HENOCH-SCHÖNLEIN PURPURA IN AN ADULT: A CASE REPORT AND LITERATURE REVIEW
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Henoch-schönlein purpura (HSP) is a small-vessel leukocytoclastic vasculitis associated with deposition of immunoglobulin A (IgA) within the vessel walls. It classically affects children, however it may also occur in adults. HSP presents with the classic tetrad of palpable purpura, arthritis, abdominal pain, and nephritis. Renal involvement more commonly occurs in adult patients and can have long-term complications. We report a case of HSP in an adult presenting with palpable purpura and hematuria.

CASE PRESENTATION
A 72-year-old Caucasian male presented with a new rash that appeared during a one-month period. He initially noticed “red spots” from the waist down and had mild neck pain. He was treated with oral and intramuscular corticosteroids. The lesions continued to spread involving his arms and lower abdomen. The lesions on his legs began to cluster and ulcerate. He had associated pruritus and pain. He denied any recent illness, infection, fevers, chills, or abdominal pain.

His past medical history was significant for chronic obstructive pulmonary disease and diabetes mellitus type 2. The patient was a former smoker and admits to occasional alcohol consumption. His family history is unremarkable. His medications include albuterol inhaler, tiotropium inhaler, and metformin 500mg BID. He is allergic to penicillin.

Clinical examination of the lower extremities revealed 1+ pitting edema and symmetrically distributed, erythematous, non-blanching petechial macules and papules on the lower extremities (Figure 1). Confluent areas of purpura with central necrosis were also present (Figure 2). The eruption involved his lower extremities, thighs, buttocks, abdomen, and arms.

Routine lab work, including complete blood count (CBC) and comprehensive metabolic panel, were unremarkable.
Urinalysis showed mild protein (300mg/dl), moderate blood, microscopic white blood cells (WBC), 10-20s/hpf, and microscopic red blood cells (RBC), 10-20s/hpf, with hyaline casts. Repeat urinalysis was unchanged. Chest X-ray was unremarkable. Hepatitis panel, ANA, C3, C4, rheumatoid factor, C-ANCA, and P-ANCA were negative.

Biopsies were performed with H & E staining and revealed an unremarkable epidermis with numerous neutrophils and leukocytoclasia surrounding the vessels. Fibrinoid necrosis of the vessel walls, spongiosis, and extravasation of erythrocytes were also noted (Figure 3). These findings were consistent with leukocytoclastic vasculitis. Due to constant proteinuria and hematuria, he was referred to a nephrologist who performed a renal biopsy. Needle biopsy showed mesangial proliferative glomerulonephritis with IgA on immunofluorescence.

The clinical, laboratory, and histopathologic findings were consistent with HSP. He was initially treated with topical corticosteroids, oral prednisone (60mg taper) and hydroxyzine. After no improvement, his systemic therapy was changed to dapsone 50mg daily with prednisone (35mg daily), however the petechia and purpura were persistent. Lastly, he was stared azathioprine 100mg daily. With this medication, his dermatitis began to improve and eventually resolved. His renal function remained stable throughout the course of his diagnosis and treatment.

**DISCUSSION**

HSP is a variant of leukocytoclastic vasculitis, which represents about 10 percent of all cutaneous cases of vasculitis.\(^2\) HSP occurs primarily in children, especially boys between the ages of 2 and 11.\(^3\) The incidence in children ranges from 130 to 180 cases per million, compared to a much lower incidence in adults of 3 to 14 cases per million.\(^4,5\) A slight male predominance is reported. Seasonal pattern is observed with peak incidence during the winter.\(^1\)

The etiology of HSP is not clear, but there have been associations with pregnancy, alcohol, inherited disorders like α-antitrypsin deficiency, infections (especially group A β-hemolytic streptococci), immunizations, and malignancies. HSP following penicillin, vancomycin, clarithromycin, aspirin, losartan, isotretinoin, and adalimumab have been reported.\(^1,2\) Children may present one to two weeks following an upper respiratory tract infection and may have positive anti-streptolysin O (ASO) titers.\(^3\)

Clinically HSP presents with a tetrad of palpable purpura, abdominal pain, arthralgias and hematuria.\(^1,2,3\) The cutaneous findings begin with an urticarial eruption of macules and papules that progress into purpura, hemorrhagic bullae, vesicles, ulcers, targetoid lesions, and necrosis. Necrotic lesions more commonly occur in adults.\(^6\) The eruption is
symmetrically distributed on the lower extremities and buttocks, but may involve the upper extremities, face, and trunk. Although lesions are usually asymptomatic, pruritus, burning, and pain may be reported.\(^2\)

Systemic involvement is seen in up to 80 percent of cases and affects the kidneys, joints, and gastrointestinal tract. Renal disease is the most significant predictor of long-term prognosis.\(^3\) The clinical expression of renal involvement varies from microscopic or gross hematuria to glomerulonephritis. In adults there is a relatively high incidence of renal involvement, 45 to 85 percent, compared to 20-50 percent in children.\(^2\)\(^7\) Nephritis in children occurs within the first two weeks, however in adults it may take months to appear.\(^6\) Despite initial normal urinalysis, repeat urinalyses are recommended to monitor renal function.\(^3,9\) Predictors of chronic renal insufficiency include hypertension, purpura above the waist, pyrexia, elevated ESR, proteinuria, hematuria, or high creatinine levels on initial presentation. Ten to 20 percent of adults and one percent of children with HSP nephritis will progress to end stage renal disease.\(^1,10\)

Gastrointestinal involvement is reported in 50-75 percent of cases and is manifested as colicky abdominal pain, nausea, or vomiting. Rarely gastrointestinal perforation/bleeding, pancreatitis, or intussusception may occur.\(^1,3,8\) Abdominal pain as a presenting symptom is more common in children than in adults. Joint involvement manifests with tender, painful joints, most commonly affecting the knees and ankles, but occasionally including the elbows, hands, and feet.\(^1,3\) Adults typically present with arthralgias without arthritis.\(^8\)

The diagnosis may be made clinically if the patient presents with the classic tetrad of symptoms, however, adults may only exhibit one or two of the classic symptoms. When this occurs, as in our patient, histopathological confirmation is helpful. Biopsies should be taken for H&E and direct immunofluorescence (DIF). H&E will display a perivascular inflammatory reaction with neutrophils, leukocytoclasia (neutrophil degeneration), and extravasated erythrocytes. These findings are indistinguishable from leukocytoclastic vasculitis.\(^1,3\) DIF will demonstrate IgA deposition within the vessel walls. Other laboratory investigation should include a CBC, coagulation studies, serum chemistry profile, urinalysis, and ESR.\(^1,3\) Renal biopsy should be performed in any patient with HSP and continual hematuria and/or proteinuria, as in our patient.\(^11\)

The prognosis for HSP is excellent, and full recovery without sequelae is typical.\(^3\) Approximately 90 percent will have spontaneous resolution of cutaneous lesions within weeks to months.\(^1\) Recurrent flares of HSP may occur in up to three percent of patients. End stage renal disease may occur in up to five percent of patients with nephritis. Acetaminophen or non-steroidal anti-inflammatory agents may be used for symptomatic treatment. If renal involvement occurs, more aggressive therapy with systemic corticosteroids, cyclophosphamide, colchicine, azathioprine, dipyridamole or dapsone may be warranted.\(^3\) Oral colchicine (0.6mg BID-TID) and dapsone (50-200mg/day) are helpful for skin and joint manifestations. Azathioprine (2mg/kg/day) and methotrexate (<25mg weekly) have been successful for renal involvement.\(^1\)

**CONCLUSION**

Although rare, HSP can affect adults and may not present with the classic tetrad of symptoms. Adults more commonly develop hemorrhagic bulla and areas of necrosis. Adults are also more likely to develop chronic renal insufficiency, especially if they have pyrexia, purpura above the waist, and/or an elevated ESR.\(^1\) Prognosis in children is excellent, however in adults the extent of renal involvement determines prognosis. Renal biopsy should be performed in any patient with persistent hematuria and/or proteinuria.\(^11\)

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