A Report of Henoch Schonlein Purpura in a Young Adult with Concomitant Lyme Disease

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Abstract
Henoch Schonlein Purpura (HSP), or IgA vasculitis, is characterized by immune complex deposition in small blood vessels and activation of the alternative complement pathway. HSP is typically seen after the resolution of viral illness in children though it has also occurred on rare occasions in adults. In these cases, the rash is usually self-limited. In this article, an uncommon presentation of HSP in a 19-year-old young adult with no evidence of prior upper respiratory infection (URI) and concomitant Lyme disease is presented. The patient had a two-week history of a worsening rash and pain in multiple joints before seeking medical care. The treatment course is notable for resolution of the HSP rash and joint symptoms upon treatment of HSP and underlying Lyme disease.

Introduction

Henoch Schonlein Purpura (HSP), or IgA vasculitis, is characterized by immune complex deposition in small blood vessels and activation of the alternative complement pathway.[1] HSP can have many clinical manifestations including purpura, renal damage, gastrointestinal distress, and arthritis. The rash seen with HSP is a non-blanching, palpable purpura with a normal or elevated platelet count. Lack of reduction in platelet count helps to differentiate HSP from other forms of purpura including immune thrombocytopenic purpura or thrombotic thrombocytopenic purpura. HSP is typically seen after the resolution of viral illness in children under the age of 10 years old and is more common in boys.[2] HSP has also occurred on rare occasions in adults.

HSP is a rare post-infectious sequela in children with a prevalence of 0.002%.[3] The majority of these cases are present in kids aged 2-6 years. There is extensive documentation correlating HSP with numerous viral illnesses including COVID-19,[4] respiratory syncytial virus (RSV), adenovirus, and hepatitis.[5] In children with the disease, purpura is often self-resolving in under a year. However, major complications can occur including involvement of kidneys and joints leading to kidney damage and arthritis. Gastrointestinal complications including abdominal pain, nausea, vomiting, and

hematemesis are also common symptoms. To reduce the risk of complications, steroid therapy can be initiated. In the adult population, HSP occurs incredibly rarely with a prevalence as low as 0.0000034-0.0000143%.[6,7] Adults with purpura and normal thrombocyte levels should be investigated for IgA vasculitis.[8] In this article, a case of HSP in a 19-year-old young adult with no evidence of prior URI and concomitant Lyme disease is presented.

Case Report

History

The patient is a 19-year-old male nonsmoker with no significant medical history. The patient presented with a rash (pictures enclosed) of the lower extremities and severe polyarticular pain for two weeks. The patient reported the lesions beginning on the feet and then quickly progressing to involve both legs, the right upper arm, and the left elbow. He experienced some discomfort in the groin without penile discharge or dysuria. Lesions in the groin region were consistent with those on the lower extremities. He also had significant swelling of the lower limbs. The patient denied having a preceding upper respiratory tract infection. He did not experience any pharyngitis, congestion, rhinorrhea, cough, or headache before the onset of the rash. He also denied hematuria/dysuria and abdominal pain. Pt had no reported tick bites or history of recent outdoor recreational activities.

Physical examination was unremarkable except for the skin and joints. Skin demonstrated palpable purpura beginning on the abdomen and traveling distally. His joints were exquisitely tender.

All vitals were within normal limits and urinalysis showed normal values without erythrocytes, leukocytes, or spillage of protein.

The patient was evaluated for viral illness and autoimmune causes of the rash. He was found to have negative hepatitis serology, a normal rheumatologic panel, and negative results for sexually transmitted diseases (STDs). Incidentally, the patient was found to have positive Lyme titers upon evaluation.

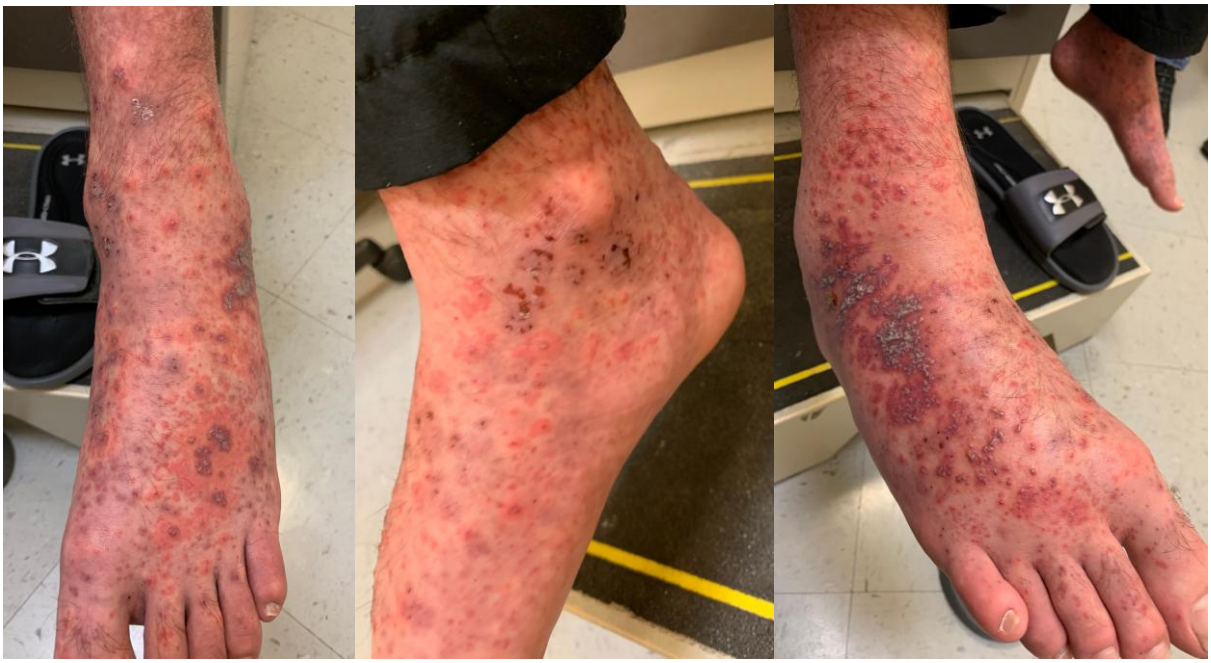

Figure 1-3: Lower extremity skin lesions.

Diagnostic Tests

CBC was normal and platelet count was within normal ranges. The patient received a rheumatologic workup which was significant only for elevated c-reactive protein. Serum ANA, RF, CCP, and ANCA were all unremarkable. Complement C3 & C4 levels were within normal ranges. Screening for STDs was also unremarkable. The patient was screened for hepatitis and results demonstrated no evidence of infection.

The patient was tested for Lyme disease and IgG titers came back positive for 18 KD (IgG), 28 KD (IgG), 30 KD (IgG), 39 KD (IgG), 41 KD (IgG), 45 KD (IgG), 58 KD (IgG), 66 KD (IgG), and 93 KD (IgG). Lyme IgG Western blot is positive if at least five of the ten IgG titers are positive; this patient demonstrated reactivity with nine of ten titers.

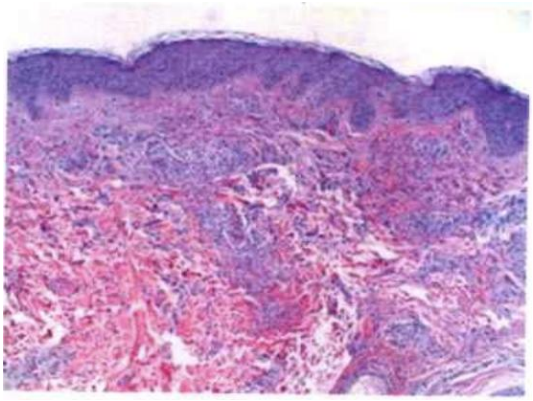

Figure 4: Punch biopsy; skin, right anterior proximal thigh.
The patient had a punch biopsy taken of the right anterior proximal thigh. Histologic findings were suggestive of a small vessel leukocytoclastic vasculitis (LCV) secondary to IgA vasculitis.

Management

The patient was placed on oral prednisone for symptom relief and saw a reduction of symptoms including resolution of purpura, pain, and swelling over the course of a month. The patient experienced rebound symptoms and a recurrence of rash after a night of alcohol intoxication when steroids were tapered down the first time. He was started on another course of steroids after this. When the Lyme titers returned, the patient was started immediately on doxycycline with a near-complete resolution of symptoms including reduced joint pain. The patient had no prior knowledge or treatment for Lyme disease.

The patient did not experience systemic symptoms of HSP and only demonstrated cutaneous signs. He did not have any renal involvement or gastrointestinal illness. The joint pain may be attributed to either HSP or advancement of Lyme disease before treatment.

Discussion

Significance

This patient was an adult male presenting with a rare form of palpable purpura most commonly seen in children. HSP has been documented in very few adults but remains a vital part of the differential for a patient with purpura.

The patient also received a diagnosis of Lyme disease. Most documented cases of HSP follow as a post-infectious sequela after a viral URI, but this patient did not have any preceding URI symptoms. Lyme disease usually manifests on the skin as erythema chronicum migrans but some patients have displayed other concomitant skin findings including Henoch-Schonlein-like purpura.[9] This patient's skin demonstrated clear palpable purpura and biopsy confirmed leukocytoclastic vasculitis.

While there is no direct correlation between Lyme disease and HSP, this patient experienced a complete resolution of rash and joint pain after receiving both steroids for HSP and antibiotic treatment for Lyme disease. Steroids can reduce inflammation and purpura in HSP but recurrence is unchanged by this treatment.[10] Steroids also do not change the likelihood for systemic complications.[11] This may suggest a need to investigate the underlying infection that resulted in autoimmune IgA.

Even though HSP is rare, it's an important differential in an adult with palpable purpura to monitor for systemic complications. HSP often correlates with URI but can also develop after other infections. In this patient, symptomatic treatment of HSP and antibiotic treatment for Lyme disease correlated with improvement of HSP rash. This case is a significant reminder to look for an underlying cause of autoimmune vasculitis to ensure adequate treatment and prevent rebound vasculitides.

The current treatment regimen for HSP involves symptomatic pain relief since the disease is self-resolving.[12] For mild pain in patients without gastrointestinal (GI) bleed or glomerulonephritis, naproxen 10-20 mg/kg split into two daily doses or ibuprofen 10 mg/kg every 6-8 hours is suggested for 3-4 weeks. Acetaminophen 10-15 mg/kg every 4-6 hours is recommended for a patient in mild pain with GI bleed or glomerulonephritis. For more severe pain with adequate oral intake, treatment is oral prednisone 1-2 mg/kg/day for 3-5 days then tapered over 3-4 weeks. For severe pain with limited oral intake, 0.8-1.6 mg/kg/day of IV methylprednisolone is suggested for 3-5 days before tapering down for 3-4 weeks.[13] In all patients, monitoring for abdominal or renal complications of IgA vasculitis is required.

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