Purpuric Rash Case Study

Subjective

HPI:

Patient is a 68-year-old male who is otherwise healthy besides being obese and having a new diagnosis of hypertension in the last year. His rash has been going on for a few weeks, this is the first time he's had it, it started on the legs and has since spread to her torso and arms in the last 2 days which prompted her ED visit, he hasn't tried any treatments and can't pinpoint anything that makes it better or worse. He's otherwise had a cold recently with fatigue, sinus congestion, rhinorrhea, and a cough, but this has been improving since it started a few weeks ago. Medications include HCTZ and ibuprofen prn for sinus headaches.

ROS:

Constitutional: Fevers, chills, fatigue, night sweats, loss of appetite, and unintended weight loss

- Fevers: cutaneous small vessel vasculitis, or a septic vasculitis
- Weight loss of 4kg $\geq$ is one of the ACR criteria for polyarteritis nodosa
- B Symptoms (Fevers, chills, night sweats): ↑ suspicion of malignancy (e.g. LCV or a lymphoproliferative disorder triggering a type 1 cryoglobulinemia)

Eyes: Eye pain, vision changes, erythema

- Both eye pain + vision changes: granulomatosis with polyangiitis
- Young child + ocular changes (conjunctivitis) think Kawasaki dz
- Elderly + vision changes/blindness = temporal arteritis

Ears, Nose, & Throat: runny nose, nose bleeds, sinus pain; ear pain or hearing loss

- Runny nose, nose bleeds, sinus pain = granulomatosis with polyangiitis
- URI/sore throat could be a sign of Strep pharyngitis = infectious trigger of CSVV

Respiratory: shortness of breath, cough (+/- productive)

- Infection/pneumonia as a CSVV trigger, but *also* remember that sick pts w/ purpura, pulmonary involvement may hint to *any* of the 3 ANCA vasculitides

Cardiac: chest pain, palpitations, and orthopnea

- CHF is the #1 killer of pts w/ Eosinophilic granulomatosis with polyarteritis

Gastrointestinal: N/V/D, abdominal pain, and hematochezia

- Gi sx can hint Henoch schonlein purpura, or maybe inflammatory bowel disease as an underlying association for a cSVV pt
- Kawasaki disease pts may experience abdominal pain and diarrhea

Genitourinary: urinary frequency, urgency, dysuria, hematuria, tenderness

- UTI as trigger for cSVV
- Gross hematuria = HSP or granulomatosis with polyangiitis
- Testicular tenderness or pain = polyarteritis nodosa in male pts

Musculoskeletal: joint pain, muscle pain, or muscle weakness

- Arthralgias can be nonspecific but are seen with cSVV, urticarial vasculitis, or 75% of Henoch-Schoenlein purpura cases
- Joint pain may be a sign of associated autoimmune disease or can occur in the cryoglobulinemias *w/out* associated autoimmune disorders as well
- Muscle pain, tenderness, or weakness can be seen in polyarteritis nodosa pts

Neurological: peripheral neuropathy, numbness, tingling, or motor weakness

- Neuro changes are present in over half of PAN, GPA, and EGPA pts

Hematology: easy bruising, bleeding gums, epistaxis, lymphadenopathy

- Easy bruising, bleeding gums, or epistaxis = coagulation or platelet abnormalities
- While swollen glands or lymph nodes → think malignancy or infection

Skin: pruritis, pain, and location of new lesions

Objective

Physical Exam:

Vital Signs: HTN can be a sign of polyarteritis nodosa or EGPA

Ears, Nose, Throat: examine mucosa of the nose and mouth for ulcerations or the pathognomonic strawberry gums of granulomatosis with polyangiitis

Genitourinary: testicular tenderness = polyarteritis nodosa in male pts

MSK: peripheral (UE + LE) motor strength

Neuro: sensation, gait

Skin: Crucial features: 1) size of purpura 2) +/- palpable 3) +/- livedoid pattern

Other: Look for urticarial lesions = urticarial vasculitis. Also take note of whether lesions appear on areas susceptible to the cold, such as the ears.

Labs:

There is no protocol for workup (patient dependent). Should be guided by your PE and ROS.

Tier 1: CBC, CMP, UA as general screening.

Tier 2: If unresolved, consider LFT's, stool guiac, coagulation profile, Sed Rate, CRP, ANA, rheumatoid factor, ASO titers, HIV, Hepatitis serologies

Tier 3: C or P ANCA, cryoglobulins, anti-phospholipid ab, C3 & C4 complement levels, SPEP, UPEP, Peripheral Blood Smear

Other work-up: Chest X-ray, punch biopsy

Clinical Reasoning

CBC:

- Platelet count: elevated in essential thrombocytosis or low in a variety of inherited or acquired platelet disorders (e.g. ITP)
- Anemia: polyarteritis nodosa or Kawasaki disease patients, or abnormalities suggestive of a lymphoma (e.g. impressive lymphocytosis or pancytopenia)
- Eosinophilia: cholesterol emboli or EGPA

CMP:

- Renal involvement w/ elevated creatinine → Henoch-Schoenlein purpura, polyarteritis nodosa, granulomatosis with polyangiitis, and microscopic polyangiitis

UA:

- Hematuria in Henoch-Schoenlein purpura, granulomatosis with polyangiitis, microscopic polyangiitis, and polyarteritis nodosa
- Detects glomerulonephritis by detecting red cells, red cell casts, and protein in the urine

Coagulation Profile:

- ↑ in PT, PTT, or INR = info on coagulation status

ESR & CRP:

- Clue for systemic involvement, but they are especially helpful for Kawasaki disease and temporal arteritis

Complement Levels:

- Helpful w/ cryoglobulinemia → low in 90% of those pts
- Helpful w/ urticarial vasculitis and systemic lupus, since low complement levels are associated with systemic involvement

Rheumatoid Factor:

- Positive in over 70% of cryoglobulinemia cases → cryoglobulinemia types 2 and 3 are caused by a mix of antibodies in a patient's system

Hepatitis Panel:

- Hep B & C can be associated with polyarteritis nodosa *and* cryoglobulinemia

Tox Screen:

- If considering, cocaine (levamisole-induced vascular lesions) → tox screen can help clinch the diagnosis

Biopsy:

- You perform two lesional punch biopsies, one for H&E and the other for DIF
- Biopsy shows vasculitis present or not. If +, categorized based on the vessel size involved. If -, then consider vasculopathy → hematology referral

DIF

- IgA deposition in and around small blood vessels = Henoch-Schoenlein

- DIF will also be positive in ~ 70-80% of several other types of vasculitis, including cSVV, urticarial vasculitis, and Polyarteritis nodosa = perivascular C3 and IgM, IgA, or IgG

Assessment

1) **Cutaneous Small Vessel Vasculitis**

Plan

1) **Supportive Care**

- Treat any underlying infections, stop or switch any meds you suspect are playing a role, and then tell patient elevate and compress their legs, which is often the most severely affected area for cSVV pts
- If your cSVV patients are itchy, you can add on antihistamines, topical steroids, or other topical treatments for pruritus such as calamine lotion
- If patients aren't getting better with supportive measures or they are severely affected, that's when you reach for systemic medications such as colchicine, dapsone, prednisone, or other systemic immunosuppressants