

Case Report

Presentation of a patient with Palpable Purpuric Rash

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Abstract

Henoch Schonlein purpura (HSP) is a form of systemic vasculitis characterized by deposition of IgA dominant immune complexes in the small vessels. The triad of palpable purpuric rash on lower extremities, abdominal or renal involvement and arthritis is the typical presentation in this condition. The disease primarily affects children and is less common in adults.

We report a case of a young female who presented with classical symptoms of HSP i.e. palpable purpura on legs, arthritis and off and on abdominal pain.

Introduction

Henoch Schonlein purpura (HSP) is an acute small vessel vasculitis. It is the most common vasculitis in children with an incidence of 15 cases/ 100,000 children per year.¹ Males are affected more often than females. The disease is less common in adults.

Although the cause is unknown, IgA seems to play a central role in the pathogenesis of HSP. The clinical manifestations are the result of widespread vasculitis due to IgA deposition in vessel walls. The major clinical features include a palpable purpuric rash on the lower extremities, abdominal pain or renal involvement, and arthritis. Cutaneous purpura is the essential element in the diagnosis of HSP. The palpable purpura is characteristically 2 to 10mm in diameter and is usually present on the lower extremities.² Small petechiae may be scattered among these lesions.

Arthritis is the second most common feature present in 75% of the patients² and mostly involves the knees and ankles. Gastrointestinal involvement occurs in 50 to 75% of patients and renal involvement in 40 to 50% cases.²

There are no specific diagnostic tests available for diagnosing HSP. Laboratory studies are useful to exclude other conditions that may mimic HSP.^{3,4}

In majority of the cases, the disease is self-limited. Recurrences do occur; however they generally subside in 4 to 6 months. Renal involvement can have chronic consequences and the long term prognosis depends on the severity of renal involvement.

There is no consensus on a preferred treatment. Steroids seem to be the most commonly prescribed therapy. However, the role of pharmacologic treatment is controversial and needs further research.

Case Report

This is a case of a 31 years old lady who presented with complaints of rash on legs, off and on abdominal pain, swelling and pain in the ankles and wrists. She was taking steroids when she presented to us. Her past history revealed similar rash on legs about a year before this episode and she received topical steroids after which the rash disappeared. On examination, she was found to have palpable purpuric rash on the lower limbs.

Her laboratory work up revealed normal platelet count, coagulation studies, ASO titer, renal function and ANA profile. Based on the clinical history and normal laboratory investigations, a clinical diagnosis of HSP was made.

Steroids were gradually tapered and her symptoms resolved with time.

Discussion

Much less is known about the natural history of HSP in adults. Data is confined to small series only.^{5,6} In a study by Blanco et al,⁷ 162 patients with HSP (46 adults and 116 children) were evaluated to compare the differences in disease expression with age. Children were more likely to have preceding upper respiratory tract infection, fever and abdominal pain. Adults had symptoms of joint pain and renal involvement more frequently. However, both these groups had a benign self limited course with complete recovery in 90% cases. Unlike this study, our patient had no renal involvement, but the disease was self limited and complete resolution of symptoms was achieved.

Various case reports of adult onset HSP have also been published.^{8,9} A case report of a 40 years old patient with HSP is also reported from Pakistan;¹⁰ this patient had symptoms of abdominal pain, retrosternal burning and palpable purpura over the lower limbs. *Helicobacter pylori* was the most probable cause of HSP in this patient.

Adult onset HSP is uncommon in our population. However, the physicians should be aware of the clinical presentation of HSP and the complications associated with it.

References

1. Nielsen HE. Epidemiology of Schonlein-Henoch purpura. *Acta Paediatr Scand* 1988; 77: 125-31
2. Saulsbury FT. Clinical update: Henoch Schonlein purpura. *Lancet* 2007; 369: 976-78.
3. Saulsbury FT. Henoch Schonlein purpura in children: Report of 100 patients and review of the literature. *Medicine (Baltimore)* 1999; 78: 395-409
4. Trapani S, Micheli A, Grisolia F, Resti M, Chiappini E, Falcini F, et al. Henoch

Schonlein purpura in childhood: epidemiological and clinical analysis of 150 cases over a 5- year period and review of literature. *Semin Arthritis Rheum* 2005; 35: 143-53

5. Lee HS, Koh HI, Kim MJ, Rha HY. Henoch Schonlein nephritis in adults: a clinical and morphological study. *Clin Nephrol* 1986; 26: 125-30
6. Faull RJ, Arons I, Woodroffe AJ, Clakson AR. Adult Henoch Schonlein nephritis. *Aust N Z J Med* 1987; 17: 396-401
7. Blanco R, Martinez Taboada VM, Rodriguez-Valverde V, Garcia-Fuentes M,

Gonzalez- Gay MA. Henoch Schonlein purpura in adulthood and childhood: two different expressions of the same syndrome. *Arthritis Rheum* 1997; 40: 859-64

8. McClain DM, Maino K, Dwyer TX. Henoch-Schönlein purpura in an adult Filipino man: a case report and literature review. *Cutis* 2006; 77: 236-40
9. Knoll BM, Valles A, Hogan MC. 56- Year- old man with rash, abdominal pain, and arthralgias. *Mayo Clin Proc* 2007; 82 : 745-8
10. Shaikh Z, Ahmed A. Henoch Schonlein purpura associated with gastric helicobacter pylori infection. *Pak J Med Sci* 2004; 20 : 68-70.