Henoch Schonlein Purpura

Morning Report

February 17, 2005

Introduction

- Most common systemic vasculitic disease of childhood
- Annual incidence is 20:100,000 children
- Age varies from 6mo→adulthood; Peak incidence is 2-8 years
- Course of disease usually lasts 4-6 weeks
- Unknown etiology
- Often preceded by a respiratory infection
- Increased incidence in winter and spring
- Male:female ratio 2:1

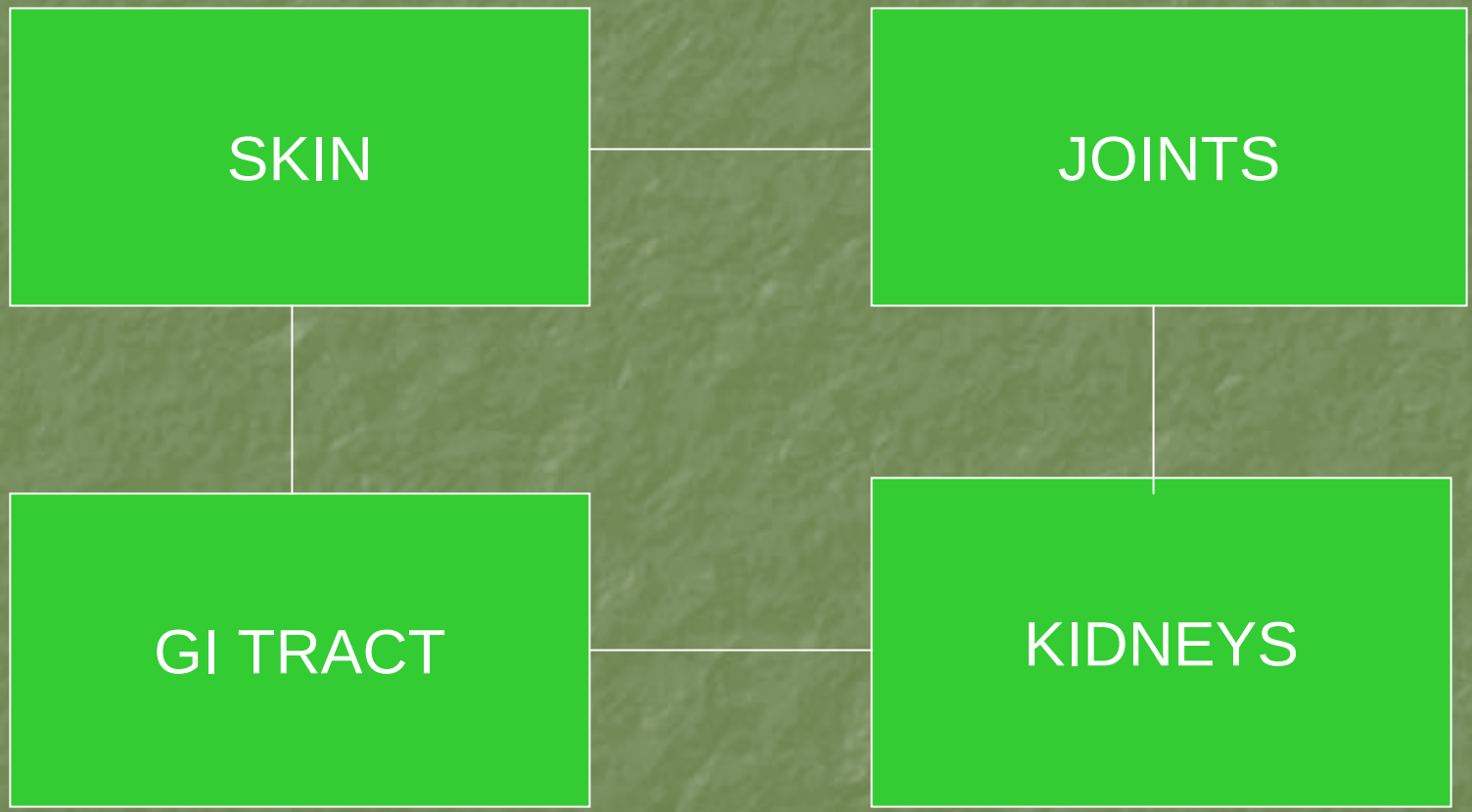
Diagnostic Criteria
American College of Rheumatology, 1990

- 2/4 criteria required for diagnosis of HSP:
  - 1. Palpable purpura without thrombocytopenia
  - 2. Age < 20yr at disease onset
  - 3. “Bowel angina”- diffuse abdominal pain, or bowel ischemia (bloody diarrhea)
  - 4. Biopsy evidence of granulocytes in the walls of arterioles or venules

Pathogenesis

- Poorly understood
- Infectious etiologies have been implicated but never confirmed
- **IgA mediated small vessel vasculitis**
- May result from a defect in the regulation of IgA synthesis in response to circulating or mucosal antigens
- Tissue deposition of IgA-containing immune complexes (+ IgG, C3) w/ secondary inflammatory response
  - Demonstrated in skin and renal biopsies
- IgA nephropathy has identical histologic findings in the kidney

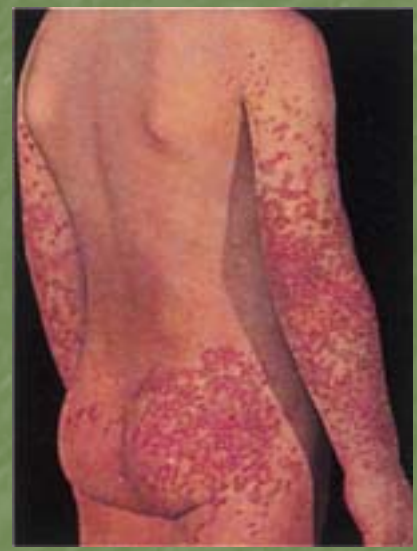
Clinical Manifestations



Symptoms can occur in any order and at any time usually over a period of several days to several weeks

Clinical Manifestations: *Skin*

- Classic rash: “palpable purpura”
- Begins as maculopapular rash→ petechiae & purpura
  - May appear urticarial at onset
- Symmetric, over extensor surfaces, buttocks
- Lesions occur in “crops” and last 3-10days
- May recur for 3-4 months
- Presenting sign in >50% of children
- **Normal platelets** & clotting studies



Clinical Manifestations: *Joints*

- Arthralgias/arthritis is present in 60-84% of cases
- Ankles and knees most common
- Prominent periarticular swelling/tenderness w/out erythema and warmth
- Effusion (if present) are serous (not hemorrhagic)
- Often most incapacitating symptom
- Symptoms are transient (resolves in a few days) w/ no permanent damage to joints

Clinical manifestations: *Renal*

- 30-70% of children will have hematuria (micro or macroscopic) with or without proteinuria
- Not predictably related to severity of extra-renal involvement
- Usually occurs few days to 1 month after onset of systemic symptoms
- Most disease is mild, but nephrotic syndrome, HTN, acute renal failure can occur
- Severity of renal impairment often correlates w/ findings on biopsy
  - Percentage of glomeruli w/ crescents may be the most important prognostic indicator

Clinical Manifestations: *GI Tract*

- GI tract involvement occurs in up to 75% of cases
- Classic presentation w/ “colicky” abdominal pain and vomiting due to local vasculitis
- Usually develops w/in 8 days of the rash (but may occurs weeks to months after rash or *before* the rash)
- Other manifestations include: intestinal hemorrhage w/ hematochezia, melena (up to 25%), occult blood (50%), intussusception, bowel performation, pancreatitis, gall bladder hydrops
- Purpuric lesions can be seen on endoscopy
- Increased risk of renal disease in pts with bloody stools

Intussusception & HSP

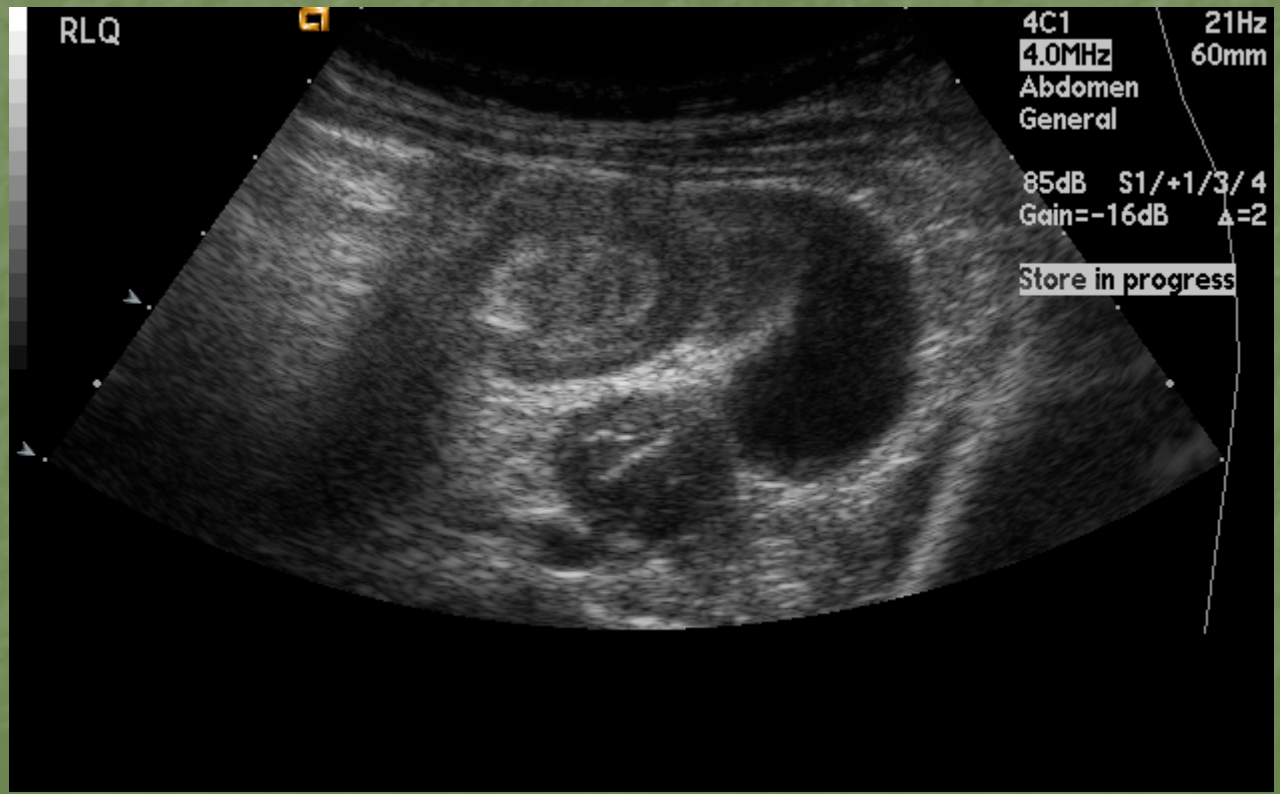
- Most frequent surgical complication of HSP
- Occurs in 1.3-13.6% of reported series
- The leadpoint is usually a **small bowel hematoma** secondary to intestinal hemorrhage and mucosal edema
- Most common types are **ileo-ileal** (51%) and ileo-colic (39%)
  - In contrast to idiopathic intussusception where 80-90% are ileo-colic
- Contrast enema reduction is often ineffective in >50% of intussusceptions in HSP b/c they are often confined to the small bowel
- Theory that underlying vasculitis of HSP makes bowel perforation more likely during contrast enema is not well supported

Intussusception & HSP

- Ultrasound is the most appropriate initial screening study in children w/ HSP and abdominal pain
  - Non-invasive, low cost
  - Helpful to follow patients serially
  - Contrast enema may be unable to reach a small bowel intussusception
  - However, contrast enema was not associated w/ complications in pts w/ HSP and intussusception

J. Schwab, et al. J Ped Surg; 2005;40

“Bull’s Eye” or “Target” Sign



Barium Enema

HSP without the P?

Fitzgerald, et al. J Ped Gastro Nutr 2000;30

- Many experts have reported cases of HSP without skin involvement
- Case reports have demonstrated late onset skin involvement (up to 18mo after dz) or no rash at all in pts presenting with predominantly GI symptoms c/w HSP
- Endoscopy and biopsy findings from small bowel mucosa were c/w HSP
- Skin biopsies from *normal appearing skin* showed IgA deposition c/w HSP

Clinical Manifestations: *Other*

- Common findings:
  - Local angioedema (dependent areas)- scrotum, eyelids, lips, over buttocks, hands/feet
  - Low grade fever
  - Fatigue
- Rare findings:
  - Cerebral hemorrhage
  - Pulmonary hemorrhage
  - Orbital hematomas, central artery thrombosis
  - Myocardial infarction
  - Priapism
  - Testicular torsion

HSP Emergencies

- Bowel perforation/Obstruction
- Renal failure
- Testicular torsion
- CNS involvement (cerebral hemorrhage)

Diagnosis

- **Clinical diagnosis**
- Laboratory findings non-specific
  - Thrombocytosis, leukocytosis, anemia common
  - Elevated IgA in 50% of patients
- Confirmation of diagnosis is accomplished by skin and/or renal biopsy
  - Skin biopsy demonstrates *leukocytoclastic vasculitis* and IgA deposition
  - Renal biopsy shows mesangial IgA deposits by immunofluorescence
    - Renal bx recommended w/ marked proteinuria and/or impaired renal function

Treatment

- **Supportive treatment** for uncomplicated disease
- Treatment recommended for patients w/ marked proteinuria, impaired renal function
  - Pulse IV methylprednisolone X 3 days, followed by oral prednisone X 3 mo
  - Early initiation of therapy may be important to prevent permanent injury
  - Other therapies used include azathioprine, cyclophosphamide, dipyridamole, plasmapheresis, IVIG
- Treatment also recommended for severe neurological involvement and considered for GI vasculitis w/ intestinal hemorrhage

Recurrent Disease

- Recurrences are common- **~1/3** of patients
- Usually mimic original episode but are less severe
- Typically occur within 4 months of resolution of the initial symptoms
- Recurrences are more likely in patients with nephritis and in older children

Prognosis

- Overall outcome is good w/ complete recovery in ~95% of patients
- Renal involvement is the most debilitating complication of HSP
  - Most pts w/ renal disease have excellent prognosis and resolution occurs w/in several months
  - ~1-5% develop persistent renal disease
    - Associated w/ nephrotic syndrome at diagnosis
  - <0.1% develop end stage disease