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-Schönlein purpura, gastrointestinal disease, Henoch-Schönlein purpura, mesenteric vasculitis, small vessel vasculitis

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, obstruction 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 palisading 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.
Inadequate sample

Case One
A 42-year-old 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. Guvacc 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 pallsiding 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-old 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 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. 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. Guvacc 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 pallsiding 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).

Table 1. Salient features of the three patients with HSP.

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

ESR: erythrocyte sedimentation rate; HPE: histopathology; DIF: direct immunofluorescence
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. 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). Although GI involvement has been reported to be either low or the same 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%. The common GI features are abdominal pain (86%), massive colorectal bleed (20%), occult blood loss (66%), vomiting (40%), and diarrhoea (20%). 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.

Two of our patients had thrombocytosis, which also has been reported earlier in patients with HSP. 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 and the finding of an increased number of circulating Ig A-secreting cells in patients with active HSP. Moreover, Ig A-dominant immune deposits can be seen in most patients with HSP, and its presence has been included in the definition adopted by the consensus conference on the nomenclature of systemic vasculitides. Cutaneous purpura is the sine qua non for the diagnosis of HSP 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, 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 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. There have been case reports suggesting that corticosteroids pulses may be helpful in patients with massive GI haemorrhage and widespread mesenteric vasculitis. 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. Timely use of corticosteroids may be successful in treating even serious forms of mesenteric vasculitis.
REFERENCES


2008 SMJ Best Research Paper Awards

The Singapore Medical Association will be presenting awards for the Best Research Paper published in the Singapore Medical Journal (SMJ) in 2008. All original research papers that are published in the SMJ during the one year period from January 1, 2008 to December 31, 2008 will be considered for this award.

The following are the judging criteria:

- The paper with the most potential impact on clinical practice
- Most rigorous study design/research methodologies
- Comprehensive data analysis and balanced discussion
- Data interpretation

Distinguished members of the medical profession will be invited to serve on our panel of judges for selecting the winning papers.

The authors of the winning papers selected by our panel of judges will receive cash prizes for the first, second and third places. Prize winners will also receive a commemorative trophy and certificate.

We thank you for your support of the SMJ. The quality of our journal depends on the quality of your submissions.