

Henoch-Schönlein Purpura in an Adult Filipino Man: A Case Report and Literature Review

LT Damon M. McClain, MC, USNR; LCDR Kimberly Maino, MC, USNR; CAPT Terrence X. Dwyer, MC, USN

GOAL

To gain a better understanding of Henoch-Schönlein purpura (HSP) to better manage patients with the condition

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Describe the presentation of HSP.
2. Identify the antecedent infections of HSP.
3. Discuss any necessary treatments for HSP.

CME Test on page 228.

This article has been peer reviewed and approved by Michael Fisher, MD, Professor of Medicine, Albert Einstein College of Medicine. Review date: March 2006.

This activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education through the joint sponsorship of Albert Einstein College of Medicine and Quadrant HealthCom, Inc. Albert

Einstein College of Medicine is accredited by the ACCME to provide continuing medical education for physicians.

Albert Einstein College of Medicine designates this educational activity for a maximum of 1 AMA PRA Category 1 Credit™. Physicians should only claim credit commensurate with the extent of their participation in the activity.

This activity has been planned and produced in accordance with ACCME Essentials.

Drs. McClain, Maino, and Dwyer report no conflict of interest. The authors report no discussion of off-label use. Dr. Fisher reports no conflict of interest.

Henoch-Schönlein purpura (HSP) is a form of cutaneous small vessel vasculitis that can involve visceral organs and is associated with deposition of immunoglobulin A (IgA)-containing immune complexes. HSP may appear after a

remote history of infection (often an upper respiratory tract infection) as a rash with palpable petechiae or purpura. In addition, a patient with HSP usually complains of arthralgia and abdominal pain. Renal involvement also is common. HSP may be confused with other systemic autoimmune diseases because they all can present with similar symptoms. Prognosis is good and recovery usually occurs without treatment. Although HSP predominately affects children, the condition also can be seen in adults. We present a case of adult-onset HSP in an otherwise healthy Filipino man and review the literature.

Cutis. 2006;77:236-240.

Accepted for publication March 31, 2005.

Dr. McClain is Head of the Medical Department, USS Oak Hill, Little Creek, Virginia. Dr. Maino is Associate Program Director, Department of Dermatology, and Dr. Dwyer is Program Director, Department of Rheumatology, both at National Naval Medical Center, Bethesda, Maryland.

Reprints: LCDR Kimberly Maino, MC, USNR, Department of Dermatology, National Naval Medical Center, 8901 Wisconsin Ave, Bethesda, MD 20889 (e-mail: klmaino@bethesda.med.navy.mil).

Henoch-Schönlein purpura (HSP) is a systemic leukocytoclastic vasculitis often noted as a skin rash with palpable petechiae or purpura. The rash of HSP usually is accompanied by arthralgia, abdominal pain, and renal disease.¹⁻³ HSP affects children at a rate of 15 cases/100,000 individuals annually⁴; the disease is believed to occur less frequently in adults, in whom the natural course of the disease is not as well understood. In adults, HSP can be confused with other autoimmune diseases such as systemic lupus erythematosus and hypersensitivity vasculitis. We present a case of adult-onset HSP and review the literature.

Case Report

A 43-year-old Filipino man presented with a 2-day history of eruptions on his legs (Figure 1) and right arm that were mildly pruritic and burned. The lesions had been preceded by 2 weeks of severe arthralgia and myalgia. The patient noted he had experienced some low-grade fevers, and he complained of mild upper abdominal pain.

A few months prior to presentation, the patient had undergone a diagnostic workup by gastroenterology for an evaluation of abdominal pain and bright red blood from the rectum. Results of stool sampling noted the presence of white blood cells. He was evaluated with a barium enema; colonoscopy was not performed. He was diagnosed with acute gastroenteritis and prescribed levofloxacin for 7 days. During a visit to his primary care physician a few weeks prior to presentation, the patient had complained of significant arthralgia. The patient denied upper or lower respiratory tract symptoms. He also denied blood in his urine.

The patient's medical history included migraines and a glucose-6-phosphate dehydrogenase deficiency. The only medication he took was sumatriptan for his migraines. The patient had no known allergies. He denied taking vitamins or herbal supplements and had no travel history or sick contacts.

Results of a physical examination showed the patient had significant difficulty ambulating but otherwise was in no acute distress. His vital signs were within reference range. Multiple, nonblanching, erythematous papules and pustules were noted on both of the patient's legs and on his right arm. No truncal lesions were noted.

Two 4-mm punch biopsy specimens were taken from the patient's left leg. Results of the histopathologic evaluation showed leukocytoclastic vasculitis (Figure 2). Results of direct immunofluorescence assays revealed granular staining (3+) of the superficial blood vessels focally for immunoglobulin A (IgA)(Figure 3). Results of a wound culture were



Figure 1. Palpable petechiae and purpura in the lower extremity of a patient with Henoch-Schönlein purpura.

negative. Laboratory examination results are listed in the Table. Results of a urinalysis revealed no protein, casts, or blood. Test results were negative for rheumatoid factor, hepatitis B and C, purified protein derivatives, extractable nuclear antigen antibodies, rapid plasma reagins, human immunodeficiency virus, cryoglobulins, antineutrophil cytoplasmic antibodies, and antinuclear antibodies; results of a throat culture were negative for β -hemolytic streptococcus. The results of a chest x-ray did not reveal abnormalities. The patient was diagnosed with HSP.

The patient was evaluated by the departments of internal medicine, rheumatology, nephrology, and gastroenterology. The patient initially was prescribed a therapeutic regimen of bed rest, acetaminophen, ranitidine, and diphenhydramine. He continued to have recurrent skin eruptions and developed more significant arthralgia and abdominal pain. The patient was given one dose of intramuscular methylprednisolone and oral prednisone. He continued to have occasional skin lesions and

mild arthralgias for a few months but currently is clear and has no residual findings.

Comment

HSP predominantly affects people younger than 20 years, and half of all affected children are younger than 5 years.¹ The tetrad of rash, arthralgia, abdominal pain, and renal disease can occur in any order over any time period,^{1,3} and all 4 symptoms do not have to be present clinically. The rash can occur anywhere on the body but usually appears symmetrically on the lower extremities as red to purple lesions. The rash typically presents with palpable petechiae or purpura but has been noted to occur with pustules, vesicles, and bullae. Additionally, the rash can be urticarial, and the Köbner phenomenon can be present. HSP can be distinguished from other autoimmune processes by skin or kidney biopsy results that show IgA deposition. If a biopsy of a skin specimen is performed, immune complexes are found almost equally in the normal-appearing skin and in the lesions of the rash.⁵ Serum complement levels also can be elevated in patients with HSP. Kawana and Nishiyama⁶ noted that increased levels of serum C5b-9 could be correlated with increased activity of HSP; however, C3, C4, and CH50 levels should not be monitored because they were not shown to be elevated consistently in HSP and thus were not reliable.⁶

Gastrointestinal tract lesions have been seen when there is no evidence of characteristic skin findings⁷⁻¹⁰; the lesions usually are present in the stomach, descending duodenum, and colon.^{7,11,12} The lesions can be raised, erythematous, or intermittently spaced.¹⁰ In several reports, occult bleeding was seen in 50% of the patients with HSP, and 25% of the patients had visible bleeding.^{7,9,11-13} Endoscopy can be recommended for individuals with suspected HSP because the test may reveal gastrointestinal tract findings that appear without the classic rash.^{7,9}

Renal involvement is common in patients with HSP. Hematuria with or without proteinuria is present in 30% to 70% of patients with HSP. Levy et al¹⁴ estimated that HSP nephritis is responsible for 15% of all glomerulopathies in children. A retrospective study by García-Porrúa et al¹⁵ noted that hematuria at the onset of HSP with persistence of the renal manifestations during the course of the disease, as well as anemia at disease onset (hemoglobin level <11 g/dL), onset in summer, and relapse, were positive useful predictors for renal sequelae in the adult patients. If a renal biopsy is performed, the percentage of glomeruli showing crescent formation can be correlated with outcome as well. It has been

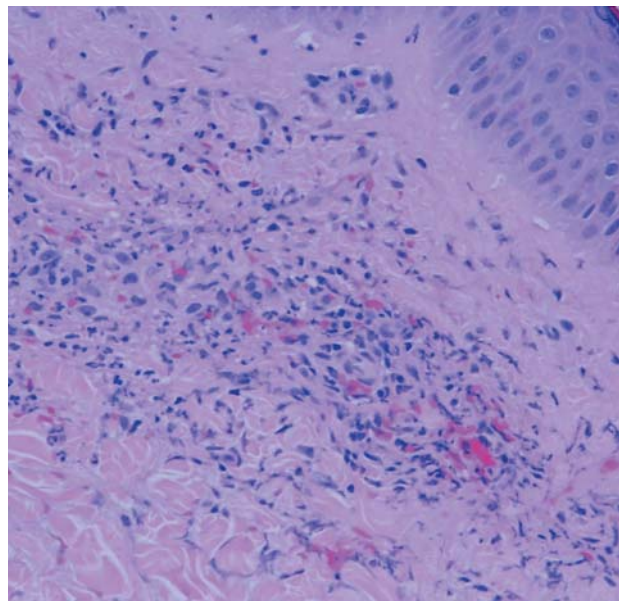


Figure 2. Leukocytoclastic vasculitis (H&E, original magnification $\times 20$).

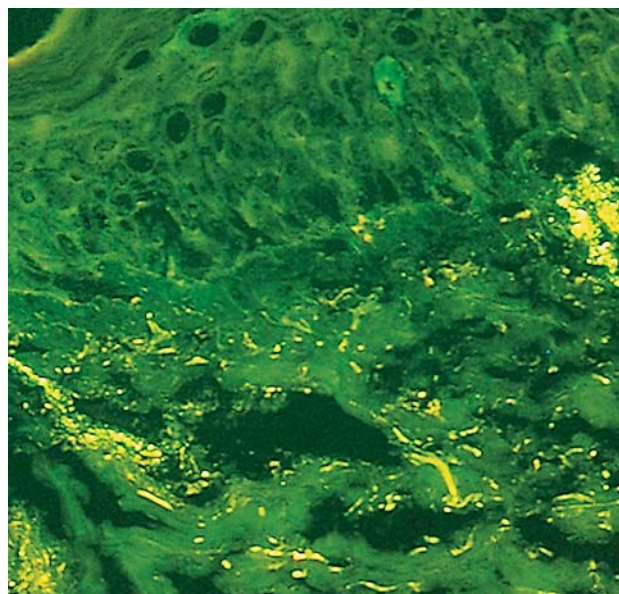


Figure 3. Positive immunoglobulin A around blood vessels (direct immunofluorescence assay, original magnification $\times 20$).

noted that patients with crescents in more than half of the glomeruli are more likely to progress to end-stage or chronic renal disease.¹⁶

The erythrocyte sedimentation rate also can be elevated in patients with HSP. Blanco et al¹⁷ believed that when comparing adults with children, an elevated erythrocyte sedimentation rate and joint pain more often were found in the adult population, whereas the children had a greater

Laboratory Test Results

Component	Patient's Level (Reference Range)
White blood cell count, k/ μ L	9.6 (4–11)
Neutrophils, %	80 (51–67)
Hemoglobin, g/dL	13.7 (14–18)
Hematocrit, %	40.3 (42–52)
Platelet count, k/ μ L	285 (150–450)
Sodium, mmol/L	139 (137–145)
Potassium, mmol/L	4.5 (3.6–5.0)
Chloride, mmol/L	103 (98–109)
Bicarbonate, mmol/L	27 (22–31)
Blood urea nitrogen, mg/dL	15 (9–21)
Serum creatinine, mg/dL	1 (0.8–1.5)
Glucose, mg/dL	123 (76–110)
Repeat fasting glucose, mg/dL	90 (76–110)
Aspartate aminotransferase, U/L	183 (17–49)
Alanine aminotransferase, U/L	203 (7–56)
Alkaline phosphatase, U/L	103 (36–126)
Albumin, g/dL	3.7 (3.5–5.5)
Total bilirubin, mg/dL	0.2 (0.2–1.3)
Creatine kinase, U/L	1411 (55–170)
C3 complement, mg/dL	199 (79–152)
C4 complement, mg/dL	35 (16–35)
Erythrocyte sedimentation rate, mm/h	28 (0–15)

occurrence of abdominal pain, fever, and a preceding upper respiratory tract infection, usually with group A streptococci.

As mentioned previously, HSP easily can be confused with other autoimmune diseases. Thus, the condition should be confirmed with a biopsy specimen from the affected area, which would show leukocytoclastic vasculitis and IgA deposition in the skin or kidney under immunofluorescence

microscopy. In many cases, patients with HSP have a history of infection that precedes the symptoms by a few days to 2 weeks. These antecedent infections include streptococcal pharyngitis, viral pharyngitis, sinusitis, urinary tract infection, bronchitis, and diverticulitis.¹⁸ Hepatitis B also has been implicated as a cause of HSP.^{19–21}

In a retrospective study comparing the clinical course of patients with HSP who were older than 20 years (adults) with those who were younger than 20 years (children), it was noted that the adults had renal involvement more frequently than the children, with 85% of the adults having a nephropathy versus 25% of the children.¹⁷ Of those patients with a nephropathy, the adults developed nephrotic syndrome more often than did the children (13% vs 3%, respectively). Additionally, 13% of the adults with HSP subsequently developed renal insufficiency; however, none of the children with HSP developed this complication. Corticosteroids were given for unresponsive lesions or nephritic involvement. Therapy was required by 63% of the adults and approximately 40% of the children, but 22% of the adults and none of the children needed cytotoxic drugs.¹⁷

The overall outcome is favorable in most patients diagnosed with HSP; one study showed a complete recovery in 89% of the adults and 94% of the children with this condition.¹⁷ Significant morbidity, however, was seen in a retrospective study of 250 patients aged 15 to 86 years who had enough renal involvement from HSP to require a biopsy. After 15 years, 11% of the patients were dependent on dialysis, and 13% had severe renal failure.²² In rare cases, patients with adult-onset HSP had fatal complications because of arrhythmia from vasculitis, which affected the right atrium, and because of massive pulmonary hemorrhage secondary to extensive petechial bleeding, which was refractory to intravenous steroids.²³ Recurrences of HSP tend to be less severe than the original outbreak and usually happen within 4 months of the original symptoms. In children, HSP recurrences have been noted in approximately 33% of patients^{3,14} and seem to be more likely in the subset of patients with nephritis.³

Most patients do not receive therapy; however, those patients with a higher percentage of crescent formation in the kidneys usually require treatment. Renal transplantation also is an option for patients with HSP with end-stage renal disease, but recurrence is seen after 5 years posttransplantation in 35% of patients.²⁴

In summary, our patient was diagnosed with HSP based on the results of a biopsy, direct immunofluorescence, and laboratory workup. His gastrointestinal

tract symptoms are resolved, and he continues to have normal urinalysis results and blood chemistries levels within reference range. He occasionally notes a few new skin lesions and has episodes of mild arthralgia. The patient will continue to be followed up closely to ensure that he does not develop renal abnormalities.

REFERENCES

1. Meadow SR, Scott DG. Berger disease: Henoch-Schönlein syndrome without a rash. *J Pediatr*. 1985;106:27-32.
2. Cameron JS. Henoch-Schönlein purpura: clinical presentation. *Contrib Nephrol*. 1984;40:246-249.
3. Saulsbury FT. Henoch-Schönlein purpura in children. report of 100 patients and review of the literature. *Medicine (Baltimore)*. 1999;78:395-409.
4. Nielsen HE. Epidemiology of Schönlein-Henoch purpura. *Acta Paediatr Scand*. 1988;77:125-131.
5. Van Hale NM, Gibson LE, Schroeter AL. Henoch-Schönlein vasculitis: direct immunofluorescence study of uninvolved skin. *J Am Acad Dermatol*. 1986;15:665-670.
6. Kawana S, Nishiyama S. Serum SC5b-9 (terminal complement complex) level, a sensitive indicator of disease activity in patients with Henoch-Schönlein purpura. *Dermatology*. 1992;184:171-176.
7. Wu CS, Tung SY. Henoch-Schönlein purpura complicated by upper gastrointestinal bleeding with an unusual endoscopic picture. *J Clin Gastroenterol*. 1994;19:128-131.
8. Nathan K, Gunasekaran TS, Berman JH. Recurrent gastrointestinal Henoch-Schönlein purpura. *J Clin Gastroenterol*. 1999;29:86-89.
9. Gunasekaran TS, Berman J, Gonzalaz M. Duodenojejunitis: is it idiopathic or is it Henoch-Schönlein purpura without the purpura? *J Pediatr Gastroenterol Nutr*. 2000;30:22-28.
10. Gunasekaran TS. Henoch-Schönlein purpura: what does the "rash" look like in the gastrointestinal mucosa? *Pediatr Dermatol*. 1997;14:437-440.
11. Nakasone H, Hokama A, Fukuchi J, et al. Colonoscopic findings in an adult patient with Henoch-Schönlein purpura. *Gastrointest Endosc*. 2000;52:392.
12. Yoshikawa N, Yamamura F, Akita Y, et al. Gastrointestinal lesions in an adult patient with Henoch-Schönlein purpura. *Hepatogastroenterology*. 1999;46:2823-2824.
13. Cappell MS, Gupta AM. Colonic lesions associated with Henoch-Schönlein purpura. *Am J Gastroenterol*. 1990;85:1186-1188.
14. Levy M, Broyer M, Arsan A, et al. Anaphylactoid purpura nephritis in childhood: natural history and immunopathology. *Adv Nephrol Necker Hosp*. 1976;6:183-228.
15. García-Porrúa C, González-Louzao C, Llorca J, et al. Predictive factors for renal sequelae in adults with Henoch-Schönlein purpura. *J Rheumatol*. 2001;28:1019-1024.
16. Habib R, Niaudet P, Levy M. Schönlein-Henoch purpura nephritis and IgA nephropathy. In: Tisher CC, Brenner BM, eds. *Renal Pathology With Clinical and Functional Correlations*. Philadelphia, Pa: Lippincott; 1994:472-523.
17. Blanco R, Martinez-Taboada VM, Rodriguez-Valverde V, et al. Henoch-Schönlein purpura in adulthood and childhood: two different expressions of the same syndrome. *Arthritis Rheum*. 1997;40:859-864.
18. Magro CM, Crowson AN. A clinical and histologic study of 37 cases of immunoglobulin A-associated vasculitis. *Am J Dermatopathol*. 1999;21:234-240.
19. Chave T, Neal C, Camp R. Henoch-Schönlein purpura following hepatitis B vaccination. *J Dermatolog Treat*. 2003;14:179-181.
20. Braun S, Ramaker J, Dippel E, et al. Schönlein-Henoch purpura associated with hepatitis B in a patient with HIV infection [in German]. *Dtsch Med Wochenschr*. 2001;126:103-107.
21. Singh PS. Henoch schönlein purpura in adults associated with hepatitis B antigen [letter]. *J Assoc Physicians India*. 1995;43:73.
22. Pillebout E, Thervet E, Hill G, et al. Henoch-Schönlein purpura in adults: outcome and prognostic factors. *J Am Soc Nephrol*. 2002;13:1271-1278.
23. McCarthy R, Rosen T, Chen SH, et al. Adult Henoch-Schönlein purpura with fatal complications. *Arch Dermatol*. 2001;137:19-21.
24. Meulders Q, Pirson Y, Cosyns JP, et al. Course of Henoch-Schönlein nephritis after renal transplantation. report on ten patients and review of the literature. *Transplantation*. 1994;58:1179-1186.

DISCLAIMER

The opinions expressed herein are those of the authors and do not necessarily represent the views of the sponsor or its publisher. Please review complete prescribing information of specific drugs or combination of drugs, including indications, contraindications, warnings, and adverse effects before administering pharmacologic therapy to patients.

CONFLICT OF INTEREST STATEMENT

The Conflict of Interest Disclosure Policy of Albert Einstein College of Medicine requires that authors participating in any CME activity disclose to the audience any relationship(s) with a pharmaceutical or equipment company. Any author whose disclosed relationships prove to create a conflict of interest, with regard to their contribution to the activity, will not be permitted to present.

The Albert Einstein College of Medicine also requires that faculty participating in any CME activity disclose to the audience when discussing any unlabeled or investigational use of any commercial product, or device, not yet approved for use in the United States.