”JSC “Astana Medical University
Department of Internal Diseases № 1

SIW

THEME:

“SCHONLEIN- HENNOCH PURPURA ”

Checked by: Baidurin S.A
Done by: Suleymenova D. 463GM.

.Astana 2018y

Henoch-Schönlein Purpura (HSP)

- is a common vasculitis of small vessels with cutaneous and systemic complications.

- It is the most common cause of nonthrombocytopenic purpura in children.

EPIDEMIOLOGY

The etiology is unknown

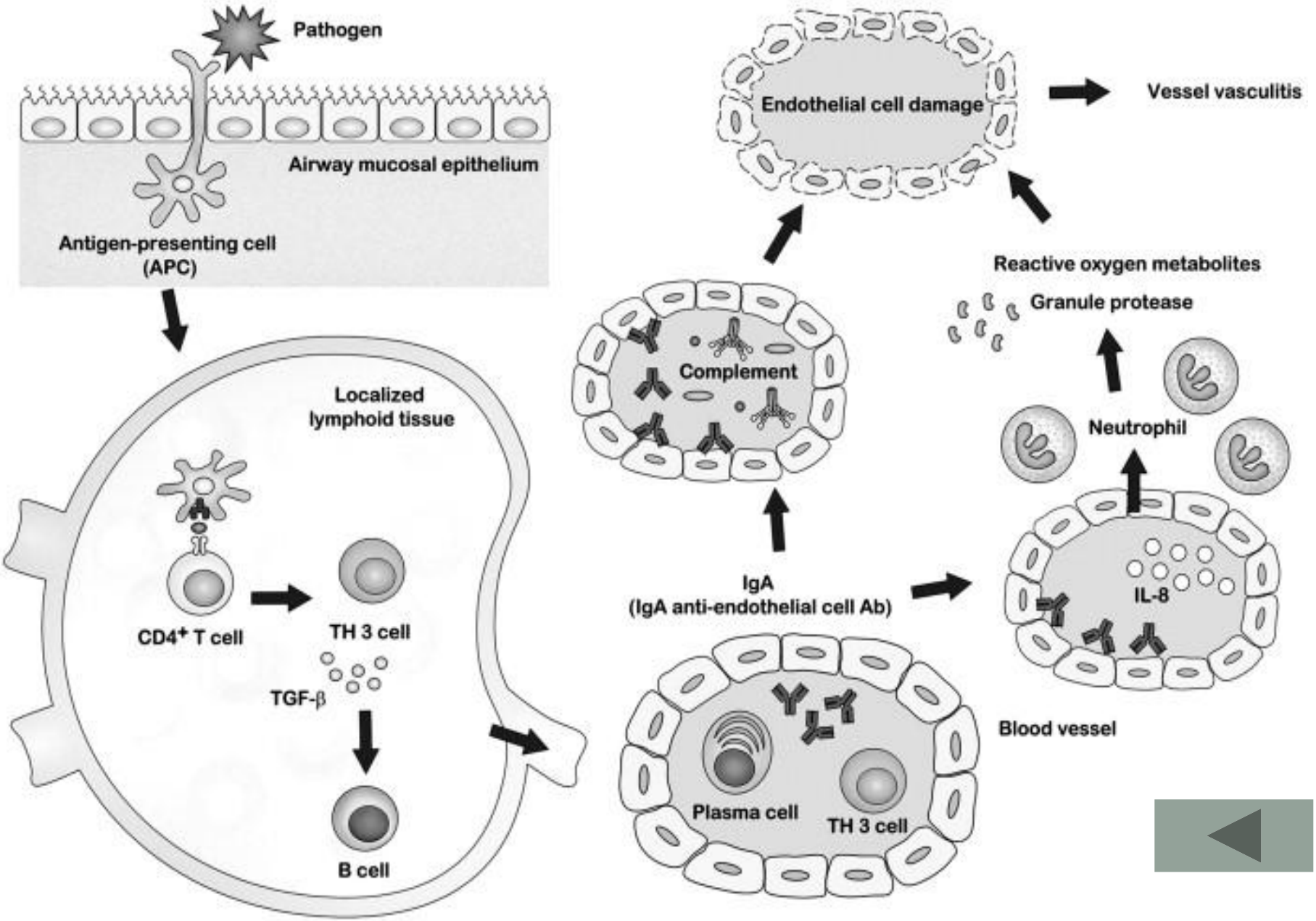
- more frequent in children than adults, with most cases occurring between 2 and 8 yr of age,
- most frequently in the winter months.
- The overall incidence is estimated to be 9/100,000 population.
- Males are affected twice as frequently as females.

PATHOGENESIS

- in specific populations, patients with HSP have a significantly higher frequency of HLA-DRB1*01 and decreased frequency of the *07 haplotype than controls.
- increased serum concentrations of the cytokines tumor necrosis factor-α (TNF-α) and interleukin (IL)-6.
- In one study, almost half of the patients had elevated antistreptolysin O (ASO) anti-bodies, implicating group A streptococcus.
- HSP is an IgA-mediated vasculitis of small vessels.
- Immunofluorescence techniques show deposition of IgA and C3 in the small vessels of the skin and the renal glomeruli;

CTLA-4 +49 A/G genotype and HLA-DRB1 polymorphisms in Turkish patients with Henoch-Schönlein purpura.

- presence of Cytotoxic T lymphocyte-associated protein 4(CTLA-4) AG genotype and HLA-DRB1*13 could be a risk factor for developing nephrotic-range proteinuria in these patients.


Pathogen
Airway mucosal epithelium
Antigen-presenting cell
(APC)
Localized
lymphoid tissue
CD4+ T cell
TH 3 cell
TGF-β
B cell
Endothelial cell damage
Vessel vasculitis
Complement
Reactive oxygen metabolites
Granule protease
Neutrophil
IL-8
IgA
(IgA anti-endothelial cell Ab)
Plasma cell
TH 3 cell
Blood vessel

The immunobiology of Henoch-Schönlein purpura.

- **group *A beta-hemolytic streptococcus*** (GAS) has widely studied and found in 20–50% of patients with acute HSP by serological tests or bacterial cultures,
- ***Bartonella henselae*** (12 of 18 HSP patient sera were positive)
- ***Parvovirus B19*** (only one of 29 HSP patients)

Other HSP-associated pathogens have been reported

- *Staphylococcus aureus*,
- *Helicobacter Pylori*,
- *Hemophilus parainfluenza*,
- Coxsackie virus,
- adenovirus,
- hepatitis A virus,
- hepatitis B virus

CLINICAL MANIFESTATIONS

- The disease onset may be acute, or insidious, with sequential occurrence of symptoms over a period of weeks or months.
- Low-grade fever and fatigue are present in more than half of affected children.
- The typical rash and the clinical symptoms of HSP are a consequence of the usual location of the acute small vessel damage primarily in the skin, gastrointestinal tract, and kidneys.

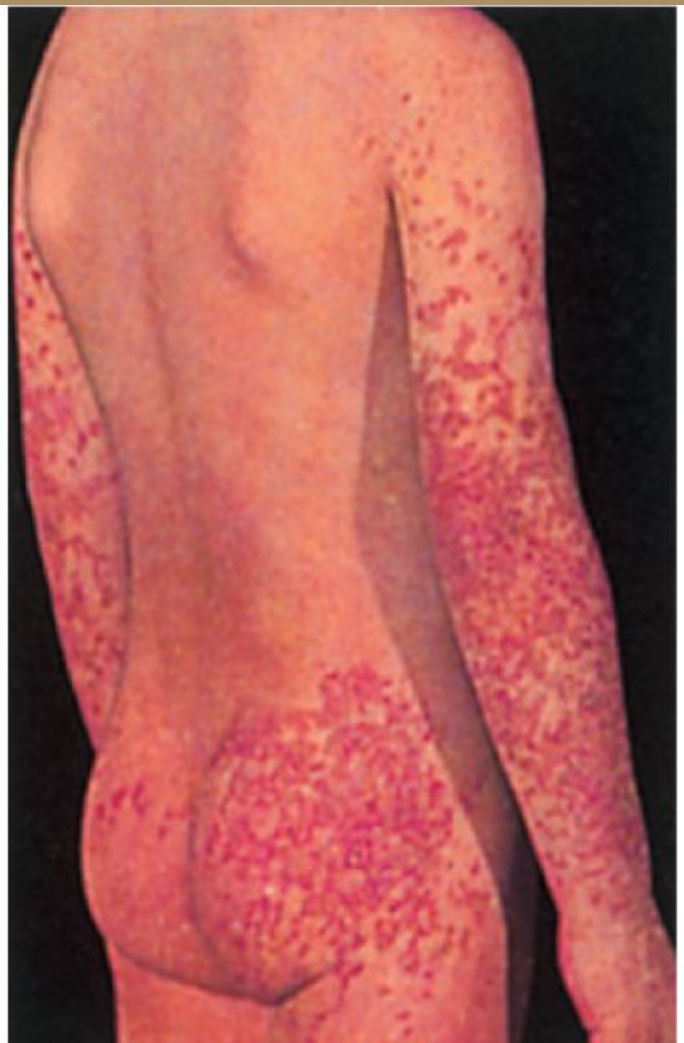
CLINICAL MANIFESTATIONS

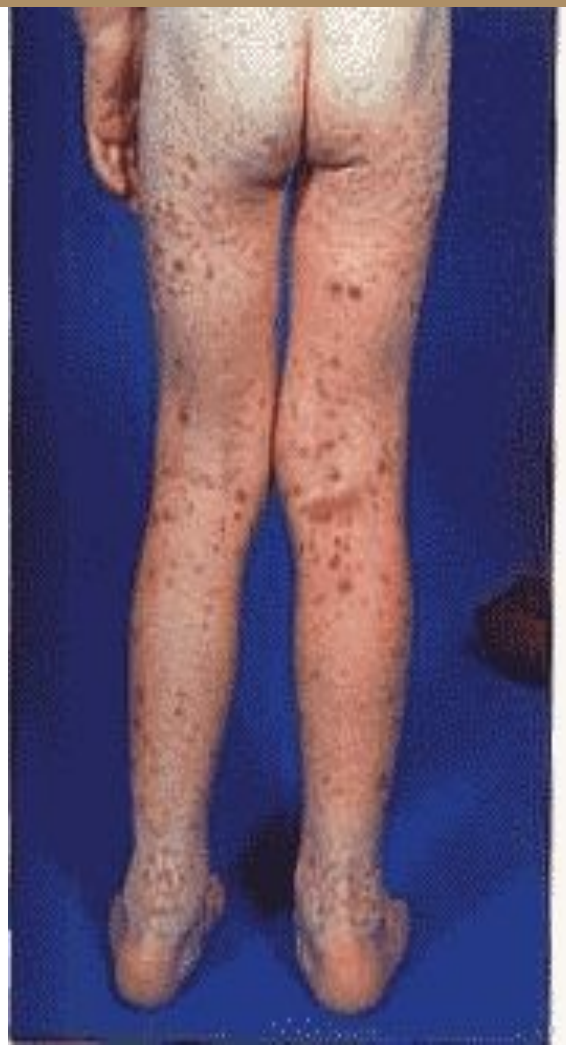
- Rash (95-100%), especially involving the legs, may not be present on initial presentation
- Subcutaneous edema (20-50%)
- Abdominal pain and vomiting (85%)
- Joint pain (60-80%), especially involving the knees and ankles
- Scrotal edema (2-35%)
- Bloody stools

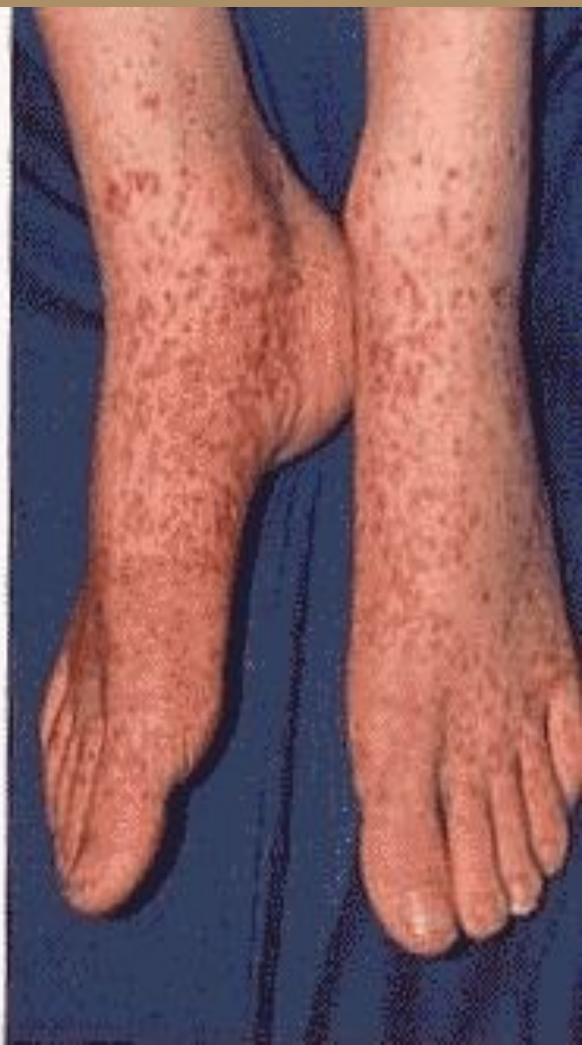
Rash

- beginning as pinkish maculopapules that initially blanch on pressure and progress to petechiae or purpura,
- characterized clinically as **palpable purpura** that evolve from red to purple to rusty brown before they eventually fade
- last from 3-10 days, and may appear at intervals that vary from a few days to as long as 3-4 mo.
- In <10% of children, recurrences of the rash may not end until as late as a yr,
- Damage to cutaneous vessels also results in local angioedema, which may precede the palpable purpura.
- Edema independent of purpura occurs primarily in dependent areas such as below the waist, over the buttocks (or on the back and posterior scalp in the infant), or in areas of greater tissue distensibility such as the eyelids, lips, scrotum, or dorsum of the hands and feet.

Rash







Arthritis

- present in more than ⅔ of children with HSP,
- is usually localized to the knees and ankles and appears to be concomitant with edema.
- The effusions are serous, not hemorrhagic,
- resolve after a few days without residual deformity or articular damage.
- They may recur during a subsequent reactive phase of the disease.

Gastrointestinal tract

- intermittent abdominal pain that is often colicky in nature.
- There may be peritoneal exudate, enlarged mesenteric lymph nodes, segmental edema, and hemorrhage into the bowel.
- More than half of patients have occult heme-positive stools,
- diarrhea (with or without visible blood), or hematemesis.
- Intussusception may occur, which may rarely be followed by complete obstruction or infarction with bowel perforation.
- If not resolved by hydrostatic reduction during a contrast study, surgical intervention is necessary.

Renal involvement

- occurs in 25-50% of children

may manifest with:

- hematuria,
- proteinuria, or both;
- nephritis or nephrosis;
- acute renal failure.

- Renal involvement at presentation may lead to chronic hypertension or end-stage renal disease in the future

Increased serum levels of insulin-like growth factor (IGF)-1 and IGF-binding protein-3 in Henoch-Schonlein purpura.

Serum IGF-1 levels were significantly higher in HSP with proteinuria than those without proteinuria and controls ($p = 0.001$ and $p = 0.001$, respectively).

Also, IGFBP-3 levels were greater in HSP with proteinuria compared to those without proteinuria and controls ($p = 0.005$ and $p = 0.0001$).

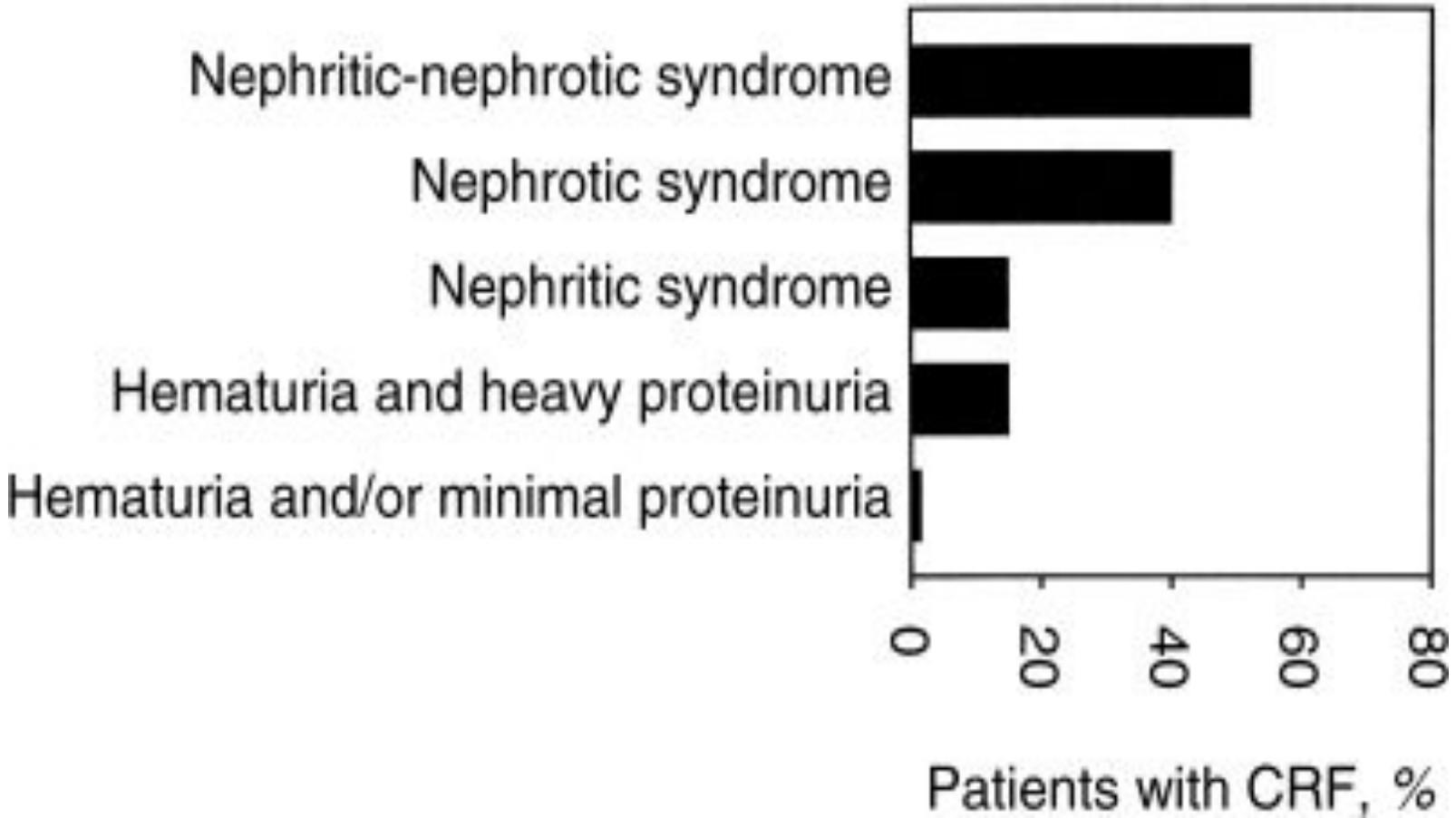
Serum immunoglobulin-A/complement-C3 ratio was higher in HSP than in the controls ($p = 0.0001$) but this ratio did not change according to proteinuria, hematuria or positive SOB.

In conclusion, IGF-1 and IGFBP-3 levels could be new markers for determination of renal involvement in HSP.

What is the difference between IgA nephropathy and Henoch-Schönlein purpura nephritis

Clinical features	IgAN	HSPN
Extra-renal symptoms	-	+
Age at onset	<15 y	>15 y
Nephritic/nephrotic syndrome	-/+	+++
Risk of chronic renal failure (CRF)	+	++
Hypersensitivity	-	+
Secondary forms	++	-/+
Endocapillary proliferation	-/+	++
Epithelial crescents	-/+	++
Perivascular glomerular IgA	-/+	++
Subepithelial/subendothelial dense deposits	-/+	++
Fibrin deposits	-/+	++
IgA-containing complexes size	7S-19S	<19S

Relationship between initial clinical signs and risk of chronic renal failure in Henoch-Schönlein purpura nephritis



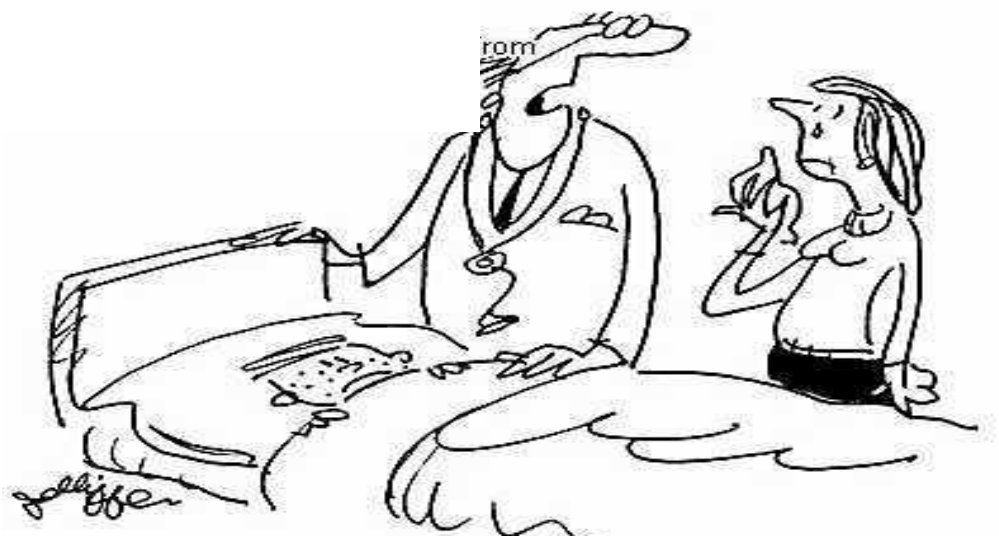
Complications of Henoch-Schönlein Purpura

Hepatosplenomegaly
- **Myocardial infarction**
- **Pulmonary hemorrhage**
- **Pleural effusion**
- **Unnecessary abdominal surgery**
- **Intussusception**
- **Hemorrhage**
- **Shock**
- **Gastrointestinal bleeding**
- **Bowel infarction**
- **Renal failure**
- **Hematuria**
- **Proteinuria**
- **Seizures**
- **Mononeuropathies**

Testicular torsion

DIAGNOSIS

Diagnostic uncertainty arises when the symptom complex of edema, rash, arthritis with abdominal complaints, and renal findings occurs for a prolonged period.

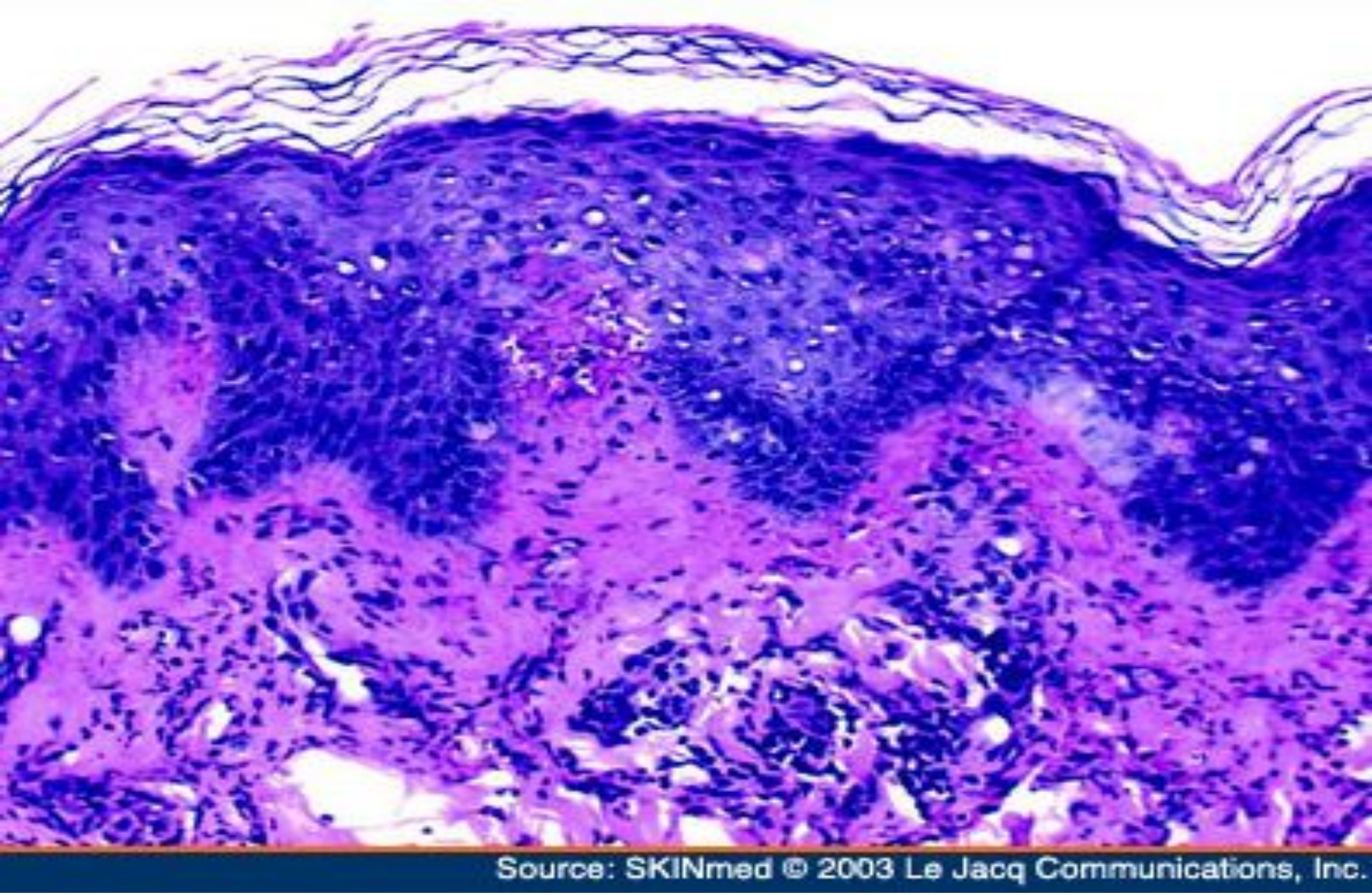


DIAGNOSIS

- Affected children often have a moderate **thrombocytosis** and **leukocytosis.**
- The erythrocyte sedimentation rate **(ESR) may be elevated**.
- **Anemia** may result from chronic or acute gastrointestinal blood loss.
- Immune complexes are often present, and 50% of patients have **elevated** concentrations of **IgA** as well as **IgM**
- **usually negative** for antinuclear antibodies (ANAs), antibodies to nuclear cytoplasmic antigens (ANCAs), and rheumatoid factor (even in the presence of rheumatoid nodules).
- **Anticardiolipin or antiphospholipid antibodies** may be present and contribute to the intravascular coagulopathy.
- **Intussusception** is usually ileoileal in location;
- **Renal involvement** manifests in red blood cells, white blood cells, casts, or albumin in the urine and azotemia

Definitive diagnosis confirmed by biopsy

- cutaneous site showing leukocytoclastic angiitis.
- Renal biopsy may show mesangial deposition of IgA and occasionally IgM, C3, and fibrin.



H & E stain of skin biopsy showing leukocytoclastic vasculitis with infiltration of neutrophils.

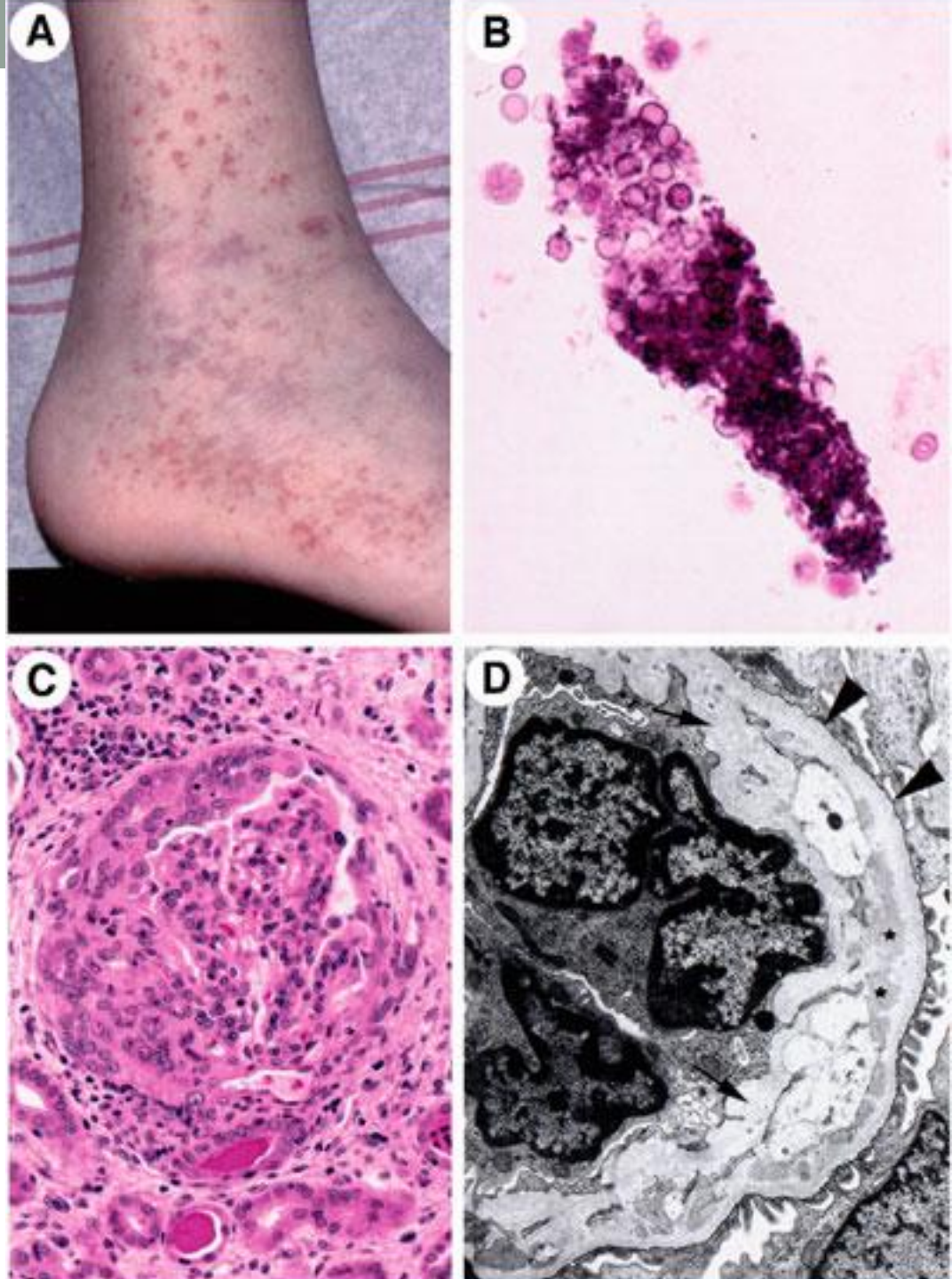
Henoch-Schönlein purpura.

A: Cutaneous purpura;

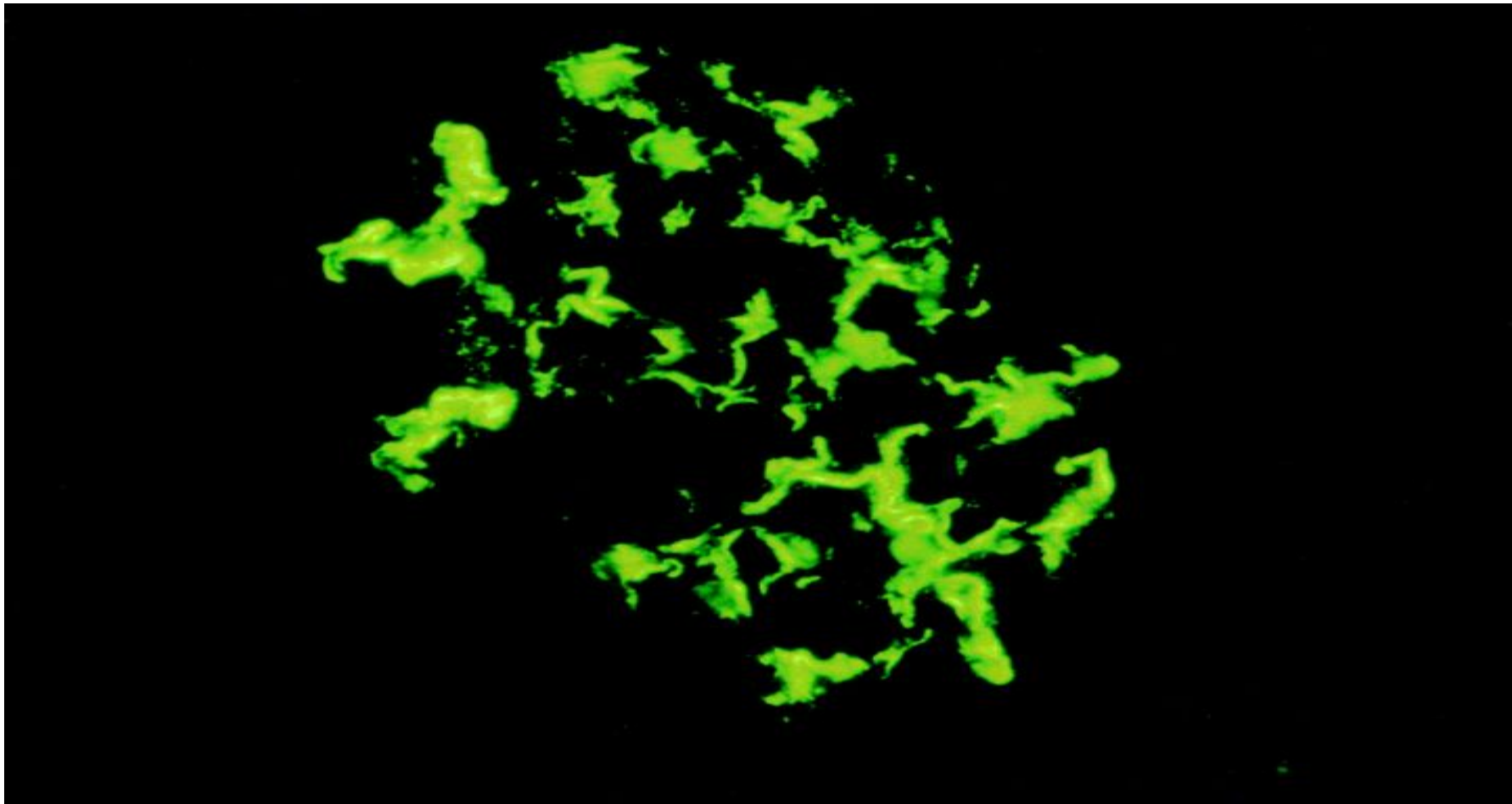
B: Urine sediment red blood cell cast;

C: Acute glomerular inflammation and crescent formation;

D: Details of basal membrane mesangial proliferation and IgA deposits



Immunofluorescence micrograph of a glomerulus from a patient with HSP nephropathy stained for the presence of IgA.



Differential Diagnosis of Henoch-Schönlein Purpura

- **Acute abdomen**
- **Meningococcal meningitis or septicemia**
- **Rheumatoid arthritis**
- **Rheumatic fever**
- **Idiopathic thrombocytopenic purpura**
- **Systemic lupus erythematosus**
- **poly-arteritis nodosa,**
- **Child abuse**
- **Drug reactions**
- **Bacterial endocarditis**
- **Rocky Mountain spotted fever**
- **familial Mediterranean fever**
- **inflammatory bowel disease.**
- **Kawasaki disease.**

Acute hemorrhagic edema (AHE)

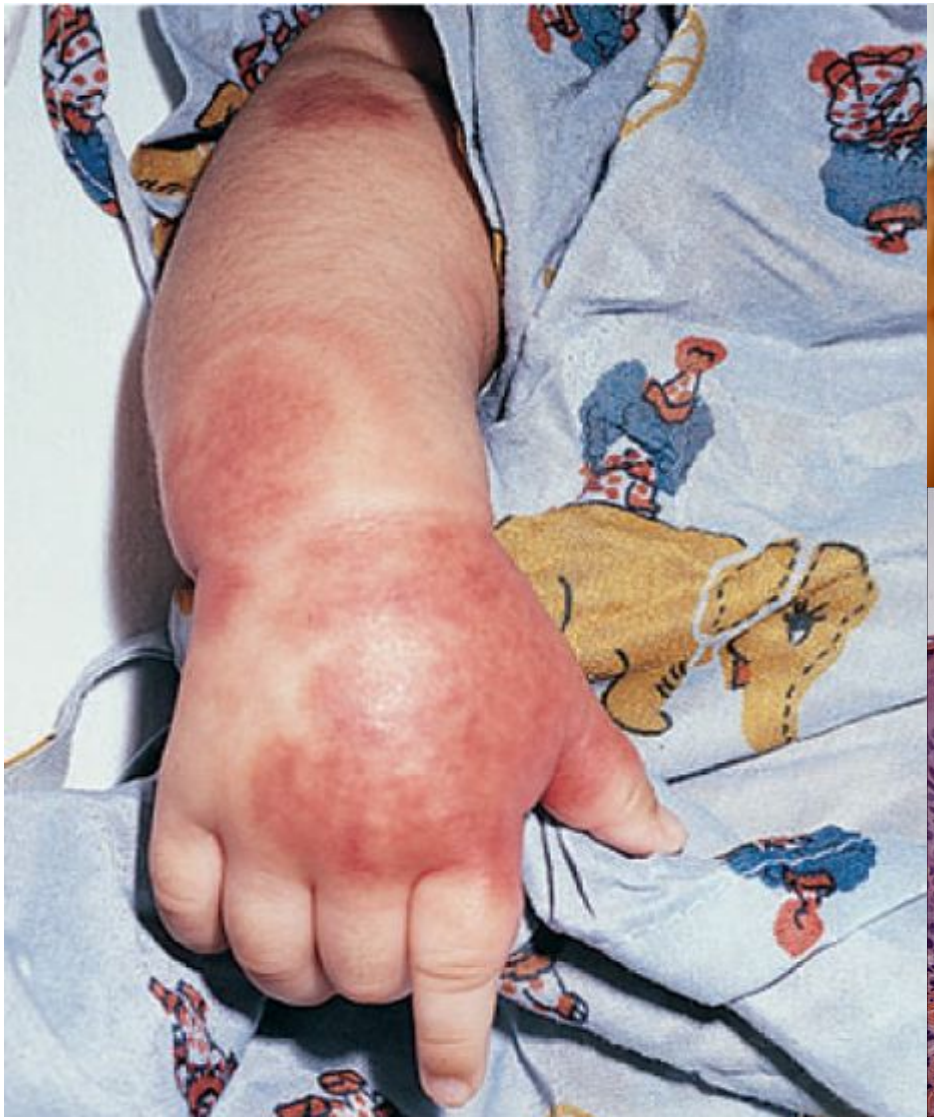
- is an acute **cutaneous benign leukocytoclastic vasculitis**
- seen in children ≤2 yr of age

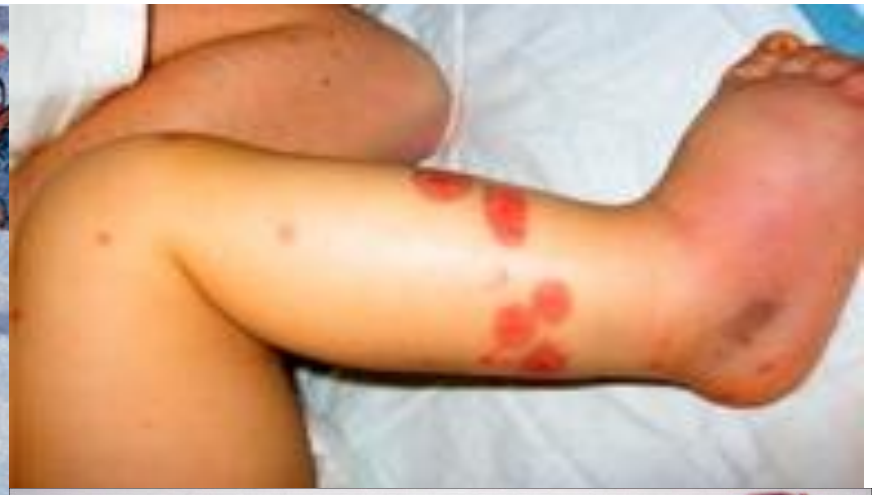
- AHE presents with fever; tender edema of the face, scrotum, hands, and feet; and ecchymosis (usually larger than the purpura of HSP) on the face and extremities

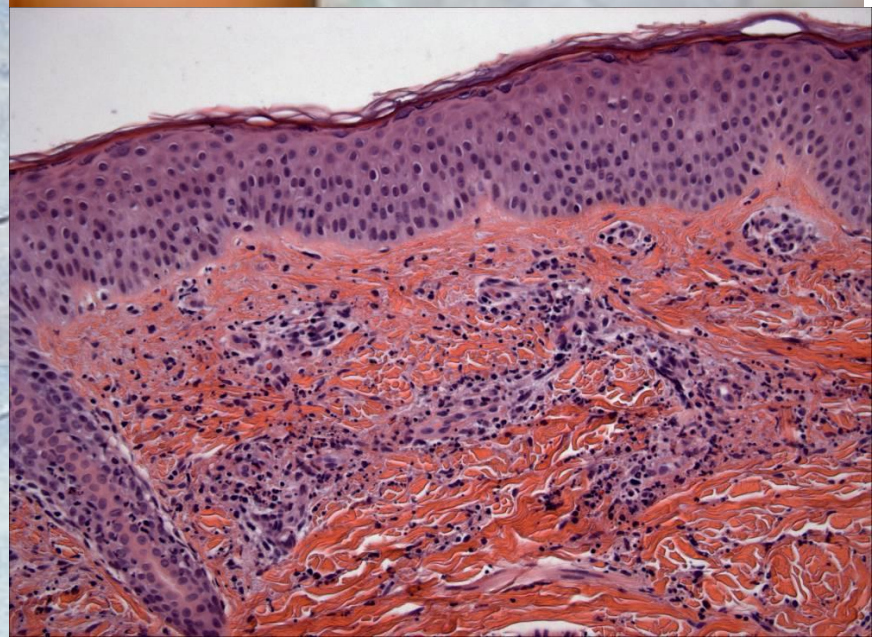
- **petechiae may be seen in mucous membranes**.
- The patient usually appears well except for the rash.
- The platelet count is normal or elevated;
- the urinalysis is normal.

The younger age, nature of the lesions, absence of other organ involvement, and biopsy may help distinguish AHE from HSP.

Acute hemorrhagic edema (AHE)







TREATMENT

Symptomatic treatment

- adequate hydration,
- bland diet,
- **pain control** with acetaminophen is provided for self-limited complaints of arthritis, edema, fever, and malaise.
- **Avoidance** of competitive **activities** and avoidance of maintaining the lower extremities in a dependent position may decrease local edema.
- If edema involves the scrotum, **elevation of the scrotum** and local cooling, as tolerated, may decrease discomfort.

TREATMENT

- Therapy with oral or intravenous **corticosteroids** (1-2 mg/kg/day) is often associated with dramatic improvement of both gastrointestinal and CNS complications.
- the effects of corticosteroids on renal manifestations are not clear.
- intussusception may be life-threatening and managed with cortico-steroids and, when necessary, hydrostatic reduction (by air or with contrast) or resection of the intussusception.

TREATMENT

- is the same as for other forms of acute glomerulonephritis
- If anti-cardiolipin or antiphospholipid antibodies are identified and thrombotic events have occurred, **aspirin** (81 mg) given once may decrease the risks associated with a hypercoagulable state.
- Rheumatoid nodules may respond to alternate-day **colchicine** (0.6 mg every other day).

Prognosis

- More than 80% of patients have a single isolated episode lasting a few weeks.
- Approximately 10-20% of patients have recurrences.
- Fewer than 5% of patients develop chronic HSP.
- Abdominal pain resolves spontaneously within 72 hours in most patients

Thank You!