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Akram Mustafa, MD

Thomas Jefferson University Hospitals, Akram.Mustafa@jefferson.edu

Edward K. Len, MD

Thomas Jefferson University Hospitals, Edward.Len@jefferson.edu

Alan Chiu, MSIII

*Thomas Jefferson University Hospitals*Follow this and additional works at: <https://jdc.jefferson.edu/tmf> Part of the [Medicine and Health Sciences Commons](#)[Let us know how access to this document benefits you](#)

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PALPABLE PURPURA IN A VIETNAMESE TEENAGE GIRL

Akram Mustafa, MD, Edward Len, MD, Alan Chiu, MSIII

A 19-year-old Vietnamese female with no significant past medical history presented to the emergency department (ED) with fevers, sore throat, generalized myalgias, arthralgias, and a worsening lower extremity rash for the past two weeks. Approximately one week after the onset of constitutional symptoms, the patient noticed a rash developing on the anterior surface of her legs. Three days prior to hospitalization, her primary care physician prescribed cephalexin for her, but she didn't recall what it was for. When her symptoms continued to worsen the next few days, she presented to the ED. In the ED, she also complained of abdominal tenderness. She had no previous hospitalizations, and her vaccinations were up to date. She also had a small tattoo noted on her neck that she reported receiving at a reputable place four years ago. She was sexually active with her boyfriend, and did not regularly use condoms. However, she denied any genitourinary symptoms.

Upon presentation, the patient had a fever and mild tachycardia. However, she was in no acute distress. Physical examination revealed diffuse palpable petechiae along the anterior part of both lower legs. The patient also had a developing rash over the anterior arms bilaterally. The rash spared her palms and soles. Abdominal exam revealed right upper quadrant (RUQ) tenderness with mild hepatomegaly. The rest of the exam was normal. On labs, patient was found to have a microcytic anemia, normal white blood cell and platelet count. Urinalysis revealed mild proteinuria and hematuria. However, the patient had normal renal function. Ultrasound of the kidneys revealed normal-appearing kidneys. Further work-up revealed mildly elevated ALT (98), AST (76), INR (1.86), and ESR (55). With the liver abnormalities, a RUQ ultrasound was also done, which revealed mild hepatomegaly but was otherwise normal. Screens for possible viral (including viral hepatitis, Monospot), bacterial (anti-streptolysin O titers, cultures), and autoimmune (antinuclear antibody, C-reactive protein) causes were all normal.

The clinical picture of palpable purpura, oligoarthritis, abdominal pain and renal involvement as demonstrated by the microscopic hematuria, and further evaluations from Rheumatology, Infectious Disease, and Nephrology led to Henoch-Schonlein purpura (HSP) as the most likely diagnosis in this patient. The exact etiology of the HSP in this patient however was unknown. The most likely triggers were either a previous viral infection or medication-induced.

Skin biopsy was not obtained because the clinical picture was deemed enough to diagnose HSP. The patient demonstrated gradual improvement with supportive care and no further work-up was necessary. The elevated liver enzymes and INR also normalized. Despite the patient's proteinuria, her renal function was normal so a renal biopsy was deemed unwarranted. On hospital day four,



Figure 1. Palpable Purpura

the patient was discharged with instructions to follow up with her primary care physician for any recurring symptoms.

Discussion

Henoch-Schonlein purpura (HSP) is a small-vessel vasculitis characterized by palpable purpura (without thrombocytopenia), abdominal pain, and arthritis. It is a syndrome predominantly seen in children; the highest occurrence of HSP is in patients between the ages of 3 and 5. Ninety percent of HSP cases are in children under the age of ten.¹ However, HSP can be seen at any age, though the occurrence in adults is rare and reported in 3.4-14.3 cases per million.³

The precise etiology of HSP is unknown. The proposed triggers are generally infectious; an upper respiratory infection often precedes HSP symptoms in 90% of cases.² Other proposed infectious agents include group A Streptococcus, MRSA, *H. pylori*, Parvovirus B 19, Hepatitis B, HIV, *Stenotrophomonas maltophilia*.³ In adults, a wider variety of antigenic stimuli have also been suggested to predispose to HSP: vaccinations, insect bites, food allergies and drugs. Medication-induced HSP is particularly more common in adults and has been seen in association with ACE-inhibitors, ARBs, NSAIDs, and antibiotics.²

Whatever the antigenic stimuli may be, the cross reactivity between the offending agent and small vessel endothelial cells leads to a leukocytoclastic vasculitis. Immunoglobulin A (IgA) has been found to be the key mediator of this reaction.² Immune complexes and chemotactic factors activate polymorphonuclear leukocytes causing inflammation and eventually necrosis of vessel walls. IgA complex depositions accumulate primarily in the skin, intestinal mucosa, joints and kidneys—precisely the organ systems behind the classic clinical features of HSP.³

The main clinical features seen with HSP are palpable purpura, abdominal pain, arthritis, and renal insufficiency. A retrospective analysis of 250 adult patients with HSP reported a purpuric rash in 96% of cases, arthritis in 61%, gastrointestinal disease in 47%, and renal disease in 32%.³ The classic skin lesions of HSP start as multiple small, painless, erythematous macular lesions that combine into palpable purpura. The rash is symmetrical and more prominent in dependent areas of the body such as the lower extremities. The arthritis/arthralgia seen in HSP is migratory and oligoarticular. The arthritis is non-deforming and the joints are not usually swollen or tender. Gastrointestinal involvement often presents as colicky pain with or without bleeding from the large or small bowel. A potentially severe complication is intussusception. Renal disease is considered the most serious manifestation of HSP, its involvement can range from microscopic hematuria to fulminant nephrotic syndrome.

The diagnosis of HSP is clinical. The European League Against Rheumatism and Pediatric Rheumatology European Society criteria include one mandatory criterion and at least one of four minor criteria. The mandatory criterion is palpable purpura predominantly in the lower limbs. In addition, there must be one of the following: diffuse abdominal pain, IgA deposition in any tissue biopsy, arthritis/arthralgia, or renal involvement. Urinalyses are often abnormal and may reveal anything from microscopic hematuria to nephrotic range proteinuria. Inflammatory markers (C-reactive protein, erythrocyte

sedimentation rate) are typically elevated. The diagnosis can be supported by a tissue biopsy that shows vasculitis with IgA immunofluorescence.^{2,3} There will also be perivascular infiltration of neutrophils.² In cases of uncertain diagnosis or severe renal disease a renal biopsy may be performed.

Treatment is symptom-directed. NSAIDs are used in mild cases for joint pain. Colchicine can be used in cases of severe skin lesions. In pediatric HSP, no guidelines exist for the management of GI and renal involvement of the disease, and far fewer recommendations exist for adults. Glucocorticoids have been used to treat GI symptoms successfully, as they have been proven to shorten the duration of these symptoms. Corticosteroids can also be used in patients with renal involvement, and in severe disease can decrease proteinuria significantly.² It should be noted, however, that there is little evidence to support the use of glucocorticoids to prevent renal disease once the diagnosis of HSP is established. 6 Leukopheresis, cytotoxic agents and immunoglobulin have been used in refractory cases.^{2,3}

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“Pink Hairy Flower.
Saint Barthelemy, French West Indies”
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