

Vasculitis

The body's defense army turns on its own blood vessel walls. Sort by vessel size (big, medium, small). Check the ANCA blood test to narrow the type. Calm the immune attack with steroids and other immune-quieting drugs before organs lose their blood supply.

THE ONE-LINE TRUTH

The immune system mistakenly attacks the walls of blood vessels, causing swelling, blockage, and bleeding that can damage any organ those vessels feed.

The chain — how it unfolds

1. Immune cells attack vessel walls everywhere. — why: In vasculitis, the immune system makes antibodies (like ANCA—antineutrophil cytoplasmic antibodies that tag white blood cells to attack vessels) or activates T-cells (the immune system's attack squad) that mistake vessel-wall proteins for foreign invaders. White blood cells swarm into the wall, release inflammatory chemicals (cytokines like TNF-alpha and interleukin-1, plus reactive oxygen species that burn tissue), and damage the endothelium (slick inner lining) and muscle layers.
2. Fever and weight loss appear first. — why: The widespread immune battle raises the body's thermostat (the hypothalamus in the brain) to create fever, which is meant to slow germs but here just signals inflammation. The fight also burns extra calories and suppresses appetite (cytokines like IL-1 and TNF-alpha turn down hunger signals), so weight drops.
3. Red or purple spots (purpura) appear on skin. — why: Small skin vessels leak red blood cells when their walls break from immune attack; the leaked blood shows as tiny red or purple dots (petechiae) or larger bruise-like patches (purpura) that do not blanch (turn white) when you press them.
4. Blood appears in urine (hematuria). — why: In the kidney, inflammation punches holes in the glomerulus (the tiny filter ball made of capillaries), so red blood cells spill through into the urine. Protein leaks too (proteinuria), making urine foamy.
5. Coughing up blood (hemoptysis) signals lung involvement. — why: Small vessels in the lung's air sacs (alveoli—the tiny balloon-like pockets where oxygen enters the blood) rupture when vasculitis hits them; blood leaks into the airway and is coughed out. This is common in granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA).

6. Numbness, tingling, or weakness in hands and feet (mononeuritis multiplex). — why: Vasculitis blocks the tiny arteries (vasa nervorum—the little supply vessels that feed peripheral nerves). Starved of oxygen, the nerve fibers malfunction—first tingling, then numbness, then weakness. Multiple separate nerves fail in a patchy pattern (not symmetric like diabetic neuropathy).
7. Severe belly pain and bloody stools. — why: When vasculitis clogs the arteries feeding the intestines (mesenteric vessels), sections of bowel lose their blood supply. The gut lining dies (ischemia, then tissue death from blocked blood flow (infarction)—tissue death), causing intense pain, bleeding into the stool, and risk of perforation (a hole that spills gut contents into the belly, leading to life-threatening infection).
8. Sudden vision loss or severe headache in giant cell arteritis. — why: Giant cell arteritis inflames the temporal artery (the vessel you can feel pulsing at your temple) and branches that feed the optic nerve (the cable carrying signals from the eye to the brain). If the optic artery clogs, the nerve dies within hours and blindness is permanent. Jaw pain when chewing (jaw claudication—the jaw muscles cramping because their blood supply is choked) is another clue that the chewing-muscle arteries are starved.

Spot it — what you see, and why

- Fever that won't go away: The inflamed blood vessel walls release chemicals called cytokines (alarm signals) that travel to the hypothalamus (the brain's thermostat) and reset your body temperature higher, like turning up a heater to fight what looks like an infection.
- Weight loss you didn't try for: Ongoing inflammation burns extra energy — like leaving a car engine running all the time — and the cytokines also kill your appetite, so you eat less and your body starts using its own stores.
- Purple or red spots on the skin (purpura): Tiny blood vessels in the skin get inflamed and leak red blood cells into the surrounding tissue, like a garden hose with holes spraying water into the dirt, leaving behind purple or red marks.
- Numbness, tingling, or weakness in hands or feet: The small arteries feeding the nerves in your arms and legs get inflamed and narrow, starving the nerves of oxygen and nutrients — like a pinched phone charger that stops electricity flowing — so the nerves can't send signals properly.
- Blood in the urine (hematuria): Inflammation hits the tiny blood vessels in the kidney's filters (glomeruli), making them leaky so red blood cells spill into the urine, turning it pink, red, or cola-colored.
- Jaw pain when chewing (jaw claudication): In giant cell arteritis, the arteries feeding the jaw muscles get inflamed and narrowed, so when you chew and the muscles need more oxygen, they don't get enough — like a cramp from running with a blocked airway — and they ache or tire quickly.

Red flags — act now

- Sudden vision loss or new severe headache in someone over 50 (possible giant cell arteritis — needs urgent steroids to save sight)
- Blood in the urine plus swelling or high blood pressure (kidney damage can spiral quickly)
- Coughing up blood or fast-worsening shortness of breath (lungs may be bleeding or severely inflamed)
- Numbness, weakness, or stroke-like symptoms (brain blood vessels may be involved)

Treat it — in this order

1. High-dose corticosteroids (e.g., prednisone 1 mg/kg/day, usually 60–80 mg daily, or IV methylprednisolone 500–1000 mg daily for severe organ-threatening disease) → Steroids shut down the immune cells that are releasing inflammatory signals and attacking vessel walls. This stops new inflammation quickly (within hours to days) and allows damaged vessels to begin healing. In giant cell arteritis, high-dose steroids prevent permanent vision loss. In ANCA vasculitis, they halt kidney and lung destruction.
2. Cyclophosphamide (IV or oral) or rituximab (anti-CD20 monoclonal antibody) for severe ANCA-associated vasculitis (GPA, MPA, EGPA with organ threat) → These drugs knock down or wipe out the B cells and other immune cells making the antibodies (ANCA) that attack vessel walls. Rituximab depletes CD20+ B cells; cyclophosphamide kills rapidly dividing immune cells. Both induce remission (stop active disease) and work deeper and longer than steroids alone, preventing relapse. Current guidelines favor rituximab over cyclophosphamide for induction in most cases because it has similar efficacy with a better long-term safety profile.
3. Plasma exchange (plasmapheresis, PLEX) for severe pulmonary bleeding (hemorrhage) or rapidly progressive glomerulonephritis with dialysis-level kidney failure → PLEX physically removes circulating ANCA antibodies and inflammatory proteins from the blood right away, before steroids and immunosuppressants have time to work. This can rescue kidney function and stop lung bleeding in the most severe cases. Some studies show benefit when creatinine is very high (e.g., > 5.7 mg/dL) or the patient needs dialysis at presentation.
4. Low-dose aspirin (75–100 mg daily) for giant cell arteritis → Aspirin inhibits platelet clumping, making blood less likely to clot inside inflamed arteries. In giant cell arteritis, inflamed vessel walls can trigger clots that block the ophthalmic artery (causing blindness) or cerebral arteries (causing stroke). Aspirin reduces this blood-starved (ischemic) complication risk.

5. Tocilizumab (IL-6 receptor blocker, subcutaneous injection weekly or every other week) for giant cell arteritis → Tocilizumab blocks interleukin-6, a key inflammatory signal that drives giant cell arteritis. Adding it to steroids lets patients taper steroids faster and stay in remission longer with lower cumulative steroid doses, reducing steroid side effects like osteoporosis, diabetes, and weight gain. FDA-approved for giant cell arteritis based on the GiACTA trial.

Your top priorities at the bedside

1. Check vision in each eye separately every 4 hours and report any new blurry vision, blind spots, or vision loss to the provider immediately — why: Giant cell arteritis can cause anterior blood-starved (ischemic) optic neuropathy (blood flow loss to the optic nerve), leading to permanent blindness within hours if steroids are delayed
2. Monitor oxygen saturation continuously and listen to lung sounds every 2 hours; report oxygen saturation below 92 percent or new crackles — why: Pulmonary bleeding (hemorrhage) (lung bleeding) from ANCA-associated vasculitis can progress rapidly to respiratory failure and death; early detection allows time for plasma exchange and mechanical ventilation if needed
3. Measure urine output every hour and check urine color; report output less than 0.5 mL per kilogram per hour or dark urine — why: Rapidly progressive glomerulonephritis (fast kidney inflammation) in vasculitis can destroy nephrons (kidney filtering units) within days, causing permanent dialysis dependence if immunosuppression is not started quickl