A Case of Henoch-Schonlein Purpura in an Adult

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BACKGROUND

Henoch-Schonlein Purpura (HSP), or IgA Vasculitis, is characterized by the clinical tetrad of palpable purpura, arthralgia/arthritis, abdominal pain, and hematuria.

The majority of cases, approximately 90%, are diagnosed in children. There is a male predominance and an increase in number of cases during the winter season. The incidence of HSP in children is approximately 20 cases per 100,000. It is less common in adults with textbooks reporting an incidence of 14 cases per million.

Triggers of HSP include medications, infections, malignancies, inflammatory disorders, and idiopathic causes. The most common triggers in children are infections, particularly viral infections, streptococcal pharyngitis, and mycoplasma. In adults, medications are the most common trigger. Virtually every class of medication can induce HSP, and the time from introduction to reaction may vary from hours to years. Therefore, it may be difficult to determine which drug is responsible.

In order to diagnose HSP the patient must demonstrate palpable purpura plus one of the following: arthritis/arthralgia, diffuse abdominal pain, a biopsy demonstrating IgA deposition or renal involvement manifested as hematuria or proteinuria.

CASE PRESENTATION

A 59-year-old previously healthy Caucasian woman presented with a one-month history of a bilateral progressive lower extremity rash associated with polyarthralgia and dependent edema (Figures 1-3).

Over the past month, she had been seen at two urgent care centers and two emergency rooms and each time was diagnosed with cellulitis and discharged on oral antibiotics and a prednisone taper. Her symptoms would improve slightly but then would flare once her prednisone taper was completed.

Review of systems was positive for arthralgia in the bilateral knees and ankles, fatigue, and burning pain in the area around the lesions. Pertinent negatives included lack of abdominal pain or melena.

Her vitals were stable except for her systolic blood pressure which was persistently elevated in the 170s to 190s. A CBC was notable for a leukocytosis of 18.500. A CMP was significant for an elevated creatinine of 1.2. A subsequent urinalysis revealed moderate hematuria and proteinuria.

Work-ups for systemic lupus erythematosus, immunofixation with electrophoresis, total serum immunoglobulins, and beta 2 microglobulin were negative. Further studies of medium vessel vasculitides and viral serology (including EBV, VZV, HIV, and a hepatitis panel) were all negative. Lower extremity wound cultures were positive for E. coli and S. aureus. Ultrasound of the bilateral lower extremities was unremarkable. Magnetic resonance imaging demonstrated diffuse edema and enhancement of the soft tissue consistent with cellulitis.

CLINICAL IMAGES

Figure 1

Figure 2

Figure 3

PATHOLOGY

Figure 4 & 5: H&E examination revealed dense neutrophilic infiltration with RBC extravasation and complete obliteration of blood vessels consistent with leukocytoclastic vasculitis.

Figure 6: Direct Immunofluorescence (DIF) findings with immunoglobulin A (IgA) in the skin in an adult with Henoch-Schonlein purpura. The superficial dermis shows strong blood vessel fluorescence with IgA.

DISCUSSION

Pathogenic: HSP is due to abnormal IgA deposition in vessel walls. Abnormal IgA glycosylation leads to the formation of IgG-IgA1 immune complexes that lodge in vessel walls and activate the alternate complement pathway. Complement activation induces neutrophil chemotaxis with a resulting release of proteolytic enzymes that cause endothelial injury. The post capillary venules of the dermis undergo intense neutrophilic vascular inflammation. The resulting inflammation and extravasation of red blood cells can be appreciated clinically as palpable purpura.

Clinical Findings: It is important to evaluate patients with palpable purpura for joint involvement, gastrointestinal involvement, and renal involvement. Up to 75% of adult patients with HSP will have joint involvement that typically affects the knees and ankles. The vasculitis that occurs in HSP may affect the vessels in the GI tract resulting in bowel ischemia and edema causing the patient to feel severe abdominal pain.

Renal involvement is seen in 40-75% patients with HSP and usually presents as proteinuria, hematuria or new onset hypertension. Virtually all patients with palpable purpura above the waist will have renal involvement. In addition, up to 30% of adults will develop chronic renal insufficiency, especially if the palpable purpura occurs above the waist or if there is an elevated erythrocyte sedimentation rate.

Diagnosis: Punch biopsies were performed and submitted for histopathological and direct immunofluorescence (DIF) examination. Hematoxylin & eosin (H&E) staining of the specimens revealed dense neutrophilic infiltrate with red blood cell extravasation, hemorrhage, and complete obliteration of the blood vessels consistent with leukocytoclastic vasculitis (Figures 4 & 5). DIF revealed perivascular IgA, C3, and fibrin deposits consistent with IgA Vasculitis (Figure 6).

These biopsy results in the setting of the clinical findings of palpable purpura, arthralgia, proteinuria, hematuria, and hypertension confirmed the diagnosis of adult Henoch-Schonlein Purpura.

TREATMENT & MANAGEMENT

Due to severity of disease, renal involvement and joint involvement the following therapy was initiated:

• Solumedrol pulse therapy: 500 mg IV daily infusion x 3 days
• Mycophenolate Mofetil 1,500 mg twice daily
• Prednisone 80 mg qAM

The patient was instructed to follow up with a dermatologist, nephrologist, rheumatologist, and internist at an academic center so that her care could be more easily managed between the different specialties.

REFERENCES


