IgA vasculitis presenting as abdominal pain and rash

Anil Kopparapu, MD, MPH , Diane Jarrett, EdD , and Shashank Kraleti, MD

Department of Family and Preventive Medicine, University of Arkansas for Medical Sciences, Little Rock, Arkansas

ABSTRACT

A 57-year-old white man presented with acute abdominal pain and rash without any prodromal symptoms. The skin biopsy confirmed immunoglobulin A (IgA) vasculitis with small vessel vasculitis and perivascular IgA, C3, and fibrin deposition. IgA vasculitis is diagnosed most commonly in children and presents in adults in only 10% of cases. Treatment is usually supportive care, and interventions may be required to avoid complications such as nephrotic syndrome or acute gastrointestinal bleeding. Clinicians should be aware of IgA vasculitis as a potential cause for abdominal pain and rash in adult populations.

KEYWORDS Abdominal pain; Henoch-Schönlein purpura; IgA vasculitis; palpable purpura

Immunoglobulin A vasculitis (IgAV) is characterized by the classic tetrad of palpable purpura, arthritis or arthralgia, hematuria, and abdominal pain.¹ It primarily affects children under 10 years old (90% of cases) and is rare in adults.² We present the case of a man who presented with complaints of abdominal pain and rash, which was subsequently diagnosed as IgAV.

CASE REPORT

A 57-year-old white man presented to the emergency department with a rash of 5 days' duration and constant, generalized abdominal pain for 1 day. He denied nausea, vomiting, or diarrhea. The rash started on his legs and gradually spread to his upper extremities and abdomen. A review of systems was positive for fatigue, chronic back pain, and headache. He had known diabetes mellitus type 2, hyperlipidemia, bipolar disorder, and chronic low back pain. His medications included metformin, famotidine, naproxen, valproic acid, and ziprasidone. He was a former smoker, having quit 20 years ago. Though he had a significant history of alcohol use, he quit about 6 months earlier. His father had premature coronary artery disease.

His vital signs were within normal limits. Multiple nonpruritic, erythematous petechial and purpuric macules and papules were noted over the abdomen, lower back, and upper and lower extremities (*Figure 1*). Mild to moderate tenderness was noted in the right lower quadrant of the abdomen. The remainder of the physical exam was within normal limits. Significant

Figure 1. Palpable purpura involvement of the (a) abdomen and (b) bilateral lower extremities.

laboratory findings were a mildly elevated white blood cell count (11.90 K/ μ L) and an elevated erythrocyte sedimentation rate (40 mm/hr) and C-reactive protein (20.20 mg/L). Urinalysis was negative for leukocyte esterase, nitrite, blood, glucose, and protein. Urine and blood cultures revealed no growth. Serology findings were nonreactive/negative for hepatitis A IgM, hepatitis B surface antigen, total anti-hepatitis B core antibody, anti-hepatitis B core antibody IgM, hepatitis C virus, and HIV P24 antigen screening and antibodies. No infectious etiology was noted. IgA levels (298 mg/dL), complement levels, renal function tests, and coagulation profile were normal (prothrombin time 12.4 seconds, activated partial thromboplastin time 28.6 seconds, international normalized ratio 1.1).

Corresponding author: Anil K. Kopparapu, MD, MPH, Department of Family and Preventive Medicine, University of Arkansas for Medical Sciences, 521 Jack Stephens Dr. #530, Little Rock, AR 72205 (e-mail: akkopparapu@uams.edu)

Color versions of one or more of the figures in the article can be found online at www.tandfonline.com/ubmc.

Received February 27, 2019; Revised June 3, 2019; Accepted June 5, 2019.

Figure 2. Granular immunoglobulin A deposition within the vascular wall of the papillary dermis (arrow). Direct immunofluorescence 400 $\times$.

A punch biopsy of the left thigh showed small vessel vasculitis. Direct immunofluorescence revealed perivascular IgA, C3, and fibrin deposition consistent with IgAV (Figure 2). A computed tomography scan of the chest showed no concerning pulmonary nodules or masses. Abdominal computed tomography scan showed nonspecific mesenteric and retroperitoneal adenopathy and cholelithiasis without evidence of acute cholecystitis; the appendix was normal and retrocecal in position. After 2 days of 60 mg oral prednisone, the patient's rash and abdominal pain resolved. He completed a 2-week course of prednisone and reported complete resolution of the rash and abdominal pain.

DISCUSSION

IgAV is an immune-mediated disorder characterized by a generalized vasculitis involving the small vessels of the skin, gastrointestinal tract, kidneys, and joints.^{1,2} Ninety percent of cases are seen in the pediatric population. The incidence of IgAV in children is approximately 6 to 22 per 100,000 person-years.³ The incidence in adults varies from 3.4 to 14.3 cases per million.⁴⁻⁶

IgAV is thought to be multifactorial, with genetic, environmental, infectious, and antigenic contributors.¹ IgAV typically has a prodrome that includes headache, anorexia, and fever, followed by symptoms such as rash, abdominal pain, bloody stools, and joint pain. A nonmigratory arthritis occurs in 75% of patients, and abdominal pain occurs in 65% of patients with IgAV.⁵ Renal disease is the most serious sequela, occurring in 40% to 50% of patients.⁶ The differential diagnosis includes thrombocytopenic purpura, idiopathic thrombocytopenic purpura, acute glomerulonephritis, meningococcal infections, bacterial endocarditis, hypersensitivity vasculitis, Wegener's granulomatosis, systemic lupus erythematosus, and other viral exanthemas.

There is no definitive blood test to diagnose IgAV. Routine laboratory tests are usually within normal ranges. The characteristic finding of IgAV is leukocytoclastic vasculitis accompanied by IgA immune complexes within affected organs. Skin biopsy is essential in confirming the diagnosis. Renal biopsy may be helpful and should be performed when nephrotic syndrome is suspected and when renal function deteriorates.⁷ Potential complications of IgAV include glomerulonephritis, nephrotic

syndrome, gastrointestinal hemorrhage, alveolar hemorrhage, cerebral hemorrhage, neuropathy, seizures, orchitis, anterior uveitis, myositis, and myocarditis.

Most patients recover quickly with supportive care, including adequate hydration and follow-up.⁸ Hospitalization should be strongly considered with severe abdominal pain, significant gastrointestinal bleeding,⁹ or marked renal insufficiency. Corticosteroids are often used to treat subcutaneous edema and nephritis. Corticosteroids, cyclophosphamide, plasma exchange, and intravenous immunoglobulin need to be considered in severe gastrointestinal hemorrhage,^{9,10} severe pulmonary involvement,¹¹ or neurological involvement.

IgAV should be considered as a diagnosis after a negative initial evaluation in adults presenting with rash and abdominal pain. Appropriate tissue pathology is essential to confirm the diagnosis. Treatment is supportive because the disease course is usually benign, but interventions may be required to avoid complications.

ORCID

Anil Koppurapu <http://orcid.org/0000-0002-4727-4573>

Diane Jarrett <http://orcid.org/0000-0002-9968-7900>

Shashank Kraleti <http://orcid.org/0000-0002-7363-8510>

1. Piram M, Mahr A. Epidemiology of immunoglobulin A vasculitis (Henoch-Schönlein). *Curr Opin Rheumatol*. 2013;25:171–178. doi:10.1097/BOR.0b013e32835d8e2a.
2. Hocevar A, Rotar Z, Jurčić V, et al. IgA vasculitis in adults: the performance of the EULAR/PRINTO/PRES classification criteria in adults. *Arthritis Res Ther*. 2016;18:58. doi:10.1186/s13075-016-0959-4.
3. Gardner-Medwin JM, Dolezalova P, Cummins C, Southwood TR. Incidence of Henoch-Schönlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. *Lancet*. 2002;360:1197–1202. doi:10.1016/S0140-6736(02)11279-7.
4. Younger DS. Epidemiology of the vasculitides. *Neurol Clin*. 2019;37:201–217. doi:10.1016/j.ncl.2019.01.016.
5. Blanco R, Martínez-Taboada VM, Rodríguez-Valverde V, García-Fuentes M, Gonzalez-Gay MA. Henoch-Schönlein purpura in adulthood and childhood. Two different expressions of the same syndrome. *Arthritis Rheum*. 1997;40:859–864. doi:10.1002/art.1780400513.
6. Saulsbury FT. Clinical update: Henoch-Schönlein purpura. *Lancet*. 2007;369:976–978. doi:10.1016/S0140-6736(07)60474-7.
7. García-Porrúa C, González-Louzao C, Llorca J, González-Gay MA. Predictive factors for renal sequelae in adults with Henoch-Schönlein purpura. *J Rheumatol*. 2001;28:1019–1024.
8. Ronkainen J, Koskimies O, Ala-Houhala M, et al. Early prednisone therapy in Henoch-Schönlein purpura: a randomized, double-blind, placebo-controlled trial. *J Pediatr*. 2006;149:241–247. doi:10.1016/j.jpeds.2006.03.024.
9. Yamazaki T, Akimoto T, Iwazu Y, et al. Henoch-Schönlein purpura complicated with severe gastrointestinal bleeding. *CEN Case Rep*. 2015;4:106–111. doi:10.1007/s13730-014-0148-8.
10. Uluca Ü, Ece A, Şen V, Yel S, Tan I, Karabel D. Management of intestinal bleeding with single-dose cyclophosphamide in Henoch-Schönlein purpura. *Pediatr Int*. 2015;57:498–500. doi:10.1111/ped.12670.
11. Stienstra Y, Fijen JW, Tervaert JW, Dankbaar H, Zijlstra JG, van der Werf TS. Pulmonary hemorrhage with respiratory insufficiency in Henoch-Schoenlein purpura [in Dutch]. *Ned Tijdschr Geneesk*. 2000;144:617–622.