonly presents with palpable purpura, arthralgia, discolored urine, and acute kidney injury (AKI).^{2–5} Approximately two-thirds of patients present with gastrointestinal (GI) symptoms including abdominal pain, nausea, vomiting, and edema.³ These symptoms are caused by ischemia and edema, and in some cases, arthralgia can lead to infarction, intussusception, or perforation of the bowel.^{3,6} IgAV is relatively common in childhood, accounting for up to 45% of pediatric vasculitides, whereas it is a rare vasculitis in adults.^{1–3} Typically, adults who develop IgA vasculitis have a more severe disease course and require more aggressive therapies.^{1,2,4} Any portion of the GI tract may be affected; however, the small bowel, and in particular the duodenum, is the most commonly affected site.² Because IgAV can cause nonspecific GI symptoms and inflammation throughout the GI tract, it can be difficult to differentiate from inflammatory bowel disease (IBD), especially from Crohn's disease.^{5,7}

CASE REPORT

A 56-year-old white man with a medical history of hypertension and diabetes mellitus developed worsening abdominal pain for 1 month. Before his symptoms, he had daily bowel movements with no GI bleeding. He developed symptoms of constipation and was started on polyethylene glycol by his primary care physician with initial improvement. He subsequently developed a bilateral lower extremity rash and was diagnosed with a leukocytoclastic vasculitis by a dermatologist. The patient was treated with oral sulfamethoxazole-trimethoprim.

His rash improved; however, because of worsening abdominal pain, he presented to the emergency department. On evaluation, his vital signs were unremarkable. His abdominal examination revealed a soft, nontender, nondistended abdomen with bowel sounds present. The dermatological examination revealed a nonpalpable purpura on the posterior aspect of his bilateral lower extremities.

Initial laboratory evaluation revealed a leukocytosis of $14.5 \times 10^9/L$ and AKI with a creatinine of 2.15 mg/dL. Computed tomography with intravenous contrast of the abdomen revealed terminal ileitis (Figure 1). Evaluation for multiple myeloma, cryoglobulinemia, and antineutrophil cytoplasmic antibody-associated vasculitis was negative. Initially, his kidney function improved with rehydration, and his abdominal pain resolved with an aggressive bowel regimen.

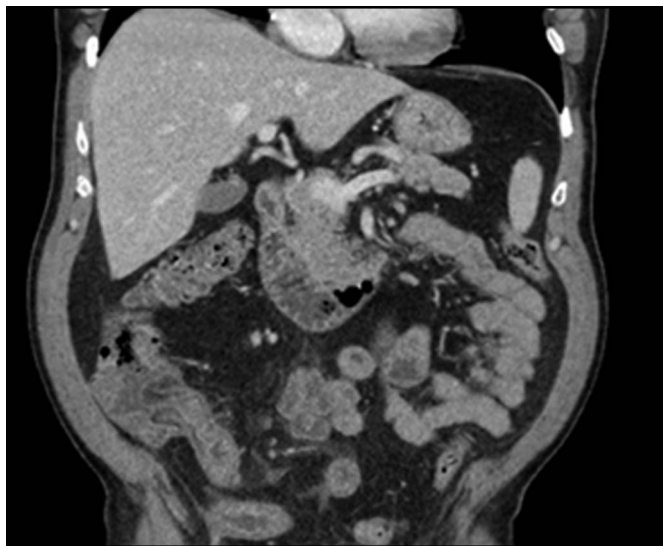


Figure 1. Computed tomography showing terminal ileitis.

He was admitted to the observational unit and evaluated by the gastroenterology team. Further laboratory evaluation revealed a C-reactive protein of 75 mg/L and an erythrocyte sedimentation rate of 21 mm/hr. Because of these findings, a colonoscopy was performed which showed terminal ileitis and a 4-mm cecal adenomatous polyp (Figure 2). Terminal ileal biopsies revealed an ulcer with focally active ileitis and hemorrhage (Figure 3). Given these findings, recurrent AKI renal biopsy was performed which showed IgA glomerulonephritis (Figure 4). He was given 1,000 mg methylprednisolone and then 500 mg methylprednisolone the following day, and subsequently, his kidney function improved with complete resolution of abdominal pain. He was discharged home on 90 mg prednisolone daily.

Although IgAV is classically associated with children, it can be a rare diagnosis in adults. The classic presentation of rash,

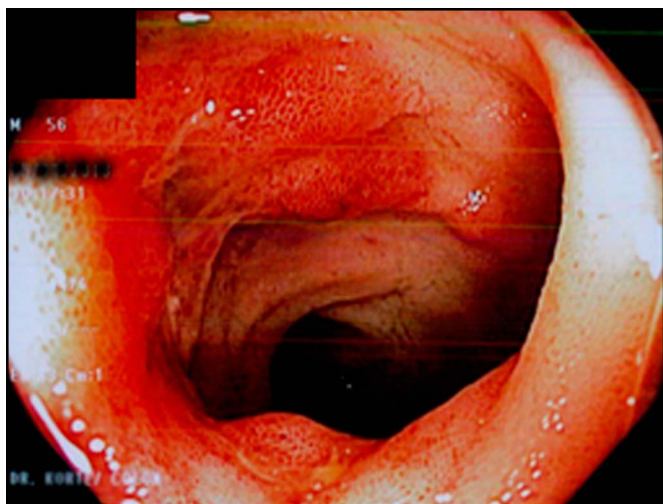


Figure 2. Colonoscopy showing terminal ileitis and a 4-mm polyp.

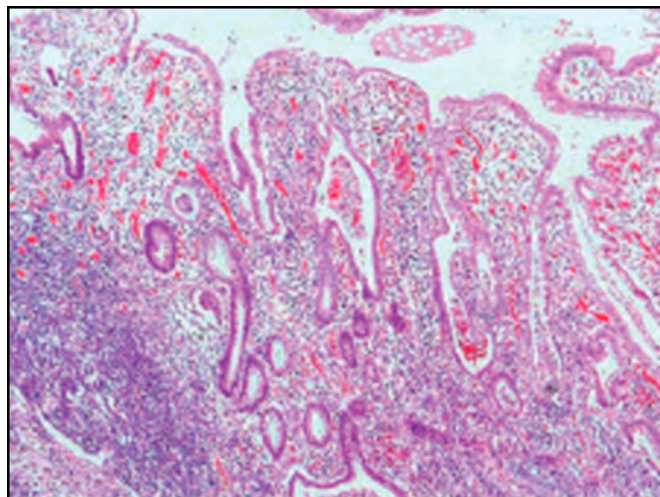


Figure 3. Terminal ileum biopsy showing ileitis.

arthralgia, and GI symptoms can mimic the presentation of IBD, especially Crohn's disease. The classic computed tomography findings for IgAV involving the GI tract are multiple focal areas of bowel thickening with skip lesions, mesenteric edema, and nonspecific lymphadenopathy and therefore is not helpful in distinguishing it from IBD.⁸ Serologic testing also cannot distinguish between the 2 diseases because one study found that nearly two-thirds of the cases of IgAV in adults had C-reactive protein levels of greater than 8 mg/dL.⁹

IgAV is the most common vasculitis to affect the upper GI tract with one study finding it to account for 56.8% of all cases.¹⁰ Although the duodenum is the most common location affected by IgAV, any portion of the GI tract may be affected, including less commonly the terminal ileum.¹¹ Because of their similar presentations, endoscopic evaluation can aid in making a diagnosis because IgAV will classically have endoscopic findings

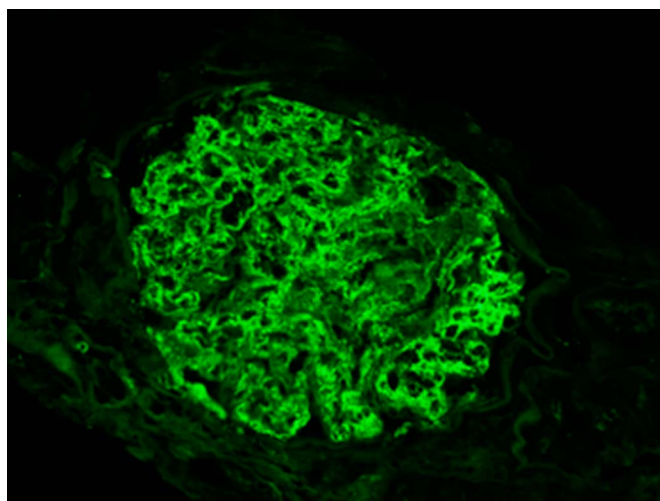


Figure 4. Renal biopsy with immunofluorescence staining showing IgA deposits.

of diffuse mucosal erythema, petechiae, hemorrhagic erosions, and longitudinal ulcers.^{8,11,12} A biopsy is often needed to help establish a distinction between IBD and IgAV, although there has been at least one case report of IgAV with endoscopic biopsies consistent with IBD.¹³ Although biopsy of the leukocytoclastic rash which accompanies IgAV will distinguish it from IBD, up to 14% of the cases will present with abdominal symptoms before the manifestation of a rash.¹⁴

It is important to make an accurate diagnosis between IgAV and IBD because the long-term treatment is different for the diagnoses. One study found that the most commonly used treatment in IgAV being renin-angiotensin-aldosterone system blockers and corticosteroids.¹⁵ Although many medications can be used for the long-term treatment of IBD, that same study found that azathioprine was the only medication also used in the treatment of IgAV and was only found to be used in 6.1% of all adults with IgAV.¹⁵

Early initiation of appropriate treatment has shown to improve the prognosis of renal complications associated with IgAV.^{4,16} Because adults face a more severe disease course compared with pediatric patients and require more intense treatment, it is important to avoid a delay in making a diagnosis of IgAV. Although rare, findings of terminal ileitis can be a sign of vasculitis, even in adult patients and those with slightly atypical presentations. This should prompt physicians to include vasculitides into their differential.

DISCLOSURES

Author contributions: M. Chadwick wrote the manuscript and is the article guarantor. L. Shamban and J. Macksood edited the manuscript.

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