CLINICAL VIGNETTE

Henoch-Schönlein Purpura: Presenting with Petechial Rash, Fever and Sore Throat

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Case Report

A 29-year-old woman with polycystic ovarian syndrome presents with a rash, fever and sore throat. The patient reports symptoms started several days prior with a petechial red rash on her bilateral distal lower extremities. The rash was initially flat and painless without pruritus. However, the rash is now deeper purple, painful, raised with a few new blisters. She also notes a sore throat, fever and some mild aching and swelling of her toes and ankles. She has no prior history of tonsillitis, arthritis or allergies. She denies cough, rhinorrhea, abdominal pain, diarrhea, hematochezia or melena, headache, chest pain, shortness of breath, hematuria or dysuria. She is not on any medications.

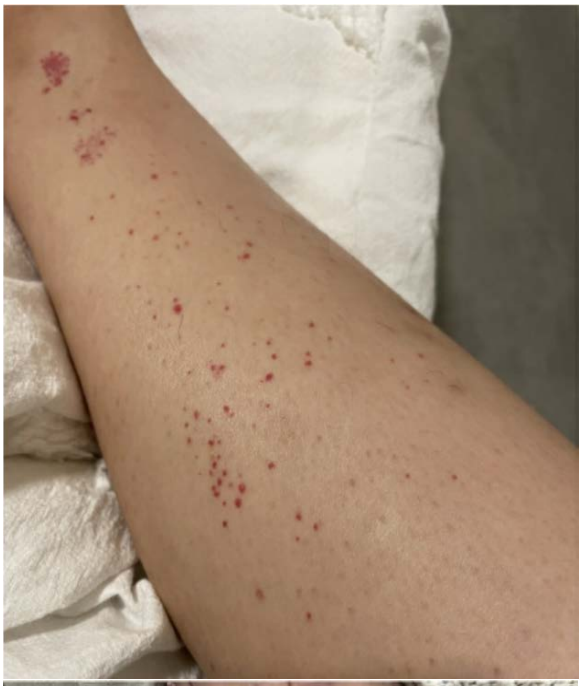
Social history includes recent marriage, actively family planning with no smoking, drug or alcohol use. Her family history includes hepatitis B and liver cancer in her father.

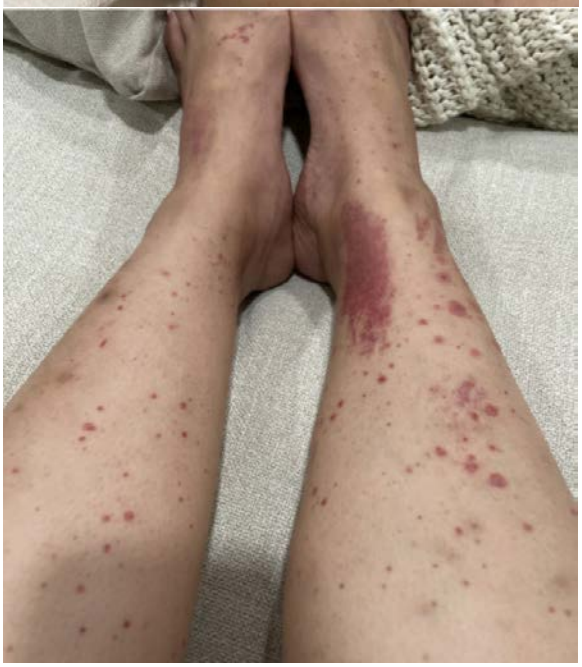
Physical Examination

On examination, she was normotensive, temperature of 37.4 Celsius, heart rate 99 bpm and SpO2 of 99%. She appeared comfortable, oropharyngeal examination, was remarkable for an erythematous, swollen left tonsil with white exudate, and midline uvula. There was no cervical lymphadenopathy and lungs were clear without stridor or rhonchi. Cardiac and abdominal exam were unremarkable. Her extremities were notable for a palpable petechial rash with violaceous plaques on her right ankle. A few scattered vesicles were present, but no bullae or pustules (see photos). She also had mild swelling and tenderness of her 3rd and 4th toes bilaterally, with faint warmth but no induration.

Initial Laboratory Values

White Blood Cell Count 5.9 x 103/uL with a normal differential, Hemoglobin 14.6 g/dL, Hematocrit 45.1%, Platelet 282,000. Sodium 138, Potassium 4, Chloride 102, bicarbonate 22, BUN 8, Creatinine 0.58, Glucose 87. Point of care rapid strep A testing was positive. Infectious monoclonal antibody was negative. Urinalysis was notable for 1+ blood and 1+ protein. Total urine protein/creatinine ratio was elevated at 0.6 mg/dL. SARS-Cov-2 PCR testing was negative.





Treatment Course

The patient was treated with amoxicillin 500mg by mouth three times a day for a 10-day course, and her sore throat and fever quickly improved. She was referred to dermatology and nephrology clinics for suspected immunoglobulin A (IgA) vasculitis with concern for renal involvement.

She presented to dermatology clinic 2 weeks later, after completing the 10-day course of amoxicillin. The rash was healing, with only faint hyperpigmented papules and macules on her distal extremities. Skin biopsy was therefore deferred. Nephrology elected to send additional vasculitis laboratory tests including pANCA, cANCA, viral hepatitis serology, ANA, dsDNA, cryocrit, HIV, C3/C4-, with all test returning normal. Kidney ultrasound showed normal kidneys with no detectable kidney stones, hydronephrosis or masses. After several weeks, repeat creatinine remained normal and urinalysis and urine total protein/creatinine ratio had all normalized.

Discussion

Henoch-Schönlein purpura (HSP) is an IgA mediated vasculitis most common in children but also affecting adults. Incidence of HSP is 10 to 22 per 100,000 annually.[1] Clinical manifestations may include purpura, joint pain, and abdominal pain.[2,3] Pediatric cases of HSP are generally mild and self-limiting, particularly in children younger than 2-years-old. On the other hand, adult cases may be more severe due to complications of acute enteritis, gastrointestinal bleeding, and glomerulonephritis.[3,4]

HSP is caused by IgA immune complexes that deposit in small blood vessels including the skin, kidney, and intestinal lining.[2] HSP often is triggered by upper respiratory infections, including Group A streptococcus such as in our case study. Other infections linked to HSP include Coxsackie, adenovirus, hepatitis A and B, haemophilus parainfluenza, parvovirus B19, helicobacter pylori, MRSA, and campylobacter.[5] HSP generally follows an upper respiratory infection within days or weeks, presenting with rash, abdominal pain, arthritis and fever. Interestingly our patient presented with rash and arthralgias at the same time as her pharyngitis.

HSP is a clinical diagnosis as there are no specific diagnostic serologic tests. In 1990, the American College of Rheumatology developed diagnostic criteria for pediatric cases of HSP. They required two out of four criteria be present: palpable purpura without thrombocytopenia, patient 20 years or younger at presentation, bowel angina (diffuse abdominal pain or diagnosis of bowel ischemia), and biopsy showing granulocytes in the walls of the small arterioles or venules.[6] While the ACR criteria may be helpful in pediatric patients, it does not translate to adults. Aside from history and examination, laboratory testing can help confirm HSP. Laboratory testing may include complete metabolic panel, urinalysis, complete blood counts, Antistreptolysin-O titers, INR, PTT and IgA levels. Normal platelets in the setting of palpable purpura should trigger consideration of HSP. Prior strep infections noted with elevated antistreptolycin-O titers, acute kidney injury with or without proteinuria, leukocytosis, elevated IgA levels and bleeding diathesis may also be indicative. Other supportive diagnostic tests may include skin and renal biopsies that show leukocytoclastic vasculitis and membranoproliferative glomerulonephritis.[7] Other vasculitis conditions should also be evaluated while testing for HSP.

Symptomatic treatment is usually the mainstay for HSP, as most cases are self-limiting. NSAIDs should be avoided due to potential renal and GI involvement. There is controversy regarding the use of corticosteroids to prevent renal involvement. Several pediatric studies over the past 15 years have examined whether the early use of corticosteroids prevented renal complications of HSP. However, these trials found no significant benefit of corticosteroids over placebo in preventing renal involvement.[8,9] Patients with minor to no renal involvement should still be monitored for 3-6 months to ensure kidney function remains normal. On the other hand, HSP cases with renal involvement should warrant serious consideration of treatment with steroids or other immunosuppressants. A Cochrane review from 2015 examined various treatment options for kidney involvement in patients with HSP. This review found there may be a role for cyclosporine A, mycophenolate mofetil, dapsone, rituximab and cyclophosphamide. However, study sizes were small and more studies are needed to elucidate the role of these agents in treatment of serious HSP disease.[10]

Conclusion

While generally thought of as a pediatric condition, HSP should be considered in adults presenting with recent strep A or other upper respiratory illness, and concomitant palpable purpura, joint pain and abdominal pain. Primary care physicians should evaluate for renal and gastrointestinal involvement and refer early in the disease process for close monitoring. While HSP usually is self-limited, adults may be more prone to end organ complications, thus early recognition is critical.

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