IgA Vasculitis (Formerly Henoch-Schonlein Purpura)

Henoch-Schonlein purpura (HSP) is a systemic vasculitis characterized by the deposition of IgA immune complexes in the skin and other organs. This condition is the most common childhood vasculitis and typically presents after pharyngeal and upper respiratory infections. The skin, joints, and GI are the most commonly involved organs. In the skin, immune complex deposition causes palpable purpura, which are small red or purple discolorations of the skin and are seen in all cases of HSP. Arthralgia and abdominal pain are also typically seen. Involvement of the GI tract can cause gastrointestinal hemorrhage, which causes black stools. Forty percent of patients also have evidence of renal involvement with hematuria. The pathophysiology of HSP is similar to IgA nephropathy, including high serum levels of IgA and similar findings on renal biopsy. However, IgA nephropathy typically only involves the kidneys and has a predilection for young adults, while HSP is a systemic disease and involves other organs. Overall, the prognosis is good in most patients, with high rates of recovery. In children under ten years of age, the condition recurs in about one-third of the cases.

PLAY PICMONIC

Epidemiology

Most Common Childhood Vasculitis
Child with #1 Foam-finger Showing Vessels-on-fire
IgA vasculitis most commonly affects children and is the most common vasculitis seen in the pediatric population.

Etiology & Pathophysiology

Post-URI Immune Response
Post-Upper Respiratory Tract Bacteria
IgA vasculitis often follows an upper respiratory or pharyngeal infection, as immune complex formation is typically triggered by a preceding infection, most commonly streptococcal.

IgA Immune Complex Deposition
Apple-goblin In-moon Antibody Complex
The accumulation of circulating immunoglobulin A (IgA) immune complexes within small vessel walls, particularly in the skin, kidneys, and gastrointestinal tract. These complexes trigger complement activation and neutrophil recruitment, leading to inflammation, vessel damage, and tissue injury characteristic of IgA vasculitis

Type III Hypersensitivity Reaction
Hiker-sensitive-crying on (3) Tree
IgA vasculitis (Henoch-Schönlein Purpura) is a Type III hypersensitivity reaction in which galactose-deficient IgA1 forms circulating immune complexes that deposit in small vessel walls of the skin, kidneys, and gastrointestinal tract. This deposition activates complement and recruits neutrophils, producing leukocytoclastic vasculitis and tissue injury. The resulting immune complex–mediated inflammation causes the characteristic palpable purpura, arthralgia, abdominal pain, and renal involvement. Experimental models reproduce the disease using Type III hypersensitivity mechanisms, confirming that IgA1 immune complex deposition and subsequent inflammatory cascades are the central drivers of IgA vasculitis.

Clinical Features

Arthralgia
King Arthur-algae
Arthralgia refers to joint pain caused by a non-inflammatory condition. Arthralgia is seen in IgA vasculitis due to joint involvement.

Abdominal Pain
Abdominal Pain-bolts
Colicky abdominal pain caused by IgA-mediated small vessel vasculitis of the gastrointestinal tract, leading to bowel wall edema, possible bleeding, and risk of intussusception

GI Bleeding / Intussusception

GI-guy Bleeding and Intestine-telescope

Gastrointestinal bleeding results from IgA-mediated small vessel vasculitis, causing mucosal hemorrhage, while bowel wall edema can act as a lead point for intussusception, presenting with severe abdominal pain and possible “currant jelly” stools.

Renal Involvement (Hematuria and Proteinuria)

Kidney

IgA immune complex deposition in the mesangium leads to glomerulonephritis, resulting in hematuria, often with RBC casts, and mild to moderate proteinuria due to glomerular inflammation; this represents the most important complication and main determinant of long-term prognosis, typically occurring days to weeks after the rash, with pathology similar to IgA nephropathy, possible progression to nephritic syndrome, and usually normal complement levels, requiring close follow-up with serial urinalysis and renal function monitoring.

Black Stool (Melena)

Black Intestine-stool

Black, tarry stool is a sign of intestinal hemorrhage, usually in the upper part of the gastrointestinal tract.

Palpable Purpura

Palpable-paw-print from Purple-cat

Lesions appear as red or purple discolorations on the skin that do not blanch with pressure. Palpable purpura results from the recruitment and activation of neutrophils within the small vessels of the skin, leading to vessel inflammation and leakage of blood into surrounding tissues. It is a hallmark of small-vessel vasculitis, most notably IgA vasculitis. The rash typically involves the buttocks and lower extremities, but may also appear on the face, arms, or trunk.

Management & Prognosis

Self-Limiting Course

Self-closing

IgA vasculitis is usually a self-limited small vessel vasculitis in which symptoms such as palpable purpura, abdominal pain, and arthralgia resolve spontaneously within a few weeks, especially in children, although recurrence can occur, and renal involvement requires ongoing monitoring.

Supportive Care

Supportive IV bags

Management is primarily supportive, including hydration, pain control, and rest, as the disease is typically self-limited; NSAIDs may be used for arthralgia, while more severe symptoms such as significant abdominal or renal involvement may require corticosteroids.