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Case Report

Henoch schonlein purpura in adults: a case report

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ABSTRACT

We describe a case of a young adult female with Henoch-Schonlein purpura (HSP) presenting with cutaneous and gastrointestinal manifestations. Biopsy revealed a leukocytoclastic vasculitis in the skin. Steroid therapy led to initial resolution of the symptoms followed by a relapse of rashes and subsequent introduction of azathioprine. HSP is the most common childhood vasculitis and uncommonly seen in adults. Early recognition of the disease, especially in the atypical age group, as in our patient, and appropriate intervention can mitigate the disease and limit organ damage.

Keywords: Henoch-Schonlein purpura, Leukocytoclastic vasculitis, Young adults

INTRODUCTION

Henoch-Schönlein purpura (HSP) is an immunoglobin A (IgA)-mediated vasculitis affecting the vasculature of several systems like the gastrointestinal tract, renal system, skin, and joints. It mainly affects children below 10 years of age and has a male preponderance.

It is characterised by the presence of a vasculitic purpuric rash, abdominal pain, joint pain, renal injury, pulmonary inflammation, or central nervous system involvement. It is usually self-limiting in nature but complications such as gastrointestinal haemorrhage and end-stage renal failure may occur and require prompt diagnosis and treatment.

IgA nephritis affects 60% of patients and can present as asymptomatic microscopic haematuria with proteinuria or as irreversible renal failure requiring renal transplantation.[1]

Diagnosis is made on the basis of criteria set by the European League Against Rheumatism (EULAR), Paediatric Rheumatology International Trial Organisation (PRINTO), and Paediatric Rheumatology European Society (PRES) (Table 1).[2]

Table 1: Diagnostic criteria for HSP, as developed by EULAR/PRINTO/PRES.

Criterion	Description
Mandatory criterion	Purpura or petechiae with lower limb predominance
Minimum 1 out of 4 criteria	1. Diffuse abdominal pain with acute onset.
	2. Histopathology showing leukocytoclastic vasculitis or proliferative glomerulonephritis with predominant immunoglobulin A deposits.
	3. Arthritis or arthralgia of acute onset.
	4. Renal involvement in the form of proteinuria or haematuria.

CASE REPORT

A 22-year-old female presented with a seven-day history of erythematous, non-pruritic rash which started from the thighs and gradually progressed to involve whole of the lower limbs followed by the trunk and the upper limbs sparing the palms and soles.

Later the feet became swollen with moderately intense (7/10) burning pain, aggravated by ambulation. In addition, the patient complained of an acute onset of mild oligoarthropathy affecting her wrists and knees and was migratory in nature. One day later, she started complaining of vomitings which were coffee colored followed by pain abdomen which was localised to the epigastric region, severe in intensity, non-radiating and aggravating on having meals. She had no significant past history and was not on any medication.

On physical examination, there was non-tender, non-blanching purpuric rash involving both upper and lower extremities with non-pitting pedal edema (Figure 1).

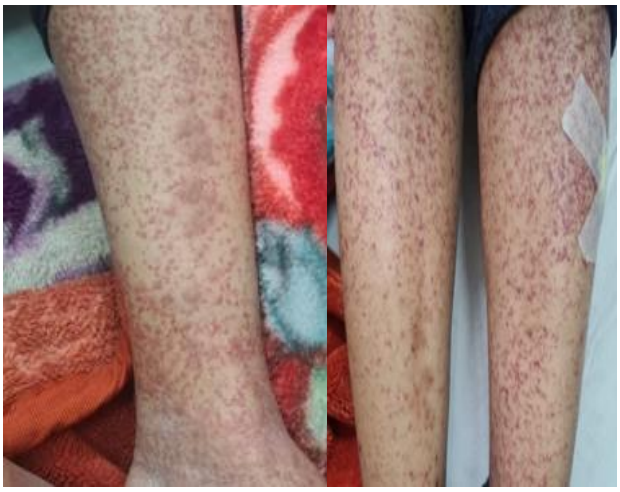

Figure 1: Purpuric rash.

There was truncal involvement. Laboratory tests showed the following: Hb- 12.5 g/dl, Hct- 42.9%, TLC- 8700, BUN- 12 mg/dl, serum creatinine- 0.5 mg/dl, urinalysis- no hematuria or proteinuria, ESR- 16 mm/h, CRP- 14.7 mg/dl, antistreptolysin O titer- 60 IU/l, ANA- negative, HbsAg- negative, Anti-HBc IgG- negative, anti HIV- negative, c-ANCA- negative, p-ANCA- negative, RA factor negative and serum immunoglobulin IgA- 368.60 mg/dl. Skin biopsy showed leukocytoclastic vasculitis of small blood vessels with extravasation of occasional RBCs seen. No fibrinoid necrosis was seen. Dermis in addition showed mild chronic inflammatory infiltrate, more so around blood vessels.

The patient was diagnosed with HSP as per American College of Rheumatology and European League Against Rheumatism (EuLAR) and Pediatric Rheumatology Society (PReS) criteria. She was started on weight based oral prednisone 60 mg once daily, with resolution of symptoms and a visible decrease in the rashes. However, there was a relapse of the rashes on the fourth day of admission and she was started with azathioprine 50 mg once daily in addition to oral prednisone. She was discharged in a satisfactory condition on the ninth day.

Differential diagnosis

Patients presenting with palpable purpura and multisystem involvement (GI, kidney and joints) without thrombocytopenia may be diagnosed as HSP. The differential diagnosis of HSP includes conditions such as Wegener's granulomatosis, infective endocarditis, IgA nephropathy, and hemolytic uremic syndrome. However, in IgA nephropathy, there is no palpable purpura.

Follow-up

Patient is now on regular follow up from last 2 months and steroids are being tapered. Patient does not have any fresh complaints. Figure 2 shows resolution of rashes on 1st month follow-up.

Figure 2: Follow-up.

DISCUSSION

Henoch-Schönlein purpura (HSP) is the most common childhood vasculitis, with an incidence of 3-27 cases per 100,000 child population. It is therefore seen regularly by pediatricians. It can present in any age, even during adulthood, but it is much more frequently seen in childhood and as such the age at peak incidence is around 4-6 years old. In childhood-onset disease, 90% of cases occur under the age of 10 years.[3] HSP is a leukocytoclastic small vessel vasculitis occurring mostly in autumn and winter after upper respiratory tract infections, medications, vaccinations, and malignancies.[4] Self-limiting by nature, HSP is harmless, but severe complications may ensue. 20% and 80% patients can have renal involvement and 7% of all HSP cases can lead to nephrotic or nephritic syndrome and 1% of all patients can develop end-stage renal failure.[5] HSP nephritis (HSPN) usually occurs within 1-2 months after the onset of HSP. Patients can also present with arthralgia and gastrointestinal disturbance. Joint involvement is commonly seen in children and adults presented more frequently with diarrhoea.[6] The pattern of joint involvement is typically oligoarthritis with no redness, warmth, or erythema. In our case, the patient

presented with acute-onset pain in four joints with crampy abdominal pain.

HSP is a clinical diagnosis but laboratory investigations and biopsy help in diagnosis of atypical cases. In addition to routine laboratory investigations, an immune panel of blood is usually done to support the diagnosis and includes ANA, ANCA, RF and factors VIII and XIII levels. Renal and/or skin biopsy may also play a role when the diagnosis is uncertain. The sensitivity of the clinical diagnostic criteria is 100% and specificity 87%, when applied to children.[2] But a few studies have reviewed these criteria to assess applicability to adults, and found a diagnostic sensitivity of 99.2% and specificity.[7] There are currently no specific biomarkers useful for diagnosis of HSP. Some biomarkers can show activity and prognosis of the disease, but none have proven clinically useful.

Skin biopsies remain the gold standard for diagnosing any cutaneous vasculitis. The hallmark feature is IgA-predominant vascular deposits. However, this is not sufficient for the diagnosis of HSP as these deposits can be found in other vasculitic syndromes, erythema nodosum and venous stasis-related conditions. The histological feature seen in skin biopsy is leukocytoclastic vasculitis primarily affecting the small superficial vessels. The vessel walls are infiltrated by neutrophil granulocytes, which partly degenerate and form nuclear dust (leukocytoclasia), located amongst extravasated erythrocytes (purpura) in the surrounding dermis. The vessel walls are thickened and might be necrotic due to exudation of neutrophils and variable amounts of fibrin. On direct immunofluorescence IgA and, eventually, complement C3 can be seen deposited in the vessel walls. Being a non-mandatory criteria, it is argued that skin biopsies are not indicated unless absolutely required. A study carried out on HSP patients with a histological diagnosis of cutaneous leukocytoclastic vasculitis found IgA positivity associated with HSP with a sensitivity of 81% and a specificity of 83%.[8]

The available treatment options aim for symptomatic relief and prevention of renal involvement. Cutaneous involvement usually requires no treatment. As HSP is characterized by IgA deposition and white cell infiltration within blood vessel walls, corticosteroids can play a role in inhibiting this inflammatory process.[9] Usually 1 mg to 2 mg per kg of oral prednisolone for two weeks is effective for abdominal and joint symptoms. However, steroids are of no help in preventing renal complications.[10] Immunosuppressive drugs (cyclophosphamide, azathioprine, cyclosporine A and mycophenolate mofetil) in combination with high-dose IV pulse steroids are recommended if there is no benefit from steroids alone and are usually recommended in rapidly progressive glomerulonephritis (RPGN) and hemorrhagic involvement of the lungs and brain. Most patients completely recover with symptom resolution within eight to 10 weeks of onset and 5% develop chronic symptoms.[11] Complete clinical resolution is more likely in patients with mild renal involvement, no neurological complications, and a disease course of less than six weeks. Disease recurrence may occur in 30% to 50% of patients as late as seven years after the initial onset.

CONCLUSION

An integrated multidisciplinary approach is needed to effectively diagnose and safely manage and monitor patients presenting with HSP. Although self-limiting in nature, HSP has the potential to manifest into life threatening conditions such as end-stage renal failure. This emphasizes the importance of early diagnosis and management. Our case sets a precedence for clinicians to consider HSP in a wider demographic profile.

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