Bilal Hameed, Uma Mahadevan, and Kay Washington, Section Editors

Abdominal Pain as the First Manifestation of a Systemic Disease

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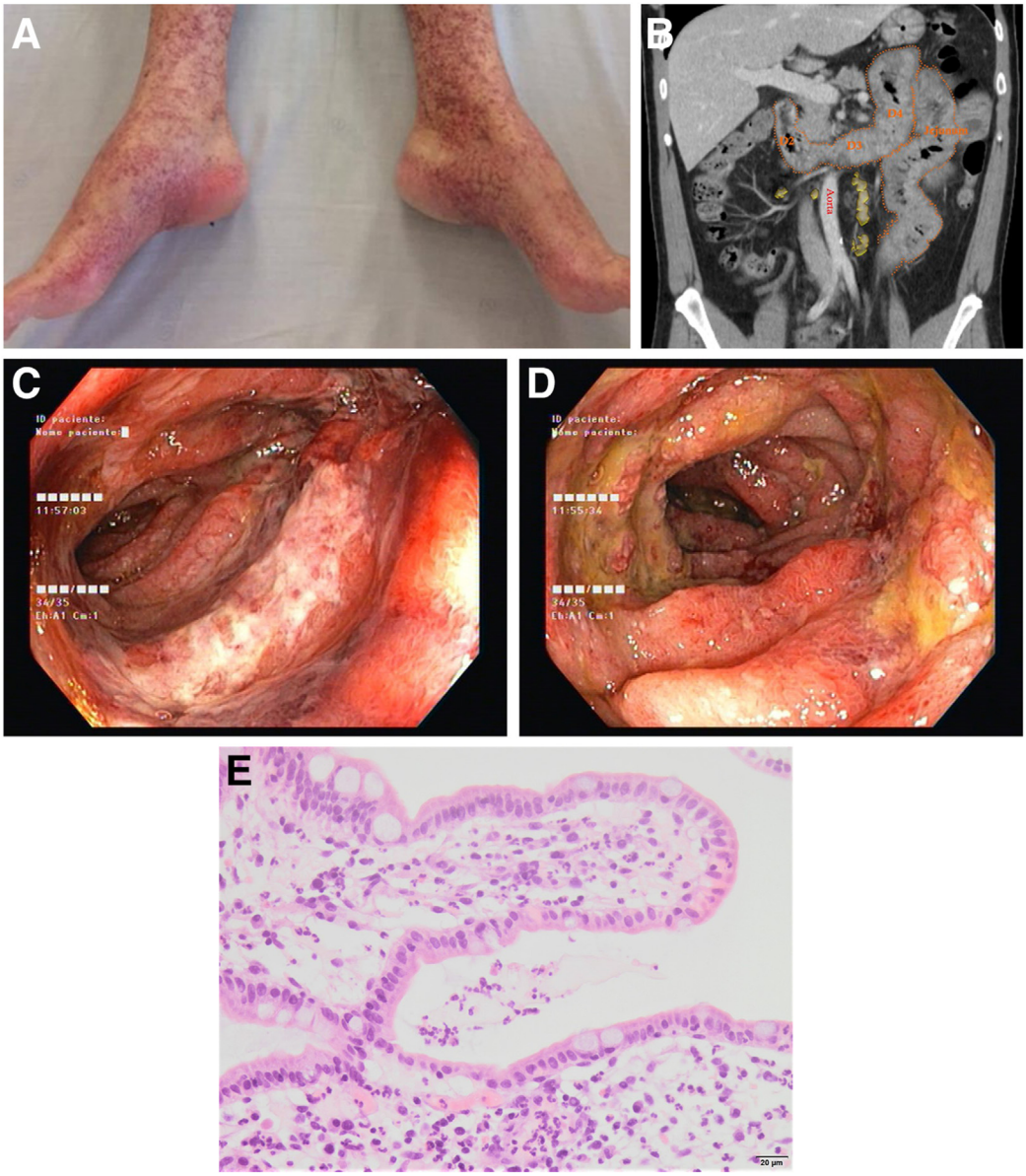

Question: A 41-year-old man without a significant previous medical history presented with a 7-day history of moderate upper abdominal pain, nausea, and anorexia. On physical examination, the patient had left upper abdominal quadrant tenderness. Two days after admission, he developed a symmetrical nonpruritic macular rash on both feet, that evolved to palpable purpura and extended to both knees (Figure *A*).

Laboratory investigation revealed neutrophilic leukocytosis (26,300/m^3 white blood cells and 20,020/m^3 neutrophils), elevated C-reactive protein (144 mg/L), hypoalbuminemia (2.5 g/dL), and folate deficiency (1.8 ng/mL). Urinalysis showed

hematuria and nephrotic-range proteinuria. The immunologic profile was unremarkable. Viral infections were excluded and blood, urine and stool cultures were negative.

A contrast-enhanced computed tomography scan showed a 10-cm-long asymmetric proximal jejunal wall thickening with mural stratification and multiple lymph node enlargements (Figure *B*). Esophagogastroduodenoscopy showed diffuse mucosal congestion and edema along duodenum with multiple erosions and deep ulcers starting in the bulb–D2 transition and worsening distally (Figure *C*, *D*). Biopsies were taken and histologic examination revealed inflammatory infiltrate with polymorphonuclears (Figure *E*).

What might be the diagnosis and how should it be managed?

Look on page 1383 for the answer and see the *Gastroenterology* website (www.gastrojournal.org) for more information on submitting your favorite image to Clinical Challenges and images in GI.

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Conflicts of interest
The authors disclose no conflicts.

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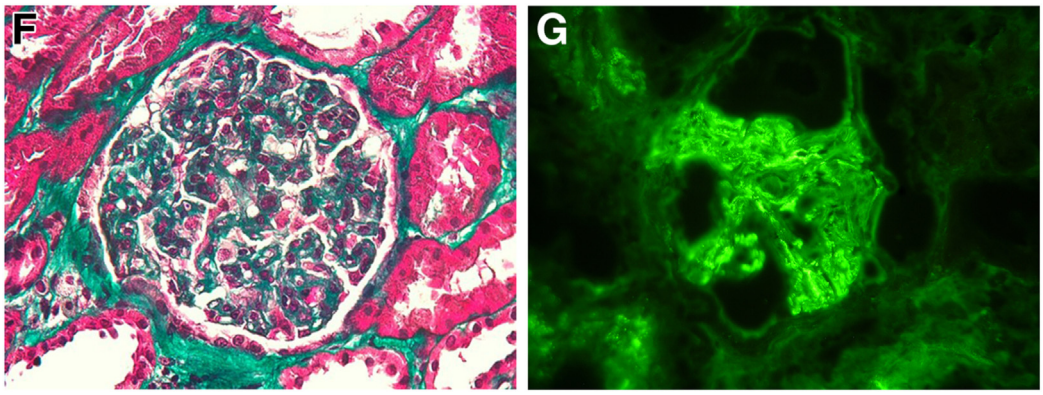
Answer to: Image 1 (Page 1381): Immunoglobulin A Vasculitis



Based on the triad of abdominal pain, nonthrombocytopenic palpable purpura, and urinalysis alterations the presumed diagnosis of immunoglobulin A vasculitis (IgAV) was made.

The purpuric lesions were biopsied and revealed leukocytoclastic vasculitis with no IgA deposits. A renal biopsy was also performed due to the urinalysis severe alterations and it confirmed the diagnosis, showing a mesangial proliferative glomerulonephritis (Figure *F*) with diffuse IgA deposits on immunofluorescence microscopy (Figure *G*).

IgAV, formerly called Henoch–Schönlein purpura, is an immune complex–mediated systemic vasculitis involving small vessels and >90% of the cases occur in children <10 years of age. In adulthood, the disease is rare, has atypical manifestations, and is more severe with a worse prognosis.[1,2]

The diagnosis of IgAV is based on clinical manifestations, typically characterized by a classic tetrad: purpura, arthralgias, abdominal pain, and urinalysis alterations. The palpable purpura is an universal finding and is predominantly located in the lower limbs. The definitive diagnosis of IgAV invariably requires histologic confirmation. The abnormal vascular IgA deposits identified by direct immunofluorescence are the defining histologic feature of IgAV.[1–3] In the presented case, the diagnosis was particularly challenging because there was no arthralgia and the abdominal pain preceded the cutaneous rash.

IgAV treatment is controversial and is usually supportive. There is some evidence to support steroid therapy in treatment of severe manifestations. However, the routine use of corticosteroids is debatable.[1–3] This patient was treated with empirical antibiotics (ceftriaxone and metronidazole) with significant clinical and imaging improvement. We hypothesize that he had an acute gastrointestinal infection overlapping IgAV, because he had elevation of acute phase markers, duodenal histology was suggestive of acute infection and there was a clearly improvement with antibiotic therapy. This hypothetical infection might have been the trigger of IgAV.

This patient was discharged asymptomatic 7 days after admission. He was referred to a nephrology outpatient clinic and treated with corticosteroids owing to severe renal involvement. He had complete response to therapy, achieving a sustained remission after treatment withdrawal.

Keywords: Enteritis; IgA Vasculitis; Vasculitis; Henoch-Schönlein Purpura.

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