Henoch-Schönlein Purpura

Henoch-Schönlein purpura (HSP) is an IgA-mediated, autoimmune hypersensitivity vasculitis of childhood. The main clinical features are skin purpura, arthritis, abdominal pain, gastrointestinal bleeding, and nephritis. The aetiology remains unknown.

Epidemiology\textsuperscript{[1, 2]}

HSP is a rare condition but is the most common form of systemic vasculitis in children; 90\% of cases occur in childhood under the age of 10 years. The peak prevalence is in children aged 4-6 years. It is rare in infants and young children. It occasionally occurs in adolescents and adults, and tends to be more severe and more likely to cause long-term renal problems when it occurs in adults.

It is estimated to affect 10 to 20 per 100,000 children per year. In the UK, the estimated annual incidence is 6-20 cases per 100,000 population\textsuperscript{[3]}. Caucasians are more often affected than other ethnic groups.

Aetiology\textsuperscript{[2, 4]}

The cause is unknown, but a mix of genetic, immune and environmental factors appear to be involved. The condition tends to be seasonal and there is often a history of recent infection. Infections preceding HSP include those involving group A streptococci, mycoplasma, Epstein-Barr virus, Coxackievirus, hepatitis A and B, parvovirus B19, campylobacter, varicella and adenoviruses. Vaccination has also been described as a trigger. There is also an association with malignancy; usually solid tumours rather than haematological malignancies, and more common in adult males.

IgA immune complexes are involved in the pathophysiology of HSP, depositing in the small blood vessels of the skin, joints, kidneys and gastrointestinal tract, causing an inflammatory reaction.

Presentation

- The disease occurs mostly in the autumn or winter months. There may be a history of a preceding upper respiratory tract infection (URTI) or less commonly a gastrointestinal infection.
- Generally, patients appear to be mildly ill, with low-grade fever.
- There is a symmetrical, erythematous macular rash, especially on the back of the legs, buttocks and ulnar side of the arms.
- Within 24 hours, the macules evolve into purpuric lesions, which may coalesce and resemble bruises. Typically the purpura are slightly raised and palpable.
- Abdominal pain and bloody diarrhoea may precede the typical purpuric rash. HSP may also cause nausea and vomiting. Such gastrointestinal symptoms precede the rash in 10-40\% of patients\textsuperscript{[1]}.
- Joint pain, especially in the knees and ankles. Joints may also be swollen and tender but permanent deformity does not occur.
- Renal involvement\textsuperscript{[5]}:
  - Affects approximately 40\% of children affected with HSP.
  - Only a small minority progresses to end-stage kidney disease.
  - Usually occurs within three months of disease onset.
  - There is usually no relationship between the severity of nephritis and the extent of the other manifestations of HSP.
  - Microscopic haematuria with mild-to-moderate proteinuria may occur.
  - Nephrotic syndrome may also occur.
  - Oliguria and hypertension are uncommon.
- Scrotal involvement may mimic testicular torsion.
- Headaches may occur, and occasionally seizures and other nonspecific neurological symptoms.

The classic tetrad is a palpable purpuric rash, joint pains, gastrointestinal symptoms and renal involvement. A number of diagnostic criteria exist, the most recent being that proposed by European League Against Rheumatism/ Paediatric Rheumatology International Trials Organisation/ Paediatric Rheumatology European Society (EULAR/PRINTO/PRES) in 2010\textsuperscript{[6]}. This set of criteria states that for a diagnosis of HSP there must be palpable purpura, which is not thrombocytopenic/ petechiae, and one or more of the following:

- Diffuse abdominal pain.
- Typical histopathology (leukocytoclastic vasculitis or proliferative glomerulonephritis with predominant IgA deposits).
- Arthritis or arthralgia.
- Renal involvement (demonstrated by quantified proteinuria or haematuria).

Differential diagnosis

- Intussusception should be considered, even if the typical purpuric rash has evolved (intussusception occurs in 3-4\% of HSP patients\textsuperscript{[7]}).
- Connective tissue diseases - eg, systemic lupus erythematosus (SLE).
Other causes of purpuric rash - eg, thrombocytopenia, meningococcal meningitis.
Other causes of glomerulonephritis.
Other causes of gastrointestinal symptoms such as inflammatory bowel disease.
Acute haemorrhagic oedema of infancy: a self-limiting condition presenting with fever, oedema and rosette-shaped, annular-shaped or targetoid-shaped purpura affecting the face, ears and extremities. [8]

Investigations
Diagnosis of HSP is clinical and not based on laboratory investigations, and there is no definitive test. The following tests may be relevant:

- Urinalysis (should always be performed): haematuria and/or proteinuria are present in 20-40% of patients [3].
- FBC: there may be raised white cell count with eosinophilia; normal or increased platelets. Helps in excluding other diagnoses such as thrombocytopenia.
- Raised ESR.
- Serum creatinine may be elevated in renal involvement.
- Serum IgA levels are often increased. This is not diagnostic.
- Autoantibody screen: connective tissue diseases.
- Abdominal ultrasound: if there are gastrointestinal symptoms - for diagnosis of intestinal obstruction.
- Barium enema: may be used to confirm and treat intussusception.
- Testicular ultrasound: assessment of possible torsion.
- Renal biopsy: if there is persistent nephrotic syndrome.
- Screening for cancer should be considered for older adults who develop HSP with no preceding infection [1].

Management
HSP is usually self-limiting and no form of therapy has been shown appreciably to shorten the duration of disease or prevent complications. Therefore, treatment for most patients remains primarily supportive and is entirely symptomatic unless there is renal involvement. This includes rehydration, pain relief, wound care for ulcerative lesions and treatment for intussusception where present.
- Non-steroidal anti-inflammatory drugs (NSAIDs) may help joint pain but should be used with caution in patients with renal insufficiency or gastrointestinal symptoms.
- Admission to hospital may be required in some cases for monitoring of abdominal and renal complications.
- There is no consensus on prevention or treatment of renal involvement. A 2015 Cochrane review found no significant benefit for treatment with steroids, antiplatelet agents or cyclophosphamide [9]. Low numbers made it impossible for the review to conclude whether there was a role for ciclosporin and mycophenolate mofetil in those with severe renal disease.
- Corticosteroids can ameliorate associated arthralgia and the symptoms associated with gastrointestinal dysfunction, but it seems there is no place for routine use and no evidence of benefit of prednisone in preventing serious long-term kidney disease in HSP [10].
- Plasma exchange is used in the management of some adults with vasculitis and idiopathic rapidly progressive nephritis but further trials are needed [1, 11].

GPs are likely to be involved in monitoring for renal involvement, usually under written guidance from the secondary care specialist. Typical monitoring advice would be [3]:

- For those with no proteinuria, blood pressure checking and urinalysis at days 7 and 14 and at 1, 3, 6 and 12 months.
- For those with proteinuria, follow-up at days 7 and 14, monthly from 1-6 months and then at 12 months.

Complications
Renal involvement occurs in up to 55% of children with HSP but is usually not serious, with manifestation ranging from microscopic haematuria and mild proteinuria to nephrotic and nephritic syndrome and renal failure [1]. Less than 1% of patients with HSP progress to end-stage kidney disease [10]. The renal prognosis is worse in older children and in adults.
- Other rare complications include myocardial infarction, pulmonary haemorrhage, pleural effusion, intussusception, gastrointestinal bleeding, bowel infarction, testicular haemorrhage or torsion, intracranial haemorrhage, seizures and mononeuropathies.
- Recurrence of symptoms may occur and does so in up to one third within 4-6 months of the initial presentation [2].

Prognosis
HSP is an acute self-limited illness and usually resolves fully without treatment, but may rarely lead to complications. Initial attacks of HSP can last for several months. Recurrence is relatively common as above.
- The majority resolve fully within four weeks [2].
- Chronic kidney disease may progress, sometimes more than ten years after the initial flare [12].
- The long-term prognosis of HSP is directly dependent on the severity of renal involvement [9].
- One study reported that adults with HSP had a higher frequency of renal insufficiency and worse renal outcomes than children [13].
Further reading & references

5. Roache-Robinson P, Hotwagner DT; Henoch Schonlein Purpura (Anaphylactoid Purpura, HSP) ...

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