Singapore Med J 2007; 48(11) : 1047

Successful treatment of severe gastrointestinal involvement in adult-onset Henoch-Schönlein purpura

Sharma A, Wanchu A, Kalra N, Singh S, Bambery P

ABSTRACT

Henoch-Schönlein purpura is a small vessel vasculitis which is uncommon in adults. The presentations of adult-onset disease are different from those seen in childhood. The commonly-recognised serious gastrointestinal complications of childhood are less well-documented in adults. We report three cases of adult-onset Henoch-Schönlein purpura with severe gastrointestinal involvement. All were men, aged 22, 35 and 42 years, respectively. Two of these patients had evidence of mesenteric ischaemia on computed tomography of the abdomen. All three patients were successfully treated with steroids.

Keywords: adult-onset Henoch-Schonlein purpura, gastrointestinal disease, Henoch-Schonlein purpura, mesenteric vasculitis, small vessel vasculitis

Singapore Med J 2007;48(11):1047–1050

INTRODUCTION

Henoch-Schönlein purpura (HSP) is a small vessel vasculitis characterised by palpable purpura, arthritis, renal and gastrointestinal (GI) involvement.[1] It is the most common vasculitic disorder of childhood, with a reported mean age of 5.9 years.[2] It is uncommon in adults. The presentations of the adult-onset disease are different than those seen in childhood. The commonly-recognised serious GI complications of childhood are less well-documented in adults. We report three cases of adult-onset HSP with severe GI involvement, which were successfully treated with steroids.

CASE REPORTS

Over a period of three years (2003–2005), three patients with adult-onset HSP were admitted to the Adult Rheumatology Unit of the Internal Medicine Department at the Postgraduate Institute of Medical Education and Research, Chandigarh, a tertiary care teaching hospital in India. They were all diagnosed to have HSP based on the American College of Rheumatology criteria.[3] A detailed history was elicited and physical examination was carried out, with special attention given to the extent of skin rash and the manifestations of inflammatory arthritis. Laboratory parameters included haematological parameters, biochemistry, urine analysis for microscopy and 24-hour urinary protein excretion, chest and abdominal radiographs and ultrasonography of the abdomen. Contrast-enhanced computed tomography (CT) of abdomen was performed in two patients.

The clinical profiles, along with laboratory and radiological data, of the three patients are shown in Table I. All the patients were men, with a mean age of 33 years. Two patients had the appearance of rash prior to joint symptoms and abdominal pain, while these appeared simultaneously in the third case. As all three patients had severe abdominal pain, obstipation and vomiting, the possibility of mesenteric ischaemia was considered. CT of abdomen was performed in Cases 1 and 3. There was mural thickening with stratification of iliac loops, and multiple vessels were seen on the mesenteric border arranged in a pallisading manner (comb sign) (Fig. 1) in Case 1. Abnormal mural enhancement of the proximal small bowel with attenuation of the superior mesenteric vein suggestive of vasculitis was seen in Case 3. Skin biopsies in two patients (Cases 1 and 2) were suggestive of leucocytoclastic vasculitis on histopathological examination, while the sample was inadequate in the third case. Direct immunofluorescence (DIF) revealed Ig A and C3 with absence of Ig G and Ig M in two patients (Cases 2 and 3). All the patients were managed with continuous Ryles tube aspiration, nil by mouth, and intravenous fluids. One patient (Case 1) was given 1 g intravenous methylprednisolone every day for three days, while the other two (Cases 2 and 3) were given oral prednisolone 1 mg/kg/day. All the patients had complete recovery from their abdominal symptoms within approximately two weeks.

Department of Internal Medicine, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

Sharma A, MD
Assistant Professor

Wanchu A, MD, DM
Additional Professor

Singh S, MD
Professor

Bambery P, MD
Professor

Department of Radiology

Kalra N, MD
Assistant Professor

Correspondence to:
Dr Aman Sharma
Tel : (91) 172 275 6681
Fax: (91) 172 274 4401
Email: amansharma74@yahoo.com

Table I. Salient features of the three patients with HSP.

Feature	Case 1	Case 2	Case 3
Age (years)/Gender	42/M	35/M	22/M
Pain in abdomen (days)	3	During admission	4
Skin rash	Buttocks, lower limbs, back	Upper, lower limbs	Lower limbs, buttocks, lower abdomen
Joints involved	Ankles, wrists, small joints of hands	Ankles, knees, elbows, wrists	Knees
Haemoglobin (g/dL)	13.7	17	13.7
Total leucocyte count (/μL)	24,100	10,400	15,200
Platelet count (/μL)	412,000	360,000	471,000
ESR (mm/1st hour)	54	12	92
Urine microscopy	Microscopic haematuria	Normal	Microscopic haematuria
24-hour urinary proteins	Nil	Nil	Nil
Skin biopsy			
HPE	Leucocytoclastic vasculitis	Leucocytoclastic vasculitis	Inadequate sample
DIF	Not done	Ig A, C3	Ig A, C3
CT of the abdomen	Mucosal oedema target sign	Not done	Mural enhancement of small bowel
Treatment	Methylprednisolone pulse (1 g/day × 3 days)	Oral prednisolone (1 mg/kg/day)	Oral prednisolone (1 mg/kg/day)
Outcome	Complete recovery	Complete recovery	Complete recovery

ESR: erythrocyte sedimentation rate; HPE: histopathology; DIF: direct immunofluorescence

Case One

A 42-year-man presented with a three-week history of rash over the lower limbs, buttocks and back, associated with bilateral ankle pain and swelling. Three days prior to admission, he developed colicky abdominal pain, which worsened while in the hospital. On examination, he was tachycardic and had severe abdominal tenderness with absent bowel sounds. There was swelling and tenderness of both wrists and at the small joints of the hands. His haemoglobin level was 13.7 g/dL, total leucocyte count (TLC) was 24,100/μL and platelet count was 412,000/μL. There was microscopic haematuria but no proteinuria. Guvaic test for stool occult blood was positive. Rheumatoid factor and cryoglobulins were negative. Skin biopsy showed evidence of leucocytoclastic vasculitis. CT of the abdomen showed mural thickening with stratification of ileal loops. Multiple vessels were seen on the mesenteric border arranged in a pallisading manner (comb sign) (Fig. 1). He was given 1 g of intravenous methylprednisolone every day for three days, followed by oral prednisolone (1 mg/kg/day). With these measures, he had complete resolution of skin lesions and abdominal symptoms over two weeks. The follow-up CT done after one month was completely normal (Fig. 2).

Case Two

A 35-year-man presented with inflammatory arthritis involving his ankles, knees, elbows and wrists, and palpable purpuric skin rash over both upper and lower extremities for ten days. He developed colicky abdominal pain and vomiting during hospitalisation. His general physical and systemic examination was normal. His haemoglobin level was 17 g/ dL, total leucocyte count was 10,400/μL and platelet count was 360,000/μL. His abdominal radiographs and ultrasonography were normal. Skin biopsy from purpuric lesions was suggestive of leucocytoclastic vasculitis and on DIF, Ig A and C3 deposits were seen. He was given oral prednisolone 1 mg/kg/day. With these measures, he recovered completely within two weeks.

Case Three

A 22-year-old man developed a purpuric rash involving his lower limbs, buttocks and abdomen, of seven days duration, associated with bilateral knee pain and swelling. He also had abdominal angina. On examination, he was tachycardic with diffuse abdominal tenderness and absent bowel sounds. His haemoglobin level was 13.7g/dL, total leucocyte count was 15,200/μL and platelet count was 471,000/μL. Guvaic test for stool for occult blood was positive. Synovial fluid total leucocyte count was 100/mm^3, with polymorphonuclear predominance. There was microscopic haematuria but no proteinuria. CT of his abdomen revealed abnormal mural enhancement

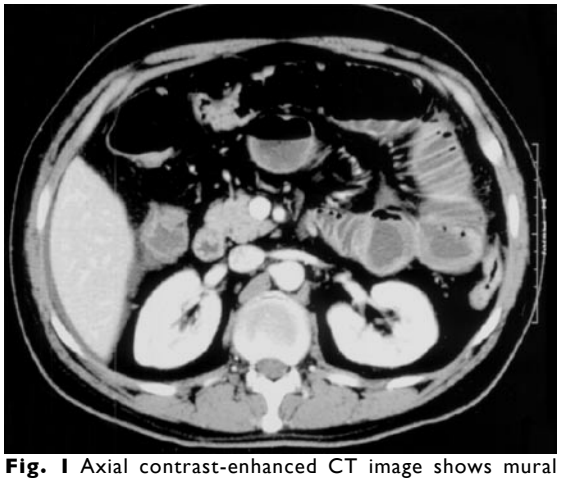

Fig. 1 Axial contrast-enhanced CT image shows mural thickening with stratification of ileal loops. There are multiple vessels seen on the mesenteric border arranged in a pallisading manner (comb sign).

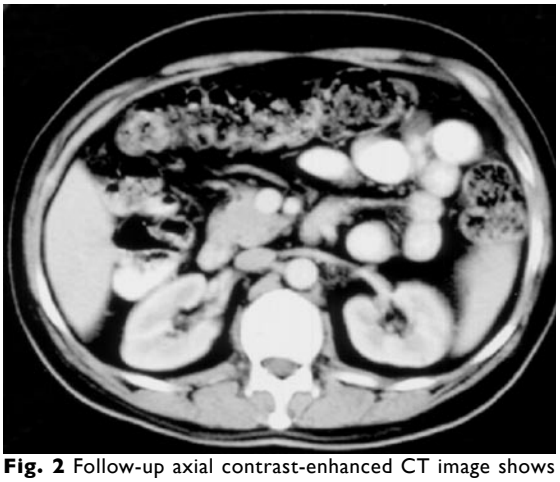

Fig. 2 Follow-up axial contrast-enhanced CT image shows normal wall thickening of the ileal loops. The vessels on the mesenteric border are also not seen.

of proximal small bowel. Skin biopsy material for histopathology was scanty but there was Ig A and C3 positivity on DIF. He was given oral prednisolone 1 mg/kg/day, and he recovered completely over the span of two weeks.

DISCUSSION

HSP is the most common acute vasculitic illness affecting children, but is relatively uncommon in adults, with reports occurring in patients up to 86 years of age.[4] Very few studies have focused on the various aspects of adult-onset HSP. Blanco et al reported a higher incidence of renal and lower GI involvement at disease onset in adult HSP, but during the clinical course, GI involvement was the same in both age groups (63.8% in children and 53.6% in adults, p is not significant). GI bleeding occurred more frequently in adults (59.1%) compared to children (28.3%, p = 0.01).[5] Although GI involvement has been reported to be either low[5] or the same[6] as in children, all our patients had severe GI involvement. The reported incidence of GI involvement in adult-onset HSP varies from 19% to 56.5%.[5,7] The common GI features are abdominal pain (86%), massive colorectal bleed (20%), occult blood loss (66%), vomiting (40%), and diarrhoea (20%).[8] Intussusception is a well-recognised complication of childhood HSP, but is rare in adults. Two of our patients (Cases 1 and 3) had transient microscopic haematuria, but none had proteinuria. This is in contrast to earlier studies in which renal involvement in adult-onset HSP were reported to be more common.[5,6]

Two of our patients had thrombocytosis, which also has been reported earlier in patients with HSP.[9] Ig A and C3 deposits were looked for and seen in two of the patients (Case 2 and 3). Ig A seems to play a pathogenic role in HSP, as supported by its presence in cryoprecipitate[10] and the finding of an increased number of circulating Ig A-secreting cells in patients with active HSP.[11,12] Moreover, Ig A-dominant immune deposits can be seen in most patients with HSP,[13] and its presence has been included in the definition adopted by the consensus conference of the nomenclature of systemic vasculitides.[14] Cutaneous purpura is the sine qua non for the diagnosis of HSP[3] and was present in all our patients. It preceded joint pains in two patients. Although there have been reports of abdominal symptoms preceding purpuric lesions by 2–14 days,[2] the onset in our patients was after the skin lesions.

The use of corticosteroids in the treatment of HSP was first reported in the 1950s,[15] but to date, there has not been prospective controlled studies on the use of corticosteroids in treatment of HSP. However, abundant clinical experience supports the value of corticosteroid therapy in ameliorating GI symptoms in HSP.[2] There have been case reports suggesting that corticosteroids pulses may be helpful in patients with massive GI haemorrhage and widespread mesenteric vasculitis.[16] One of our patients was administered methylprednisolone pulse for three days, while the other two were treated with oral prednisolone. All three recovered completely. In conclusion, HSP is uncommonly seen in adults and can have serious GI involvement, such as small bowel infarction, which may be fatal.[17] Timely use of corticosteroids may be successful in treating even serious forms of mesenteric vasculitis.

REFERENCES

1. Patrignelli R, Sheikh SH, Shaw-Stiffel TA. Henoch-Schönlein purpura. A multisystem disease also seen in adults. Postgrad Med 1995; 97:123-4, 127, 131-4.
2. Saulsbury FT. Henoch-Schönlein purpura in children. Report of 100 patients and review of the literature. Medicine (Baltimore) 1999; 78:395-409.
3. Mills JA, Michel BA, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of Henoch-Schönlein purpura. Arthritis Rheum 1990; 33:1114-21.
4. Kassutto S, Wolf MA. Clinical problem-solving. A wrinkle in time. N Engl J Med 2003; 349:597-601.
5. Blanco R, Martínez-Taboada VM, Rodríguez-Valverde V, García-Fuentes M, González-Gay MA. Henoch-Schönlein purpura in adulthood and childhood: two different expressions of the same syndrome. Arthritis Rheum 1997; 40:859-64.
6. García-Porrúa C, Calviño MC, Llorca J, Couselo JM, González-Gay MA. Henoch-Schönlein purpura in children and adults: clinical differences in a defined population. Semin Arthritis Rheum 2002; 32:149-56.
7. Tancrede-Bohin E, Ochonisky S, Vignon-Pennamen MD, et al. Schönlein-Henoch purpura in adult patients. Predictive factors for IgA glomerulonephritis in a retrospective study of 57 cases. Arch Dermatol 1997; 133:438-42.
8. Novák J, Márki-Zay J, Csiki Z, et al. [Schoenlein-Henoch purpura in adulthood (gastrointestinal manifestation and endoscopy)]. Z Gastroenterol 2001; 39:775-82. German.
9. Saulsbury FT, Kesler RW. Thrombocytosis in Henoch-Schönlein purpura. Clin Pediatr (Phila) 1983; 22:185-7.
10. Garcia Fuentes M, Chantler C, Williams DG. Cryoglobulinaemia in Henoch-Schönlein purpura. Br Med J 1977; 2:163-5.
11. Casanueva B, Rodriguez-Valverde V, Merino J, Arias M, Garcia-Fuentes M. Increased IgA-producing cells in the blood of patients with active Henoch-Schönlein purpura. Arthritis Rheum 1983; 26:854-60.
12. Casanueva B, Rodriguez-Valverde V, Luceño A. Circulating IgA producing cells in differential diagnosis of Henoch Schönlein purpura. J Rheumatol 1988; 15:1229-33.
13. Hené RJ, Velthuis P, van de Wiel A, et al. The relevance of IgA deposits in vessel walls of clinically normal skin. A prospective study. Arch Intern Med 1986; 146:745-9.
14. Jennette JC, Falk RJ, Andrassy K, et al. Nomenclature of systemic vasculitides. Proposal of an international consensus conference. Arthritis Rheum 1994; 37:187-92.
15. Ashton H, Frenk E, Stevenson CJ. Therapeutics. XV. The management of Henoch-Schonlein purpura. Br J. Dermatol 1971; 85:199-203.
16. Wang L, Huang FC, Ko SF, Cheng MT. Successful treatment of mesenteric vasculitis caused by Henoch-Schönlein purpura with methylprednisolone pulse therapy. Clin Rheumatol 2003; 22:140-2.
17. Chan JC, Li PK, Lai FM, Lai KN. Fatal adult Henoch-Schönlein purpura due to small intestinal infarction. J Intern Med 1992; 232:181-4.