

A 6-year-old boy is brought to the physician by his mother with abdominal pain that began 4 days earlier. The pain is diffuse and associated with nausea. The boy has had no fever, vomiting, or changes in bowel habits. He had a rash that was initially erythematous and macular but has now become confluent. Physical examination shows a diffusely tender abdomen without rebound or guarding. A nonblanching rash is noted on his lower extremities. The patient's right knee is swollen and he complains of pain with passive range of motion. Laboratory results are as follows:

#### Complete blood count

Hemoglobin	13.5 g/dL
Platelets	350,000/ $\mu$ L
Leukocytes	9,100/ $\mu$ L
Neutrophils	68%
Eosinophils	1%
Lymphocytes	25%
Monocytes	6%

#### Serum chemistry

Sodium	140 mEq/L
Potassium	3.6 mEq/L
Chloride	98 mEq/L
Bicarbonate	23 mEq/L
Blood urea nitrogen	18 mg/dL
Creatinine	1.3 mg/dL
Calcium	9.2 mg/dL
Glucose	118 mg/dL

#### Urinalysis



### Serum chemistry

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### Urinalysis

Specific gravity	1.022
Protein	+1
Blood	Moderate
Glucose	Negative
Ketones	Negative
Leukocyte esterase	Negative
Nitrites	Negative

Which of the following glomerular abnormalities is most likely present in this patient?

- ☐ A. Glomerular basement membrane thickening
- ☐ B. Linear deposition of immunoglobulin G on the basement membrane
- ☐ C. Localized areas of mesangial sclerosis and collapse
- ☐ D. Mesangial deposition of immunoglobulin A
- ☐ E. Podocyte fusion

Submit



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#### Urinalysis



Glucose 118 mg/dL

Urinalysis

Specific gravity	1.022
Protein	+1
Blood	Moderate
Glucose	Negative
Ketones	Negative
Leukocyte esterase	Negative
Nitrites	Negative

Which of the following glomerular abnormalities is most likely present in this patient?

- ☐ A. Glomerular basement membrane thickening [5%]
- ☐ B. Linear deposition of immunoglobulin G on the basement membrane [11%]
- ☐ C. Localized areas of mesangial sclerosis and collapse [4%]
- ☒ D. Mesangial deposition of immunoglobulin A [72%]
- ☐ E. Podocyte fusion [8%]

Proceed to Next Item

Explanation:

User Id: [redacted]

Henoch-Schönlein purpura	
Pathogenesis	<ul style="list-style-type: none"><li>IgA-mediated leukocytoclastic vasculitis</li></ul>
Clinical manifestations	<ul style="list-style-type: none"><li>Palpable purpura</li><li>Arthritis/arthralgia</li><li>Abdominal pain, intussusceptions</li><li>Renal disease similar to IgA nephropathy</li></ul>



Explanation:

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Henoch-Schönlein purpura	
Pathogenesis	<ul style="list-style-type: none"> <li>• IgA-mediated leukocytoclastic vasculitis</li> </ul>
Clinical manifestations	<ul style="list-style-type: none"> <li>• Palpable purpura</li> <li>• Arthritis/arthralgia</li> <li>• Abdominal pain, intussusceptions</li> <li>• Renal disease similar to IgA nephropathy</li> </ul>
Laboratory findings	<ul style="list-style-type: none"> <li>• Normal platelet count &amp; coagulation studies</li> <li>• Normal to ↑ creatinine</li> <li>• Hematuria +/- RBC casts +/- proteinuria</li> </ul>
Treatment	<ul style="list-style-type: none"> <li>• Supportive (hydration &amp; NSAIDs) for most patients</li> <li>• Hospitalization &amp; systemic glucocorticoids in patients with severe symptoms</li> </ul>

RBC = red blood cell; NSAIDs = nonsteroidal antiinflammatory drugs.

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This child's clinical presentation of abdominal pain, **lower-extremity purpura**, arthritis, and hematuria is most consistent with Henoch-Schönlein purpura (HSP), an immunoglobulin A (IgA)-mediated vasculitis of the small vessels. The **rash** typically consists of **palpable purpura** and is distributed symmetrically over the lower legs, buttocks, and arms. **Arthralgias/arthritis** most commonly affect the knees and ankles. These symptoms are usually transient, and there is no permanent damage to the joints. **Colicky abdominal pain** (presumably due to local vasculitis) is present in the majority of patients.

**Renal involvement** occurs in approximately 20%-50% of children and may occur as late as 4-6 weeks after onset of the illness. Most patients have relatively mild disease characterized by microscopic or macroscopic hematuria, red cell casts, and mild-to-moderate proteinuria (typically non-nephrotic range) with a normal or only slightly elevated serum creatinine. However, more severe complications including nephrotic syndrome, hypertension, and acute renal failure, may occur. The diagnosis of HSP is made clinically in pediatric patients with a classic presentation. However, in children with atypical presentations, a renal biopsy may be helpful to confirm the diagnosis and will



patients with severe symptoms

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**(Choice A)** Membranous nephropathy is characterized by thickening of the basement membrane that affects all glomeruli. Membranous nephropathy is far more common in adults than children.

**(Choice B)** Goodpasture syndrome, which is caused by antibodies directed against the basement membrane, typically presents with pulmonary hemorrhage and glomerulonephritis. It is seen in both adolescents and adults. Linear deposition of immunoglobulin G on the basement membrane is seen in Goodpasture syndrome.

**(Choice C)** Focal segmental glomerulosclerosis is the most common cause of nephrotic syndrome in adults in the United States. It is characterized by localized regions of mesangial sclerosis and basement membrane collapse. As suggested by the name, not all glomeruli are affected in this disease.

**(Choice E)** Minimal change disease is the most common form of idiopathic nephrotic syndrome in children and often presents with edema and hematuria. Electron microscopy findings include fusion or flattening of the podocytes ("foot processes").

#### Educational objective:

Henoch-Schönlein purpura is an immunoglobulin A (IgA)-mediated vasculitis of the small



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#### **Educational objective:**

Henoch-Schönlein purpura is an immunoglobulin A (IgA)-mediated vasculitis of the small vessels that is most common in children. Classic manifestations include **palpable purpura** on the lower extremities, **arthralgias**, **abdominal pain**, and **renal disease**. Immunofluorescence microscopy shows IgA deposition in the kidney.

#### **References:**

1. [Henoch Sch?nlein purpura in childhood: epidemiological and clinical analysis of 150 cases over a 5-year period and review of literature.](#)
2. [Clinical course of extrarenal symptoms in Henoch-Sch?nlein purpura: a 6-month prospective study.](#)



Media Exhibit

h-Schonlein purpura

