Henoch-Schonlein Purpura

BY

Dr Arshad Malik

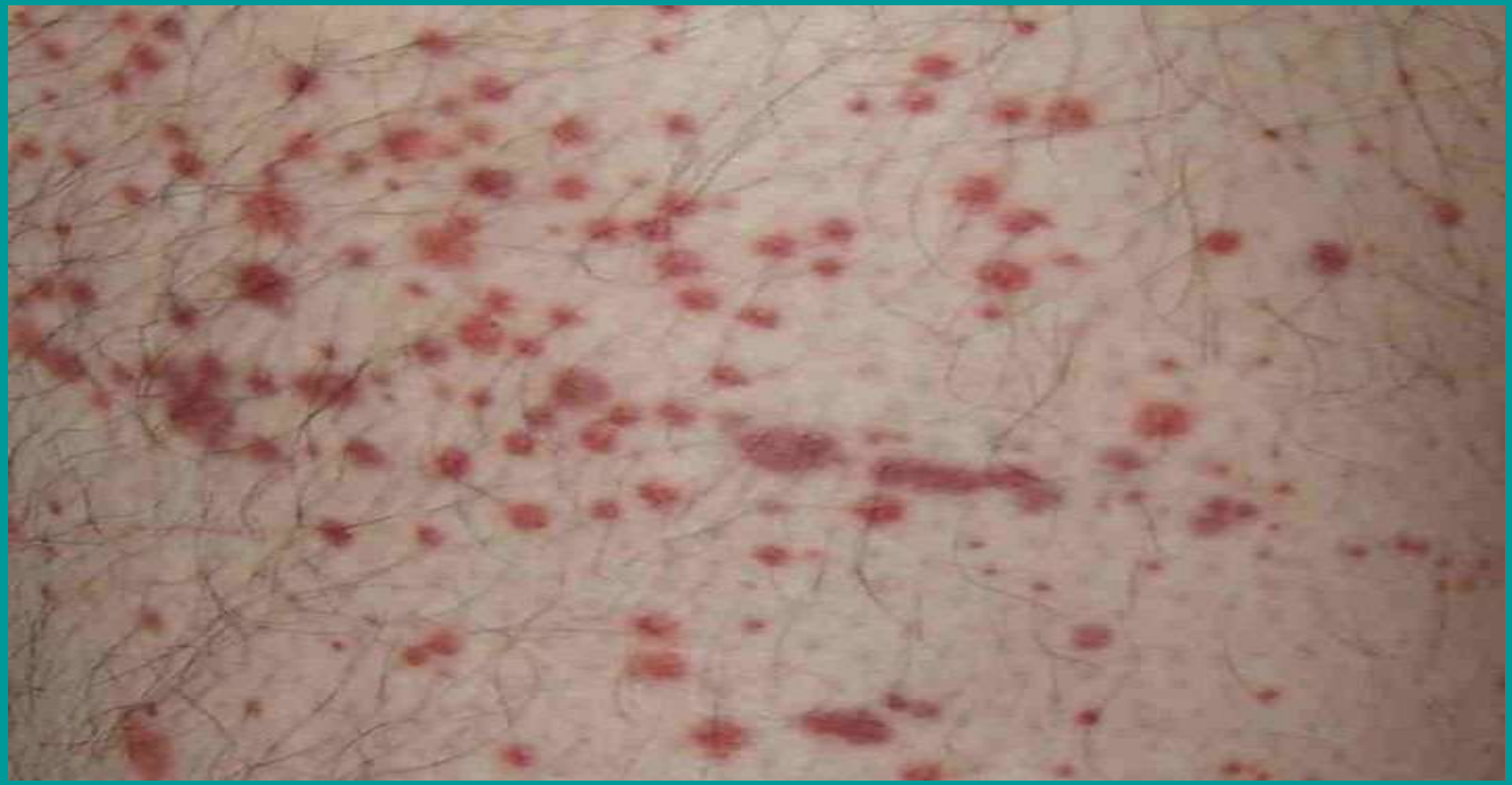
Introduction

- The disease is eponymously named after Eduard heinrich Henoch (1820-1910), a German pediatrician, and his teacher Johann Lukas Schonlein (1793-1864), who described it in the 1860s.

Cont…

*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.

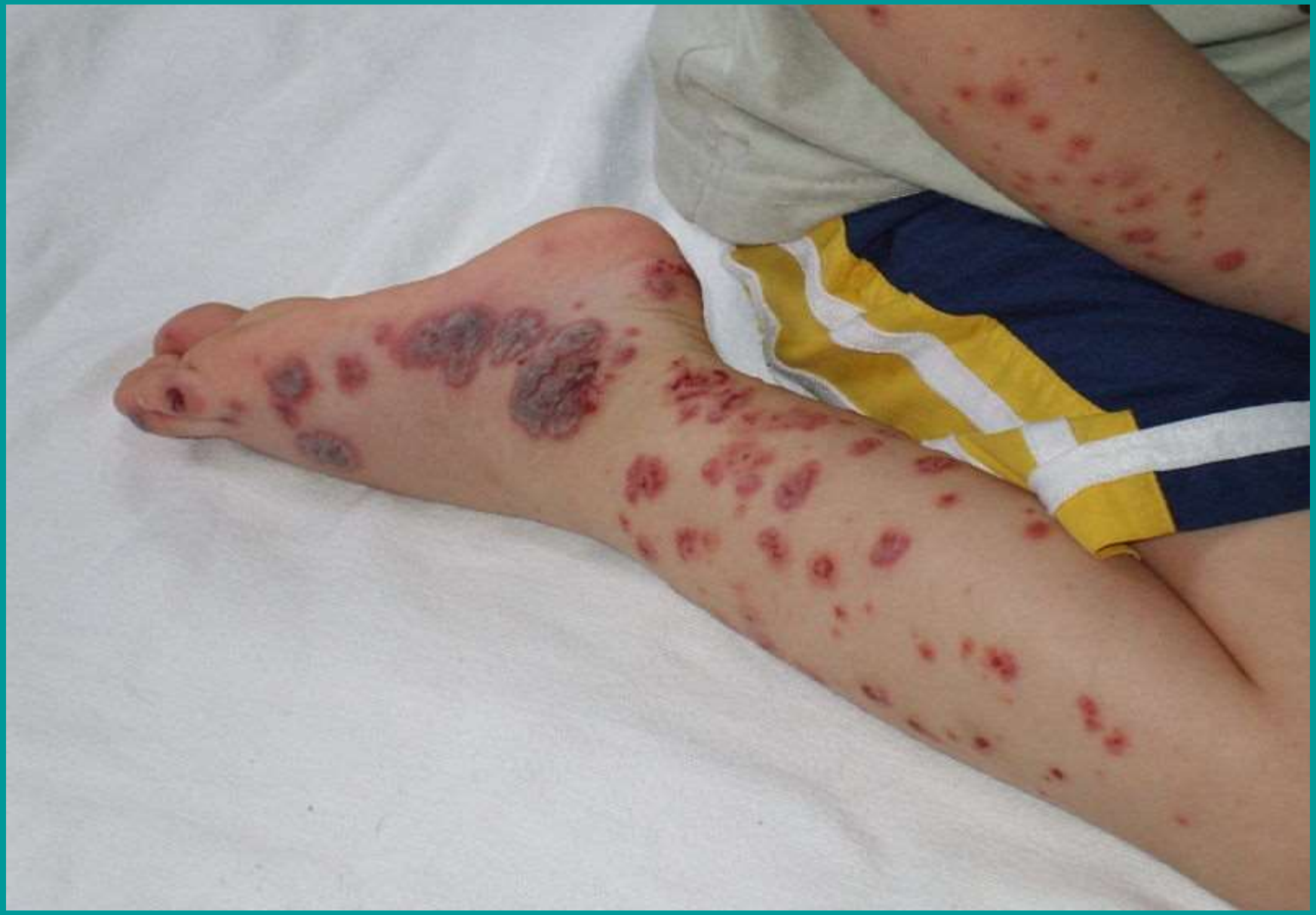
Cont…..

* 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.



Cont……

- **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 then 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 anti bodies may be present.

Cont…

- **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.

Cont…

- ***Meningococcemia.***
- ***Kawasaki disease***..unremitig fever, maculopapular rash which is prominent on lower extremities and peripheral arthritis.
- ***JRA***.. Salmon pink rash disappearing and is maculopapular. Sweling doesn't extend beyond the joint.
- ***AHE***.. leulkocytoclastiC vasculitis seen in children < 2yr.fever , tender edema(of face, scrotum, hands& feet.) & echymosis.

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 corticsteroids(1-2 mg/kg/day).

Cont…

- **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% dev serious renal disease.
- Death rarely occurs due to bowel infarction, CNS or renal disease.

Recent advances

british journal of nephrology

- **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