Vasculitis in the Hospital Setting

Vasculitis refers to a group of disorders characterized by inflammation of blood vessels, leading to tissue ischemia and organ dysfunction. This guide provides physician assistant (PA) students with a comprehensive framework to understand the clinical presentation, diagnostic approach, types, treatment, complications, and prognosis of vasculitis in the hospital setting, with case scenarios to apply the knowledge.

Introduction and Pathophysiology

- Vasculitis is caused by immune-mediated inflammation of blood vessel walls, resulting in vessel damage, occlusion, or aneurysm formation. The pathophysiology involves:
- **Immune Dysregulation:** Autoimmune activation (e.g., autoantibodies like ANCA in small vessel vasculitis) or immune complex deposition (e.g., in cryoglobulinemic vasculitis) triggers inflammation.
- **Vessel Wall Injury:** Inflammatory cells (e.g., neutrophils, lymphocytes) infiltrate vessel walls, releasing cytokines (e.g., TNF-α, IL-1) and causing endothelial damage.
- **Ischemia and Necrosis:** Vessel narrowing or occlusion leads to downstream tissue ischemia; aneurysm formation can cause rupture or bleeding.
- **Systemic Effects:** Inflammation and organ dysfunction (e.g., glomerulonephritis, pulmonary hemorrhage) result from widespread vessel involvement.
- Vasculitis can affect vessels of any size (small, medium, large) and often presents as a multisystem disease requiring urgent hospital management.

Clinical Presentation

General Symptoms:

Constitutional: Fever, weight loss, fatigue, night sweats.

Musculoskeletal: Myalgias, arthralgias, arthritis.

Organ-Specific Symptoms:

- **Skin:** Palpable purpura (small vessel), livedo reticularis, ulcers, nodules.

- **Lungs:** Cough, hemoptysis, dyspnea (e.g., pulmonary hemorrhage in ANCA vasculitis).
- **Kidneys:** Hematuria, proteinuria, acute kidney injury (AKI) (e.g., glomerulonephritis in GPA).
- **Nervous System:** Peripheral neuropathy (mononeuritis multiplex), stroke, seizures (CNS vasculitis).
- **GI:** Abdominal pain, GI bleeding (mesenteric vasculitis).
- **ENT:** Sinusitis, nasal crusting, epistaxis, hearing loss (e.g., GPA, EGPA).

When to Suspect Vasculitis

- **Multisystem Involvement:** Unexplained symptoms affecting ≥2 organ systems (e.g., rash + AKI, hemoptysis + neuropathy).
- **Constitutional Symptoms with Organ Dysfunction:** Fever, weight loss with elevated inflammatory markers (e.g., ESR, CRP) and organ-specific findings.
- **Palpable Purpura or Rash:** Suggests small vessel vasculitis (e.g., HSP, cryoglobulinemia).
- **Rapidly Progressive Organ Failure:** AKI (glomerulonephritis), pulmonary hemorrhage, or mononeuritis multiplex.
- **Risk Factors:** Hepatitis B/C (PAN, cryoglobulinemia), autoimmune diseases (e.g., RA, SLE), drug exposure (e.g., hydralazine-induced ANCA vasculitis).

Labs and Diagnostic Studies

Initial Labs:

- **CBC:** Anemia (chronic disease, hemorrhage), leukocytosis/leukopenia, eosinophilia (EGPA).
- **Inflammatory Markers:** ESR, CRP elevated in active disease.
- **Renal Function:** Elevated BUN/Cr, hematuria, proteinuria (glomerulonephritis).
- **Liver Function Tests (LFTs):** Elevated in hepatitis-associated vasculitis (e.g., HBV-PAN).
- **Urinalysis:** Red blood cell (RBC) casts, hematuria (renal vasculitis).

Specific Tests:

- **ANCA (Anti-Neutrophil Cytoplasmic Antibodies):** c-ANCA (PR3, GPA), p-ANCA (MPO, MPA, EGPA).
- **ANA, Anti-dsDNA, Complement (C3/C4):** Low complement in SLE-associated vasculitis, cryoglobulinemia.

- **Cryoglobulins:** Positive in cryoglobulinemic vasculitis (often HCV-related).
- **Hepatitis Serologies:** HBV (PAN), HCV (cryoglobulinemia).
- **Blood Cultures:** Rule out infection (e.g., endocarditis mimicking vasculitis).

Diagnostic Studies:

- **Biopsy:** Gold standard; site depends on presentation (e.g., skin, kidney, lung, nerve).
  - **Small vessel:** Leukocytoclastic vasculitis (HSP), granulomas (GPA, EGPA).
  - **Medium vessel:** Necrotizing vasculitis without granulomas (PAN, Kawasaki).
- **Imaging:**
  - **CT/MRI:** Assess organ involvement (e.g., CT chest for pulmonary hemorrhage, MRI brain for CNS vasculitis).
  - **Angiography:** For medium/large vessel vasculitis; aneurysms (PAN), stenoses (Takayasu).
  - **Chest X-Ray/CT:** Cavitary lesions, nodules (GPA), infiltrates (EGPA).

Other:

Urine Drug Screen: Rule out cocaine (mimics vasculitis with nasal destruction).

Electromyography (EMG)/Nerve Conduction Studies: For mononeuritis multiplex.

Different Types of Vasculitis and Their Manifestations

Small Vessel Vasculitis:

- Granulomatosis with Polyangiitis (GPA):
  - **Manifestations:** Upper respiratory (sinusitis, epistaxis), pulmonary (hemoptysis, nodules), renal (glomerulonephritis), c-ANCA/PR3 positive.
- Microscopic Polyangiitis (MPA):
  - **Manifestations:** Renal (glomerulonephritis), pulmonary (hemorrhage), peripheral neuropathy, p-ANCA/MPO positive.
- Eosinophilic Granulomatosis with Polyangiitis (EGPA):
  - **Manifestations:** Asthma, eosinophilia, sinusitis, neuropathy, pulmonary infiltrates, p-ANCA/MPO positive.

- Henoch-Schönlein Purpura (HSP):
  - **Manifestations:** Palpable purpura (lower extremities), arthralgias, abdominal pain, renal (IgA nephropathy), IgA deposition.
- Cryoglobulinemic Vasculitis:
  - **Manifestations:** Purpura, arthralgias, neuropathy, glomerulonephritis, often HCV-related, cryoglobulins positive.

Medium Vessel Vasculitis:

- Polyarteritis Nodosa (PAN):
  - **Manifestations:** Mononeuritis multiplex, skin (nodules, ulcers), GI (mesenteric ischemia), renal (aneurysms), often HBV-related.
- Kawasaki Disease:
  - **Manifestations:** Fever, rash, conjunctivitis, strawberry tongue, coronary artery aneurysms (children).

Large Vessel Vasculitis:

- Takayasu Arteritis:
  - **Manifestations:** Pulselessness, hypertension (aortic involvement), stroke, young women, aortic arch stenosis.
- Giant Cell Arteritis (GCA):
  - **Manifestations:** Headache, jaw claudication, vision loss (temporal artery), polymyalgia rheumatica, elderly patients.

Treatment

General Principles:

Immunosuppression: Mainstay of therapy to halt vessel inflammation.

Supportive Care: Address organ-specific complications (e.g., dialysis for AKI, oxygen for hypoxia).

Infectious Disease Prophylaxis: TMP-SMX (PCP prophylaxis) with high-dose steroids/cyclophosphamide.

Small Vessel Vasculitis:

- GPA, MPA, EGPA:
  - **Induction:** High-dose steroids (methylprednisolone 500-1000 mg IV daily x 3 days, then prednisone 1 mg/kg/day PO) + cyclophosphamide 2 mg/kg/day PO or 15 mg/kg IV q2-3 weeks.

- **Maintenance:** Azathioprine 2 mg/kg/day PO or rituximab 375 mg/m^2 IV weekly x 4 doses; taper steroids over 6-12 months.
- **Severe Cases:** Plasma exchange for pulmonary hemorrhage, rapidly progressive glomerulonephritis (RPGN).

- HSP:
  - **Supportive:** Often self-limiting; NSAIDs for arthralgias.
  - **Severe (Renal/GI):** Prednisone 1 mg/kg/day PO x 4-6 weeks.
- Cryoglobulinemic Vasculitis:
  - **Treat HCV:** Direct-acting antivirals (e.g., sofosbuvir/ledipasvir).
  - **Severe:** Rituximab 375 mg/m^2 IV weekly x 4 doses, steroids for organ-threatening disease.

Medium Vessel Vasculitis:

- PAN:
  - **Induction:** Prednisone 1 mg/kg/day PO + cyclophosphamide (as above).
  - **Maintenance:** Azathioprine or methotrexate 15-20 mg PO weekly.
  - **HBV-Related:** Antivirals (e.g., entecavir), avoid cyclophosphamide if possible.
- Kawasaki:
  - **Acute:** IVIG 2 g/kg IV over 10-12 hours + aspirin 80-100 mg/kg/day PO; steroids for refractory cases.

Large Vessel Vasculitis:

- Takayasu Arteritis:
  - **Induction:** Prednisone 1 mg/kg/day PO x 1-3 months, taper slowly.
  - **Maintenance:** Methotrexate 15-20 mg PO weekly or tocilizumab (IL-6 inhibitor) 162 mg SC weekly.
- GCA:
  - **Induction:** Prednisone 40-60 mg PO daily (vision loss: methylprednisolone 1 g IV daily x 3 days).
  - **Maintenance:** Taper over 1-2 years; tocilizumab for steroid-sparing.

Hospital Management of Vasculitis Complications

Sick vasculitis patients often present with acute organ-threatening complications requiring hospital management.

Pulmonary Hemorrhage (e.g., GPA, MPA):

- **Supportive:** Oxygen, intubation if massive hemoptysis or respiratory failure (low tidal volumes 6 mL/kg).
- **Immunosuppression:** Methylprednisolone 1 g IV daily x 3 days, cyclophosphamide 15 mg/kg IV.
- **Plasma Exchange:** 7-10 sessions over 2 weeks for diffuse alveolar hemorrhage.
- **Monitoring:** Daily chest X-ray/CT, hemoglobin (bleeding), ABG for oxygenation.

Rapidly Progressive Glomerulonephritis (RPGN):

- **Supportive:** Dialysis if AKI (e.g., Cr >5 mg/dL, hyperkalemia); fluids for volume management.
- **Immunosuppression:** Methylprednisolone 1 g IV daily x 3 days, cyclophosphamide 15 mg/kg IV.
- **Plasma Exchange:** For severe renal involvement (e.g., dialysis-dependent).
- **Monitoring:** Daily Cr, urine output, urinalysis (RBC casts resolution).

Mononeuritis Multiplex:

- **Immunosuppression:** Prednisone 1 mg/kg/day PO, cyclophosphamide for severe cases.
- **Supportive:** Pain control (e.g., gabapentin 300 mg PO TID for neuropathic pain), physical therapy.

Sepsis (Steroid/Cyclophosphamide Risk):

- **Antibiotics:** Broad-spectrum (e.g., piperacillin-tazobactam 4.5 g IV q6h + vancomycin 15 mg/kg IV q12h) if infection suspected.
- **Supportive:** Fluids (30 mL/kg), vasopressors (e.g., norepinephrine) for septic shock.
- **Monitoring:** Blood cultures, lactate, adjust immunosuppression (e.g., hold cyclophosphamide).

GI Involvement (e.g., Mesenteric Vasculitis):

- **Supportive:** NPO, IV fluids, monitor for ischemia (e.g., lactate, CT for pneumatosis).
- **Immunosuppression:** Prednisone 1 mg/kg/day IV, cyclophosphamide if severe.

- **Surgery:** Urgent laparotomy if perforation or infarction (rare but high mortality).

Complications

Organ Damage:

- **Renal:** End-stage renal disease (ESRD) from glomerulonephritis (GPA, MPA).
- **Pulmonary:** Chronic respiratory failure, pulmonary hypertension (PH) from recurrent hemorrhage.
- **Neurological:** Stroke, permanent neuropathy (PAN, EGPA).

Treatment-Related:

- **Infections:** High-dose steroids/cyclophosphamide increase risk (e.g., PCP, sepsis).
- **Steroid Side Effects:** Osteoporosis, diabetes, Cushingoid features.
- **Cyclophosphamide:** Hemorrhagic cystitis, infertility, malignancy (e.g., bladder cancer).

Vascular:

- **Aneurysms (PAN, Takayasu):** Risk of rupture, bleeding.
- **Thrombosis:** Vessel occlusion causing ischemia (e.g., stroke, mesenteric ischemia).

Prognosis

- **GPA, MPA:** 5-year survival 70-80% with treatment; poor prognosis if untreated (90% mortality within 1 year).
- **EGPA:** Better prognosis (5-year survival >85%); cardiac involvement worsens outcomes.
- **HSP:** Self-limiting in children (90% resolve); adults may have chronic kidney disease (10-20%).
- **PAN:** 5-year survival 80% with treatment; HBV-related PAN worse if untreated.
- **Takayasu:** 5-year survival >90%; risk of aortic complications (e.g., dissection).
- **GCA:** Good prognosis with treatment; vision loss (15-20%) if untreated.

Key Pearls

- **Presentation:** Multisystem (rash, AKI, hemoptysis); constitutional symptoms (fever, weight loss).
- **When to Suspect:** ≥2 organ involvement, palpable purpura, RPGN, elevated ESR/CRP.
- **Diagnosis:** ANCA, biopsy (leukocytoclastic vasculitis, granulomas), imaging (CT, angiography).
- **Types:** Small (GPA, MPA, EGPA), medium (PAN, Kawasaki), large (Takayasu, GCA).
- **Treatment:** Steroids + cyclophosphamide (induction), azathioprine/rituximab (maintenance); plasma exchange for severe cases.
- **Hospital Complications:** Pulmonary hemorrhage (plasma exchange), RPGN (dialysis), sepsis (antibiotics, adjust immunosuppression).
- **Prognosis:** Variable; better with treatment, poor if untreated or severe organ involvement.

References

UpToDate: “Vasculitis: Diagnosis and Management” (2025). UpToDate Vasculitis

ACR: “Guidelines for the Management of Vasculitis” (2024). ACR Guidelines

EULAR: “Recommendations for ANCA-Associated Vasculitis” (2023). EULAR Guidelines

NEJM: “Advances in the Treatment of Giant Cell Arteritis” (2024). NEJM GCA

Case Scenarios

Case 1: A 45-Year-Old Male with Hemoptysis and AKI

Presentation: A 45-year-old male presents with 2 weeks of cough, hemoptysis, and fatigue, now with decreased urine output. Exam shows T 38°C, BP 140/90 mmHg, RR 24/min, SpO2 92% on room air, crackles in lungs, 1+ edema.

Labs/Imaging: Cr 3.5 mg/dL (baseline 1.0), urinalysis: RBC casts, hematuria. c-ANCA/PR3 positive. CT chest: Bilateral nodules, cavitary lesions.

Diagnosis: Granulomatosis with Polyangiitis (GPA) → Hemoptysis, AKI, c-ANCA, cavitary lesions.

Management: Admit to ICU. Start methylprednisolone 1 g IV daily x 3 days, then prednisone 1 mg/kg/day PO. Cyclophosphamide 15 mg/kg IV. Initiate plasma exchange (7 sessions) for pulmonary hemorrhage. TMP-SMX 1 DS tab PO daily (PCP prophylaxis). Monitor Cr (improves to 2.0 mg/dL), chest X-ray (nodules resolving). Transition to rituximab maintenance. Discharge with rheumatology follow-up.

Case 2: A 62-Year-Old Female with Vision Loss

Presentation: A 62-year-old female presents with 1 week of headache, jaw claudication, and sudden vision loss in the right eye. Exam shows T 37.5°C, tender right temporal artery, visual acuity 20/200 right eye, 20/30 left eye.

Labs/Imaging: ESR 80 mm/h, CRP 50 mg/L. Temporal artery biopsy: Giant cell arteritis.

Diagnosis: Giant Cell Arteritis (GCA) → Headache, vision loss, elevated ESR, biopsy confirmation.

Management: Admit for urgent treatment. Start methylprednisolone 1 g IV daily x 3 days (vision loss), then prednisone 60 mg PO daily. Aspirin 81 mg PO daily (reduce ischemic risk). Monitor vision (no further loss), ESR/CRP (decreasing). Taper prednisone over 1-2 years. Add tocilizumab 162 mg SC weekly at 3 months (steroid-sparing). Discharge with rheumatology follow-up.

Case 3: A 50-Year-Old Male with Abdominal Pain and Neuropathy

Presentation: A 50-year-old male with hepatitis B presents with 3 weeks of abdominal pain, weight loss, and foot drop. Exam shows T 38°C, BP 150/90 mmHg, diffuse abdominal tenderness, livedo reticularis, right foot weakness.

Labs/Imaging: HBV positive, LFTs elevated, EMG: Mononeuritis multiplex. Angiography: SMA aneurysms.

Diagnosis: Polyarteritis Nodosa (PAN, HBV-Related) → Abdominal pain, neuropathy, aneurysms, HBV.

Management: Admit for treatment. Start prednisone 1 mg/kg/day PO + entecavir 0.5 mg PO daily (HBV suppression). Cyclophosphamide 2 mg/kg/day PO (severe GI/neurologic involvement). Monitor LFTs (improving), abdominal pain (resolves). Transition to azathioprine maintenance. Discharge with hepatology/rheumatology follow-up.

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