

An Unusual Case of Henoch-Schönlein Purpura

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Abstract

Vasculitis has protean manifestations with etiological non-specificity of histological lesions. The natural history of Henoch-Schönlein purpura in adults is less well established. We report an adult female patient who presented with palpable skin purpura and was evaluated to have IgA nephritis who remains stable on treatment with cyclophosphamide and steroids.

Introduction

Vasculitis, inflammation of vessel walls has many causes, although they result in only a few histologic patterns of vascular inflammation. These protean clinical manifestations, combined with the etiologic non specificity of the histologic lesions, complicate the diagnosis of specific forms of vasculitis. This is problematic because different vasculitides with indistinguishable clinical presentations have very different prognosis and treatments.



Before treatment After treatment

Fig. 1 : Purpura on lower limbs

Table 1 : Investigations

Urine routine	Albumin 3+ RBCs:6-8/hpf Granular casts
24 hour urinary protein	3.2 gram/ day
CT/ BT	2'10''/6' 10''
B Urea and creatinine	42 mg/dl & 0.9mg/dl
Liver function tests	Within normal range
Total count; Differential count	10,700 cells/cumm; P81 L16 M0
Platelet count	2.93 lakh/ cumm
Prothrombin time	Test: 18; control: 15 seconds
HIV/ HBsAg/ anti HCV/ANA/ ANCA	negative
Skin biopsy revealed	Leukocytoclastic vasculitis
Renal biopsy revealed	Mesangial hypercellularity, endocapillary proliferation with mild interstitial edema. No evidence of crescents or basement membrane thickening.
Immunofluorescence:	Mesangial deposits of IgA (4+) and C3(2+) deposition
Upper GI endoscopy revealed	Vasculitis with submucosal hemorrhage

Henoch-Schönlein purpura (HSP) is a leukocytoclastic vasculitis involving small vessels with the deposition of immune complexes containing IgA. It is characterized by the association of skin, joint, and gastrointestinal manifestations that may occur in successive episodes. In addition to these manifestations, renal involvement is common, and the long-term prognosis depends on its severity.

HSP primarily affects children, and its incidence is approximately 15 cases/100,000 children per year; it is less common in adults. Although HSP has been extensively studied in children, much less is known about its natural history in adults. Apart from a recent multicenter Italian study,¹ data on this disease in adults are confined to small series with relatively short follow-up. In adults, however, the incidence of HSP and the severity of its clinical manifestations appear not to be the same as in children.

Among cases of glomerulonephritis, HSP is only responsible for 0.6 to 2% of adult nephropathies.² The incidence of renal involvement in adults varies from 45 to 85% of cases, depending on the data for patients and the definition of renal involvement.² The risk of progression to renal insufficiency, which ranges from 5% to 15% in children, seems to be much higher in adults, approximately 30%. No study has yet been able to demonstrate the ability of any treatment to prevent progression to renal failure in either children or adults.

We report an adult female diagnosed with HSP nephritis. This case comes to the fore as ACR criteria for HSP restricts the age to less than 20 years.³

Case Report

A 32 year old married lady presented with rash of 2 weeks duration which was insidious in onset, non-pruritic, non-

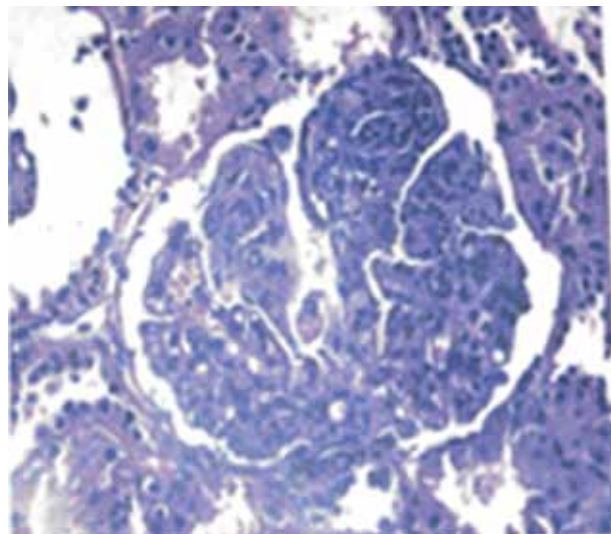


Fig. 2 : Light microscopy: Mesangial hypercellularity, endocapillary proliferation with mild interstitial edema

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Table 2 : According to various studies following are the differences between children and adults with HSP

Features	Children	Adults
Gender distribution	Equal	Male> female
Seasonal	fall and winter	summer and winter
Peviousn URTI	Common	Less common
Diarrhoea	Less common	common
Abdominal pain	common	Less common
Fever	common	Less common
Joint pain	Less common	common
Leucocytosis,	Less common	common
Thrombocytosis	common	Less common
Renal involvement	Less common	Frequent & severe
Hospital stay	Shorter (4.3 days)	Longer (10 days)
Outcome	Very good (93.9% recovery)	Good (89.2% recovery)

blanching, pink-purple raised rash. It started in the lower limbs and progressed over three days duration to involve upper limbs, sparing the face, trunk and mucosa. Desquamation in extremities began at the end of first week. It was associated with facial puffiness and hand and feet edema.

She had mild intermittent fever at the onset of illness which subsided with use of antipyretics. She also had upper abdominal burning pain which increased with food intake and was associated with two episodes of Hematemesis of <50 ml each. She had epistaxis for 2 days.

Relevant drug history included use of over the counter analgesics, antipyretics and antibiotics (quinolones) for her complaints. There was no h/o diarrhea, sore throat or prior jaundice; no h/o decreased urine output.

On examination, she had facial puffiness and palpable tender purpura restricted to extremities and buttocks sparing the face, trunk, palms and soles and mucosa (Fig. 1). Her blood pressure was in the borderline hypertensive range which progressed to frank hypertension in ensuing two days. At the end of first week she had a second crop of purpura with similar distribution. Her systemic examination was normal.

With these details a provisional diagnosis of small vessel vasculitis? Drug induced vasculitis ? HSP was made and she was further investigated. Meanwhile she was on symptomatic treatment.

Her investigations revealed: (Table 1) and Renal biopsy: (Fig. 2)

In the presence of characteristic palpable non thrombocytopenic purpura, abdominal pain and arthralgia with facial puffiness, hypertension, sub-nephrotic range proteinuria and renal biopsy evidence of IgA nephropathy she was diagnosed with Henoch Schonlein purpura with glomerulonephritis and was treated with iv methyl prednisolone 0.5 g for 5 days and continued with oral steroids; she was started on ACE inhibitors for her proteinuria. She has shown significant improvement both in the resolution of skin lesions and abdominal complaints.

Discussion

Henoch-Schönlein purpura preferentially involves venules, capillaries, and arterioles. Susceptibility to HSP may have a genetic origin. Several reports suggest that deficiency of complement 4 (C4) from deletion of C4 genes predisposes

patients to IgA nephropathy and HSP nephritis.⁴ It has been suggested that the IL-1 receptor antagonist (IL1RN2) allele might be a genetic marker shared by patients with HSP nephritis and a group of IgA nephropathy patients with recurrent gross hematuria.

Henoch-Schönlein purpura is most frequent in childhood, with a peak incidence at five years old. The disease often begins after an upper respiratory tract infection.⁶ Purpura, arthralgias, and colicky abdominal pain are the most frequent manifestations. Gastro intestinal manifestations may precede the disease in 14%-36% patients. The rash may rarely present as hemorrhagic vesicles and bullae and desquamation may occur. Localized edematous swelling of the subcutaneous tissues of the lower extremities and hands is frequently observed and does not correlate with the presence or degree of proteinuria. Coppo *et al.*² has shown that children differ from adults by more frequent joint manifestations.. Cutaneous necrosis is uncommon in children and affects fewer than 5% of cases. In contrast necrotic purpura is much more common in adults. Approximately half the patients have hematuria and proteinuria, but only 10 to 20 percent have renal insufficiency. Rapidly progressive renal failure is rare. Pulmonary disease and peripheral neuropathy are uncommon.

The difference between adult and childhood HSP has been described in Table 2.

The diagnosis of HSP is now restricted to the specific clinicopathological entity caused by vascular IgA-dominant immune complexes. Renal biopsy is especially useful in distinguishing HSP from other disorders and for assessing prognosis and need for treatment in patients with renal disease.. IgA ANCA have been reported in patients with HSP and IgA nephropathy by some⁵ but not by others. The misuse of this term for patients with ANCA-associated small-vessel vasculitis who present with purpura, abdominal pain, and nephritis is particularly problematic, because these patients do not have a good prognosis and should be treated quickly with immunosuppressive therapy.

The average duration of the disease is 1 month, even though it may run a protracted course over several years and there is a tendency for recurrences.⁶ Successive episodes of purpura are common during the first weeks of disease but rarely occur beyond the third month. A single attack seldom lasts more than 1 week, and attacks of purpura are associated inconstantly with exacerbation of renal disease. Although the disease is usually self-limited with a good eventual outcome, the glomerulonephritis associated with HSP may uncommonly lead to renal failure, more so in adults. Hypertension, proteinuria and initial renal insufficiency were associated with adverse renal outcome. Hence persistent proteinuria should be aggressively treated with angiotensin-converting enzyme inhibitors.

Childhood onset has a self limited disease course. Microscopic hematurias usually have complete renal recovery. The most accurate prognostic factors are histological presence of crescents, interstitial fibrosis, and extensions of mesangial deposits to include dense subepithelial deposits. The risk is highest in children with crescents in more than half of the glomeruli. In adults even fewer than 50% crescents augurs an unfavorable course. The course of persistent renal sequelae and further renal flare-ups cannot be precisely predicted by histology, and long-term follow-up is warranted.

The overall prognosis is excellent; supportive care suffices for most patients. The main long-term morbidity is from progressive

renal disease. End-stage renal disease develops in approximately 5% of patients. Treatment for aggressive Henoch-Schönlein purpura glomerulonephritis is controversial.

Corticosteroids, immunosuppressive drugs, and anticoagulant therapy have been tried with contradictory results, but a recent study suggests that combined therapy with corticosteroids and azathioprine may be beneficial.

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