# An adult male with abdominal pain and skin rash

**Jay Patel MD, Imran Umer MD, Anand Reddy MD,
Yasir Ahmed MD, Craig Spellman DO, PhD**

## ABSTRACT

*Henoch-Schönlein purpura is generally a disease of children and uncommon in adults, especially after the age of 40. It is characterized by leukocytoclastic vasculitis skin rash, arthralgia, and gastrointestinal symptoms. In adults, glomerulonephritis may occur and the long-term prognosis is poor. We present the case of a 65-year-old man with complaints of abdomen pain and skin rash who subsequently developed renal failure. He was diagnosed with adult onset Henoch-Schönlein purpura and survived after treatment with glucocorticoids and hemodialysis.*

***Key words:*** Henoch-Schönlein purpura, HSP, IgA vasculitis, leukocytoclastic vasculitis, acute renal failure, skin rash

## INTRODUCTION

Henoch-Schönlein purpura (HSP) involves the small blood vessels and presents with a purpuric skin rash, arthralgia, and gastrointestinal symptoms. HSP is usually an autoimmune disease in children and occurs after viral infections. HSP is rare in adults, is more severe, and carries a poor prognosis compared to children.[1] Glomerulonephritis is seen in approximately 50% of patients and is associated with a poor long-term prognosis.[2] Mortality in adults with HSP is usually due to gastrointestinal complications, including ischemic colitis, perforations, and bleeding.[3] Here we report a case of adult onset HSP who developed renal failure and survived after a prolonged hospital course requiring supportive care, glucocorticoid therapy, and temporary hemodialysis.

***Corresponding author:*** Yasir Ahmed MD
***Contact Information:*** Yasir.ahmed@ttuhsc.edu
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## CASE PRESENTTION

A 65-year-old Caucasian man with history of hypertension presented with abdominal pain and a skin rash with swelling involving both lower extremities for one week. Prior to admission, he visited an urgent care clinic and received an intramuscular injection of methylprednisolone. He developed nausea and vomiting with increased abdominal pain and was admitted in our hospital. He also complained of diffuse arthralgia but denied fever, chills, diarrhea, recent travel, and contact with ill persons. He denied any prior history of skin rashes or lesions. Vital signs included temperature 98°F, blood pressure 130/82 mmHg, pulse rate 92 beats/minute, respiratory rate 20 breaths/minute, and oxygen saturation 95% on room air. Physical examination was significant for diffuse abdominal tenderness and petechial skin lesions involving the legs and feet. (Figure-1)

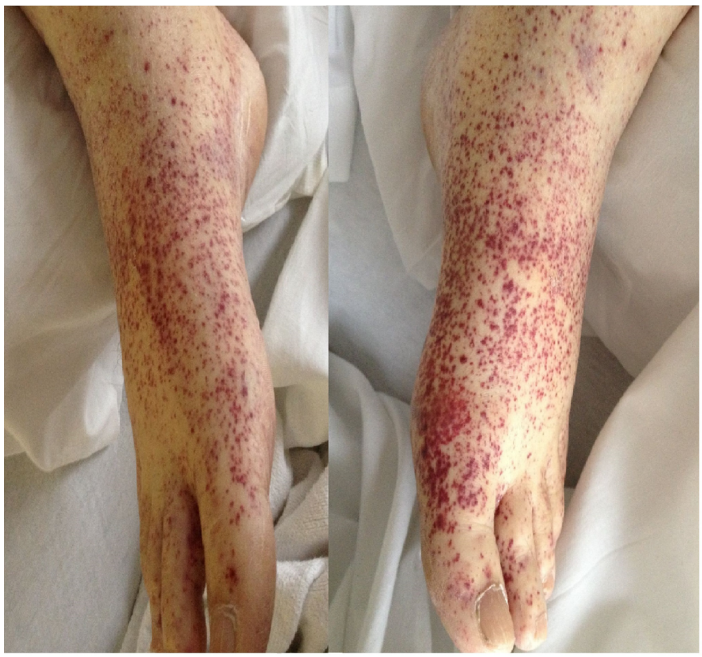

**Figure 1.** Right and left foot and ankle showing petechial and purpuric skin rash

The initial laboratory studies revealed white blood cell count 17x $10^3$/μL (polymorph 91%, lymphocytes 6%), hemoglobin 14 gm/dL, platelets 41x$10^3$/μL, erythrocyte sedimentation rate 8 mm/hr. Liver function tests and serum electrolytes were normal except for creatinine of 1.6 mg/dL. Urine studies showed protein 100 mg/dL and 10 red blood cells per high power field. Repeat urine protein studies showed proteinuria at 2014 mg per 24 hrs. A computed tomography scan of the abdomen reported nonspecific thickening of the large intestine and consistent with early ischemic colitis. Empiric antibiotic therapy was begun and an infectious disease work up was done for HIV, hepatitis B and C, and bacterial infections. By hospital day three, the petechial rash extended up to the abdomen, buttocks, and hands, and anasarca was evident. During the next five days, the patient became anuric and a transaminitis developed with increased aspartate aminotransferase levels (106 IU/L) and alanine aminotransferase levels (91 IU/L). His serum creatinine increased to 5.1 mg/dl, and the BUN increased to 116 mg/dl. All infectious disease work ups were reported to be negative so antibiotics were stopped on hospital day eight. Serologic studies for vasculitis of various etiologies (ANCAs, ANA, cryoglobulins, etc.) were negative. Based on the skin lesions, the abdominal pain and renal failure, the clinical diagnosis of HSP was made. Subsequently, skin and renal biopsies were performed and leukocytoclastic vasculitis and IgA nephropathy with chronic inflammatory changes was reported, respectively (Figure 2a, 2b, 2c). Methylprednisolone 500mg intravenous twice a day was administered on hospital days 5-8, and the patient was then switched to oral prednisone 90 mg per day. Hemodialysis was started on hospital day eight for acute renal failure and anuria. The patient slowly improved with resolution of the arthralgia, anasarca, abdominal pain and skin lesions. Dialysis was discontinued after three weeks, but oral prednisone was continued and weaned off after three months. At six weeks follow up visit as an outpatient, the patient remained symptom free with return of renal function to base line.

## *Discussion*

HSP is typically a disease of children and is very uncommon in adults.[4,5] The incidence in adults varies from approximately 3 to 14 cases per million, depending upon the particular study and patient ethnicity.[6] It is an immune-mediated systemic leukocytoclastic vasculitis characterized by vascular and/or mesangial IgA deposition.

The etiology of HSP remains unknown; it may be triggered by a variety of antigenic stimuli including infections, drugs, toxins, systemic diseases and cancer.[7,8] In the present case infectious gastroenteritis was possibly a triggering factor. Episodes of macroscopic hematuria are frequently associated with acute renal failure and HSP-associated IgA nephropathy leads to severe renal failure (CrCl <30 ml/min) in 13% and end stage renal disease in up to 11% of adult patients on long term follow-up.[1] Proteinuria, hypertension, and initial kidney impairment portend an unfavorable renal prognosis with the degree of pro-

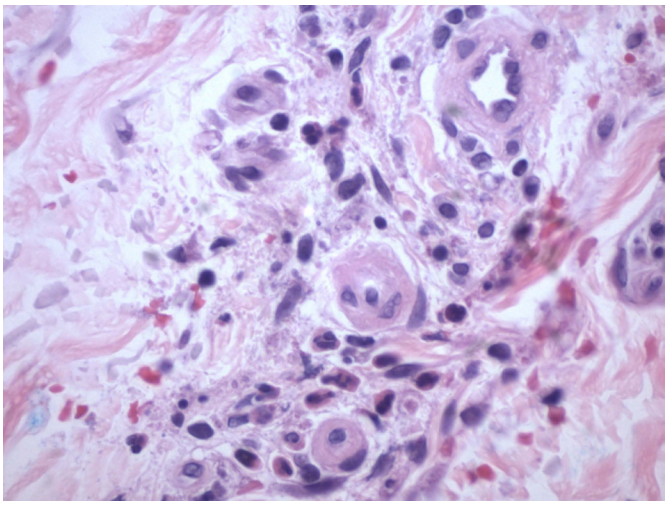

Figure 2a

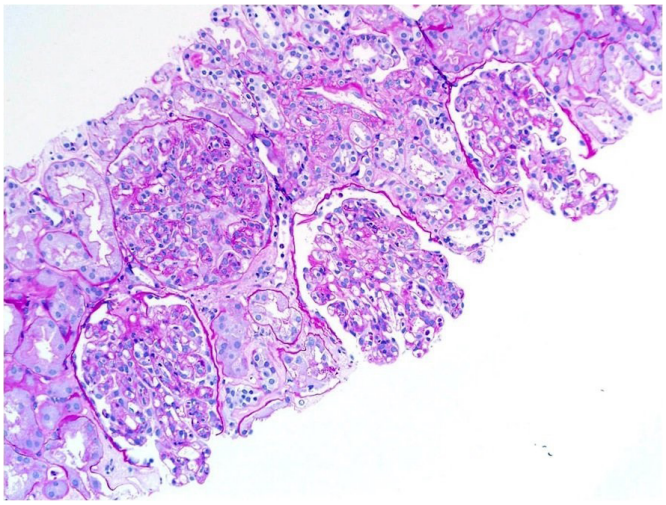

Figure 2b

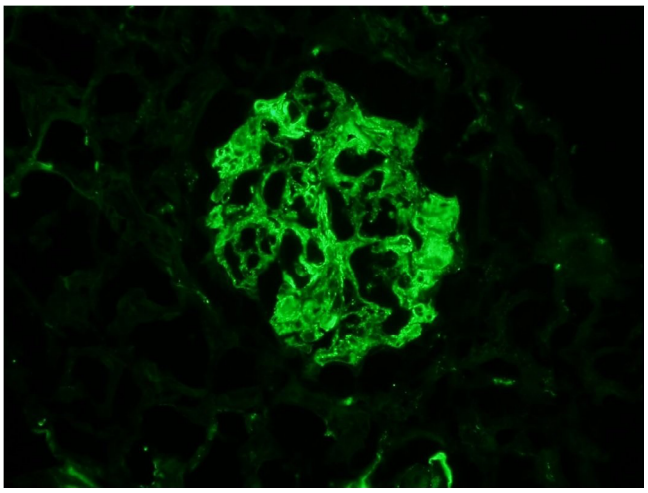

Figure 2c

**Figure 2** **a.** skin biopsy, showing leukocytoclastic vasculitis; **b.** the glomeruli reveal both mesangial and endocapillary hypercellularity with predominant neutrophils (PAS x 200). There is mild tubular atrophy and interstitial fibrosis (involving 25% of renal cortex); **c.** direct immunofluorescence microscopy with fluorescein anti-IgA labelling showed mesangial IgA deposit.

teinuria being the most significant prognostic factor.[9,10] The definitive diagnosis requires a biopsy of the skin and/or kidney. Pathognomonic features include leukocytoclastic vasculitis and IgA deposits in the dermal capillaries and IgA deposits in the mesangium.[11]

The use of glucocorticoids is thought to be beneficial for resolving the arthralgias and abdominal pain of HSP. High dose methylprednisolone may be beneficial in patients with advanced renal disease (crescentic nephritis).[12] Alternative therapies include azathioprine, high-dose IV immunoglobulin, mycophenolate mofetil, cyclophosphamide, and thalidomide.[13,14] There may be a role for plasmaphoresis in the management of HSP.[15] However, there have been no randomized clinical trials to document the efficacy of any of these therapeutic regimens.

In summary, adult onset HSP is a rare disease, often missed at initial presentation. An aggressive diagnostic approach should be employed promptly in such patients to make a timely diagnosis to achieve good clinical outcome. Early initiation of high dose steroids may prevent permanent kidney damage in patients like the present case.

**Author Affiliation:** All authors except Anand Reddy work in the Department of Internal Medicine at Texas Tech University Health Sciences Center in Odessa, TX. Jay Patel and Imran Umer are residents, Yasir Ahmed is an infectious disease specialist, and Craig Spellman is an endocrinologist. Anand Reddy works as clinical nephrologist at TTUHSC in Odessa, TX.
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