Henoch-Schonlein Purpura

By

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Introduction

• The disease is eponymously named after Eduard Heinrich Henoch (1820-1910), a German pediatrician, and his teacher Johann Lukas Schönlein (1793-1864), who described it in the 1860s.
*HSP is a common vasculitis of small vessels with cutaneous & systemic complications.

- Its etiology is unknown & often follows URTIs.
- Most incidence in age group 2 to 8 yrs.
- Males affected twice then females.
Pathogenesis

• Pathogenesis of HSP is unknown.
• Pts with HSP have significantly higher frequency of HLA-DRB1*01 and decreased frequency of *07 haplotype than controls.
• During active disease there is increased serum concentration of cytokines TNF alpha & IL-6.
* In 50% pts there is increase in ASO antibodies, implicating group A streptococcus.

* Immunofluorescence techniques show deposition of IgA & C3 in small vessels of skin & renal glomeruli.
Direct immunofluorescence for IgA
Clinical manifestations

- Onset is acute with systemic manifestations simultaneously or insidious.
- Low grade fever and fatigue.
- Rash beginning as maculo papular then progress to petechie or palpable purpura in dependant areas of body which is the hallmark of the disease.
- The lesions occur in crops last from 3 to 10 days & may appear at intervals that vary from few days to several months.
• **Edema**.. primarily in dependent areas & is independent of purpura.

• **Arthritis**.. present in more then 2/3 of children, usually involves knees and ankles. It is associated with edema.

• **GI manifestations**.. Intermittent abd pain, enlarged mesenteric lymph nodes segmental edema & hemorrhage into the bowel. More than 50% pts have occult blood in stools or hematemesis.
Cont...

- **Intussusception**.. may occur which rarely followed by complete obstruction or infarction and needs surgical intervention.

- **Renal involvement**.. Occurs in 25-50% cases and may manifest with hematuria, proteinuria, nephritis, nephrosis or ARF, which may lead to ch. HTN or end stage renal disease.
Cont...

• Hepatosplenomegaly and lymphadenopathy may also be present.
• Neurological involvement a rare but serious complication resulting in seizures, paresis or coma.
Rare complications

- Rheumatoid like nodules,
- Cardiac & eye involvement
- Mono neuropathies
- Pancreatitis
- Pulmonary or intramuscular hemorrhage.
Diagnosis

• Mainly clinical, routine lab tests are non-specific.
• Moderate thrombocytosis, leukocytosis and elevated ESR.
• Anemia may be present.
• 50% pts have elevated conc. of IgA as well as IgM.
• Anti cardiolipin or antiphospholipid antibodies may be present.
• Intussusceptions is usually ilio ileal in location, barrium enema may be used for diagnosis and non surgical reduction.
• Urine R/E shows RBCs, WBCs, casts or albumin.
• RFTs. Serum urea & creat. may be raised.
• Vessel biopsy, to confirm vasculitis.
• Renal biopsy. Mesangial depositions of IgA, occasionally Ig m, C3 & fibrin.
Differential diagnosis

• **Thombocytopenic purpura.** More common b/w age 3 to 7 yrs. Spleen is not palpable. Capillary fragility test is positive. Bleeding time is prolong & plts. count is decreased.

• **Poly arteritis nodosa.** Cutaneous lesions are different and peripheral, neurological and cardiac manifestations are more common.
• **Meningococcemia.**

• **Kawasaki disease.** unremitting fever, maculopapular rash which is prominent on lower extremities and peripheral arthritis.

• **JRA.** Salmon pink rash disappearing and is maculopapular. Swelling doesn’t extend beyond the joint.

• **AHE.** leukocytoclastic vasculitis seen in children < 2yr. fever, tender edema (of face, scrotum, hands & feet.) & ecchymosis.
Treatment

- **Symptomatic**: adequate hydration, balance diet and pain control.
- **Arthritis**: is self-limiting. Avoidance of competitive activities and pain control.
- **Edema**: elevation of scrotum & local cooling.
- **GIT**: hydrostatic reduction or resection of intussusception and oral or IV corticosteroids (1-2 mg/kg/day).
**Chronic or recurrent HSP:** IV methyl prednisolon 30mg/kg/day (max 1gm/day). For 3 days. Followed by 1 to 2 times weekly tapered acc to response.

**Renal:** as other forms of glomerulonephritis. High dose corticosteroids, & cyclophosphomide or azathioprine in pts e crescentic glomerulonephritis.
Complications

- Nephrotic syndrome.
- Bowel perforation
- Testicular torsions
Prognosis

- Self limiting disease with overall good prognosis.
- <1% develop persistent renal disease and 0.1% develop serious renal disease.
- Death rarely occurs due to bowel infarction, CNS or renal disease.
Abstract  Levels of von Willebrand factor antigen (vWF: Ag) and factor XIII activity (F XIII) were studied in relation to the severity of clinical symptoms (scored from 0 to 3) and to immunological parameters [IgA, C3, C4, and circulating immune complexes (CIC) in 16 children (7 males, 9 females, aged 3–11 years) with Henoch-Schonlein purpura (HSP) at presentation. vWF: Ag was increased in 7 patients, F XIII activity was decreased in 6. In all children we found high levels of IgA, while C3 and C4 levels were normal; CIC were elevated in 11. vWF: Ag correlated with clinical score and with IgA and CIC, probably as a result of immune-mediated endothelial cell damage. The haemostatic alterations observed in HSP are important for understanding the pathophysiology of the disease.
THANKS