Henoch-Schönlein Purpura

Background

- Henoch-Schönlein Purpura (HSP) is the most common cause of non-thrombocytopenic purpura in children.
- The cause is unknown, but it is an IgA-mediated vasculitis of small vessels, usually following an upper respiratory tract infection (URTI).
- It is more common in children than in adults, with a peak incidence in the 2-8 year age group, and a seasonal peak in winter.
- The onset of the illness may be acute, with simultaneous involvement of several organ groups, or insidious, with sequential occurrence of features over weeks to months.
- It is usually a self-limiting disease.
- Many children will need only a urine test and can be sent home with GP follow up.

Assessment

Clinical Features

- The child is generally well-looking
- There is often a history of a recent URTI
- Small-vessel vasculitis may involve several organ groups, in particular the skin, joints, gastrointestinal tract and kidneys
- Rarely, there may be CNS or pulmonary involvement
### Rash
- Initially blanching pink maculopapules which may be discrete or confluent
- Progresses to petechiae or purpura, which are often raised (“palpable purpura”)
- The distribution of the purpura typically involves gravity-dependent body areas, particularly the buttocks, legs and extensor surfaces of the arms.

### Oedema
- Vasculitis of dermal vessels also results in angio-oedema
- Non-pitting, often painful, oedema of dependent areas (hands and feet) as well as the eyelids, lips or scrotum.

### Arthritis
- Occurs in two-thirds of patients
- Self-limiting serous joint effusions which resolve over several days
- Typically involving the gravity-dependent joints (elbows, wrists, knees, ankles) – swollen, painful joints; may cause difficulty in weight-bearing

### GIT
- Abdominal pain is common, and is generally intermittent and colicky in nature
- Diarrhoea (with occult blood) is common
- Occasionally, frank haematemesis or melaena may occur
- More serious, but infrequent, complications include spontaneous bowel perforation or intussusception

### Renal
- 90% of patients have microscopic haematuria, but this is persistent or recurrent in only 5% of cases
- Serious complications may include acute glomerulonephritis, nephrotic syndrome, acute renal failure (<1%), or isolated hypertension
- Renal involvement may be a late manifestation, up to 6 months after initial presentation.

### Differential Diagnosis
- Meningococcal septicaemia
- Thrombocytopenia
- Other vasculitides
- Patients initially presenting with only with abdominal pain or arthralgia may pose a diagnostic challenge
- Patients initially presenting with haematuria only should have other causes of haematuria excluded

### Investigations
- There is no diagnostic investigation for HSP. Investigations are used to detect complications or other causes of purpura

#### Urine
- Microscopic haematuria is common (90%)
- If macroscopic haematuria is present, urine should be checked for RBC casts (nephritis) and protein (nephrotic syndrome)

**Consider**
• FBC to exclude thromocytopaenia as cause of purpura if diagnosis is uncertain
• Blood culture, white cell count, CRP, meningococcal PCR if meningococcal septicaemia is suspected
• Urea, creatinine and electrolytes if renal impairment is suspected
• Complement levels and ASO-titres if nephritis is present

**Management**

Most patients with HSP will be managed as an outpatient with symptomatic treatment (analgesia) and follow up.

- Document BP
- Surgical consult for abdominal complications or testicular pain (testicular pain from vasculitis may be difficult to differentiate from testicular torsion)
- Abdominal X-Ray or ultrasound may be needed to exclude abdominal complications
- There is some evidence to suggest that prednisolone 1 mg/kg/day for 2 weeks benefits in abdominal and joint pain (and oedema)
  - There is very limited evidence in preventing renal complications
  - The decision whether to start prednisolone should be made by the admitting Paediatric Consultant

**Admission criteria**

- Abdominal complications (intussusception, perforation, haematemesis or bloody stools)
- Renal complications (nephritis, nephrotic syndrome, hypertension, renal failure)
- Symptomatic relief (severe joint pain or abdominal pain, painful oedema)

**Referrals and follow-up**

- Patients should receive follow up with their GP or Paediatrician for at least 6 months to ensure symptom relief and disease resolution
- Check BP and urinalysis weekly for one month, then monthly for 6 months after presentation as renal disease may present late
  - If still present at 6 months refer to the Renal team
- Provide [Henoch-Schonlein Purpura Health Facts](#)

**Nursing**

Routine nursing care.
Tags

abdominal, angio oedema, arthritis, blanching, haematuria, henoch schonlein purpura, hsp, maculopapules, oedema, petechiae, purpura, rash

References