30 - Vasculopathy

Background:

-Conditions with blood vessel damage in the absence of vasculitis

-Often due to problems with clotting, typically an inherited or acquired issue with platelets or the coagulation pathway

Inherited coagulopathies

- Factor 5 Leiden: Factor V that is resistant to degradation by protein C → coagulation cascade activated
- Protein C/S deficiency: incapable of deactivating factors V & VIII = "broken brakes" →coagulation cascade activated
- Antithrombin III mutation: Blocks factors II & X = broken brakes!
- Hyperhomocyteinemia: 2-4x risk of thrombosis
- Sickle cell disease: Acidosis or low O2 → sticky sickle cells → clotting

Acquired coagulopathies

-Anti-phospholipid syndrome: patients are more prone to forming clots, therefore look for a hx of stroke, MI, DVT, PE, miscarriages. Cutaneous changes are related to occlusion, including livedo reticularis, splinter hemorrhages, or retiform purpura.

- Anti-cardiolipin
- Lupus anticoagulant
- Anti-beta2 glycoprotein

-Liver disease: not only affects coagulation production but also has an impact on platelet function

-Type I cryoglobulinemias: lymphoproliferative disorder making monoclonal IgM = clogging up vessels → vasculopathy

-Purpura fulminans: acutely sick patient with disseminated intravascular coagulation (DIC)

PEARL: Risk factors for acquired coagulation: immobilization, obesity, cancer, pregnancy, smoking, oral contraceptives

Increased platelet destruction

- ITP (Idiopathic Thrombocytopenic Purpura): autoimmune destruction of platelets due to IgG autoantibodies coating platelets and leading to macrophage destruction in the spleen

- TTP (Thrombotic Thrombocytopenic Purpura): caused by a deficiency in ADAMS TS13, an enzyme that cleaves VW factor multimers. Since these multimers build up you get platelet aggregation and thrombosis. THINK "FAT RN": Fevers, Anemia, Thrombocytopenia, Renal, Neurologic

-DIC: massive activation of coagulation, leading to ischemia and diffuse thrombosis.

-HIT (Heparin Induced Thrombocytopenia)

Abnormal platelet function

-Medications (aspirin, NSAIDs)

-Myeloproliferative disorders

-Renal disease: chronic renal failure and may present with bruising, petechiae, and GI bleeding. Use of dialysis improves platelet function!

Embolic Disorders

-Cholesterol emboli: cholesterol fragments that dislodge from a plaque and travel downstream to plug arterioles in the skin. Can occur spontaneously or after cardiac procedures . Associated with peripheral eosinophilia!

-Oxalate emboli

Miscellaneous

-Pigmented purpuras

- Schambergs disease: affects the lower legs of middle aged adults appears as petechiae with golden-brown hemosiderin staining
- Lichenoid purpura of Gougerot and Blum: rust-colored to violaceous lichenoid papules on the legs and trunk of older men
- Purpura annularis telangiectodes; 1-3 cm annular patches with petechiae on the legs of younger women
- Lichen Aureus: solid golden to rust colored macules or papules
- Eczematous Dermatitis of Doucas and Kapetanakis: mix of eczema and petechiae showing up on older men

Diagnosis:

Biopsy will show fibrin thrombi with minimal inflammation and no leukocytoclastic vasculitis

Screening labs: PT, PTT, INR, Protein C/S, Factor V Leiden, Antithrombin III, Prothombin 22-10 gene mutations, anti-prothrombin antibodies, homocysteine levels

Treatment:

Targets underlying disease, often in conjunction with a hematologist.

Pigmented purpuras: Topical steroids + vitamin C 500mg BID + rutoside 50mg BID