

## CLINICAL VIGNETTE

# Henoch-Schönlein Purpura

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A 61-year-old female presents with pains and bruises in her legs for 2-3 weeks. She assumed that the pains were related to her chronic back problems, but the symptoms did not improve with NSAID's and rest. The pains are mostly localized in her thighs and calves but also involve her knees. She denies any injuries or trauma. She reports a recent upper respiratory infection and was prescribed antibiotics by a doctor at an urgent care center near her home. Her past medical history includes melanoma, hypertension, hepatitis B, osteopenia, thyroid cysts, spinal stenosis, degenerative disc disease, and osteoarthritis. Past surgical history includes total abdominal hysterectomy with BSO in 1990.

Her medications include Losartan 25 mg daily and Brimonidine eye drops. She also takes calcium, magnesium, and vitamin D supplements. Recently, she began to take various herbal supplements that she read about in a magazine. She doesn't recall the names of these herbs. She's allergic to penicillin, hydrocodone, codeine, and doxycycline.

**Social History:** She attends a Pilates class several times a week. She swims about once a week. She has had more difficulty exercising recently because of the back and leg problems. She has never smoked. She drinks socially. She drinks green tea each morning. She has been a vegetarian for 37 years consuming no fish, meat, nor chicken.

**Family History:** Father died at 60 in a car accident. He had hypertension. Mother died at 61 with lung cancer. She was a smoker. The patient has one sister aged 54 who has melanoma.

**Physical Examination:** Blood pressure is 116/78, pulse 64, temperature 36.6, height 5 feet 7, and weight 152.2 pounds. Body fat is 37.2%. BMI is 23.8. The patient is comfortable in no apparent respiratory distress.

**Head and neck exam:** JVP is normal. Carotids are without bruits. Conjunctivae pink. Sclerae nonicteric. Oropharynx is unremarkable. Ear canals are negative bilaterally. Carotids are without bruits. JVP is normal. Head is atraumatic and normocephalic. Lungs: Clear to auscultation and percussion. Neck: Supple and nontender. Cardiac: Regular rate and rhythm. No murmurs or gallops. PMI is normal. Abdomen: Nontender and nondistended with good bowel sounds. No masses or hepatosplenomegaly. Breast exam shows no abnormalities to inspection and palpation. Skin: Shows palpable purpura in both legs. No edema. Neurologic: Alert and oriented x2. Cranial nerves are intact II-XII. Motor and sensory exams are normal. Reflexes are grossly normal in all 4 extremities. I could not reproduce her leg symptoms today and her back is benign.

Her labs revealed a mildly elevated PT/INR and PTT. Her white count is 10.8 with a left shift. Chemistries are normal. TSH is normal. UA showed microscopic hematuria.

### *General Discussion and Epidemiology*

Henoch-Schönlein purpura (HSP) is an immune-mediated systemic small vessel vasculitis that can affect the skin, the gastrointestinal tract, kidneys, joints, as well as other organs.<sup>1</sup> HSP is an IgA-mediated vasculitis and usually presented as an acute, self-limited disorder that doesn't recur.<sup>1</sup> The prevalence of HSP is approximately 15-20 cases per 100,000 although this varies widely by country.<sup>2</sup> In some countries, HSP is a very common cause of glomerulonephritis and affects males slightly more often than females especially in children.<sup>2</sup> It most commonly presents prior to the age of ten years with a peak incidence at age five but can occur in adults of any age.<sup>2</sup> HSP tends to occur during the winter months and is rarely seen before the age of two years.<sup>1</sup> An older age of presentation is associated with a greater risk of chronic renal disease, proteinuria, and hypertension.<sup>3</sup> Abdominal pain tends to be more common in those who present at a younger age.<sup>4</sup> Children tend to present with elevated inflammatory markers such as CRP, leukocytosis, and thrombocytosis, while adults tend to present with elevated cyroglobulin levels.<sup>5</sup> Familial clusters of HSP have also been identified.<sup>6,7</sup>

### *Etiology and Pathophysiology*

The cause of HSP has not been elucidated, and it is believed to be a multifactorial disorder possibly related to various types of infections or toxins.<sup>8</sup> Infections that have been linked to HSP include Salmonella, Shigella, Legionella, Brucellosis, Hepatitis B and C, Yersinia, H. Pylori, Epstein-Barr Virus, Cholera, and Adenovirus.<sup>8</sup> Various medications linked to HSP include Erythromycin, Penicillin, Losartan, and Quinidine.<sup>8</sup> IgA is an important factor in the pathophysiology of HSP especially the IgA1 subtype.<sup>8</sup> Interleukins also appear to play a role include tumor necrosis factor, transforming growth factor, IL-1, and IL-6.<sup>9</sup> Genetics appears to play a minor role.<sup>10</sup> The use of TNF-blockers may increase the risk of HSP in certain patients.<sup>10</sup> HSP appears to be related to IgA nephropathy although the relationship between these two disorders hasn't been fully elucidated.<sup>9</sup>

### *Clinical Features*

The most common clinical manifestation in HSP are dermatologic, gastrointestinal, rheumatologic, and renal.<sup>10</sup> Specific signs and symptoms can include palpable purpura, GI

bleeding, glomerulonephritis, and abdominal pain.<sup>9</sup> Patients often present with nonspecific constitutional symptoms of fever, malaise, myalgias, headache, cough, and chills.<sup>8</sup> Besides purpura, dermatologic findings can include urticarial and macular rashes, as well as angioedema and erythema multiforme-type reactions.<sup>10</sup> The rash usually appears in waves and typical lasts for 2-4 weeks.<sup>8</sup> The rash most commonly affects the buttocks, upper thighs, feet, ankles, and lower legs.<sup>8</sup> Gastrointestinal features can include diarrhea, pancreatitis, melena, duodenal ulcers, appendicitis, bowel infarction, and ileal bleeding.<sup>11</sup> Renal involvement can present in many different ways including acute glomerular inflammation, urethritis, mesangial hypercellularity, endocapillary proliferation, and leukocyte infiltration.<sup>9</sup> Rheumatologic involvement can affect the knees, wrists, and ankles.<sup>12</sup> Scrotal involvement is also quite common.<sup>12</sup> HSP can also be associated with neurologic and psychological issues including seizures, neuropathy, mood changes, ADHD, and altered mental status.<sup>12</sup>

### Diagnosis and Testing

There is no diagnostic test with high sensitivity and specificity for HSP.<sup>12</sup> Tests are often performed to rule out other disorders.<sup>12</sup> These can include ANA, Rheumatoid factor, ESR, CBC, urinalysis, CH50, C3, C4, basic chemistries, factor 8, factor 9, IgA, IgG, PT, PTT, D-dimer, amylase, and lipase.<sup>13</sup> Imaging may include ultrasound (abdomen and scrotum), chest CT, abdomen CT, renal biopsy, skin biopsy, EGD, and colonoscopy.<sup>13</sup> Disorders that may need to be ruled out include medication reaction, bowel infarction, essential mixed cryoglobulinemia, hypersensitivity vasculitis, leukocytoclastic vasculitis, Rickettsial diseases, Waldenström macroglobulinemia, and Wegner's granulomatosis.<sup>12</sup>

### Treatment

The treatment of HSP is primarily supportive in most cases.<sup>13</sup> These supportive measures can include hydration, electrolyte monitoring, nutritional monitoring, adjustment of medications, and symptom management.<sup>13</sup> Steroids can be helpful in certain cases such as nephrotic syndrome, glomerulonephritis with crescents, scrotal edema, GI bleeding, and pulmonary hemorrhage.<sup>13</sup> Azathioprine, Cyclophosphamide, or Cyclosporine may also be used in these cases.<sup>13</sup> Plasmapheresis may also be considered especially if severe renal disease is present.<sup>14</sup>

### Prognosis

Patients with HSP generally follow a benign course and have a very good prognosis. Most patients have a complete resolution of symptoms within several months, and only 3-5% of patients have residual problems.<sup>13</sup> Relapses can sometimes occur and can last for several months.<sup>13</sup> Patients that are likely to have long-term sequelae are those with severe renal disease, GI bleeding, hematuria, proteinuria, neurologic complications, or persistent rashes.<sup>13</sup>

### Clinical Course and Follow-Up

The patient was managed conservatively with NSAID's, hydration, and skin care. Over the course of 6-8 weeks, she gradually improved and eventually had a complete recovery.

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