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Collagen Vascular Disease - Considerations for Emergent Management

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Emergent Management

• Chronic conditions with long-term management decisions made by others but
  - flares of general disease process
  - end-organ involvement
• Because of their chronicity they present with
• Occasionally determine the initial diagnosis
Where are you taking me today?

- Raynaud phenomenon
- Reactive arthritis (Reiter Syndrome)
- Rheumatoid arthritis
- Systemic sclerosis (Scleroderma)
- Systemic lupus erythematosus
- Polymyalgia rheumatica
- Polymyositis / Dermatomyositis
- Vasculitides
Raynaud Phenomenon

• Definition:
  – Exaggerated vasospasm of digital / precapillary arteries of fingers, toes, ears, nose, knees and nipples
  – Initiated by exposure to cold or emotional stress
Raynaud Phenomenon

• Etiology:
  – Unknown
  – May be first precursor of future connective tissue disorder

• Clinical Presentation:
  – Triphasic
  – Typically begins in one finger then symmetrically spreads to other fingers but usually spares the thumb
Raynaud Phenomenon

• Triphasic progression
  – White – pallor
    • Lack of arterial flow due to vasospasm
  – Blue
    • Cyanosis from blood pooling
  – Red
    • Reactive hyperemia

– Ischemic phases (white-blue) last 15-20 min
Raynaud Phenomenon

• Relative temperature shifts may be provocative

• General body chill can trigger

• Fear / anxiety can trigger
Raynaud Phenomenon

• Specific criteria
  – Symmetric episodic attacks
  – No evidence of peripheral vascular disease
  – No tissue gangrene, digital pitting, or tissue injury
  – Negative nailfold capillary examination
  – Negative ANA, normal ESR
Raynaud Phenomenon

• Nailfold capillary microscopy –
  – Place drop of immersion oil on periungual area then examine with ophthalmoscope set at diopter 40 or a dissecting microscope.
  – Should see regularly spaced capillary loops
  – Abnormal findings –
    • Enlarged or distorted capillary loops
    • Relative paucity of loops
Raynaud Phenomenon

• Treatment
  – Avoidance strategies
    • Cold, nicotine, sympathomimetics (decongestants, diet pills, herbs containing ephedra)
  – Drug therapy
    • Ca channel blockers, direct vasodilators (NTG, hydralazine, minoxidil), sympatholytics (methyldopa, reserpine, prazosin), prostaglandins, anticoagulation / antithrombotic tx (aspirin, dipyridamole, heparin, LMWH)
    • sympathectomy
Raynaud Phenomenon

- Severe ischemia
  - Warm patient (body and digits)
  - Analgesics
  - Antiplatelet tx – aspirin
  - Vasodilator tx
    - Nifedipine extended release 30-60 mg daily
    - Amlodipine 5-10 mg daily
  - Topical NTG
  - Heparin / LMWH 24-72 hrs
  - Temporary chemical sympathectomy
    - Digital or regional block – lidocaine/bupivicaine
  - IV prostaglandin administration
Reactive Arthritis (Reiter Syndrome)

• **Definition:**
  - Arthritis following a preceding infection without intra-articular presence of the pathogen i.e. not a septic joint

• **Etiology:**
  - Seronegative (rheumatoid factor negative)
  - Spondyloarthropathy (very likely HLA-B27 pos)
  - Follows a GU or GI infection
Reactive Arthritis

• Clinical Presentation:
  – Male 15 – 35 y/o
  – Asymmetric, oligoarthritis (2-4 joints)
  – Usually involves lower extremeties and sacroiliac joints
  – Skin lesions resembling pustular psoriasis on palms and soles – *keratoderma blennorrhagicum*
  – lesions on glans penis – *balanitis circinata*
Reactive Arthritis

• Classic triad of Reiter syndrome
  – Nongonococcal urethritis (Chlamydia)
  – Conjunctivitis / anterior uveitis
  – Arthritis

• Following GI infection with Shigella, Salmonella, Campylobacter, Yersinia, Clostridium difficile (?)

• Infection precedes arthritis by 2 - 6 wk
Reactive Arthritis

• Treatment:
  – NSAIDs
    • Naproxen 500 mg TID
    • Indomethacin 50 mg TID
  – Intra-articular glucocorticoids
  – Expect resolution 3 – 12 months
Rheumatoid Arthritis

• Definition:
  – Chronic, symmetric polyarticular synovial joint disease

• Nonarticular & Systemic Manifestations
  – HEENT
    • Episcleritis
      – Painless injection of episcleral vessels (self-limiting)
    • Scleritis
      – Dark red / purple discoloration with marked ocular tenderness
      – Potential for visual impairment and scleral rupture
Rheumatoid Arthritis

• Cricoarytenoid joint
  – Dysphonia, hoarseness, stridor
  – If fix in closed position could require emergent tracheostomy

• Ligamentous destruction of transverse ligament of C-2 with potential for cord compression
Rheumatoid Arthritis

• Pulmonary
  – Pleural effusion *
  – Interstitial fibrosis
  – Pulmonary nodules

• Cardiac
  – Pericarditis *
  – Pericardial effusion 10%
  – Myocarditis
  – CAD – sudden death, MI *
Rheumatoid Arthritis

- Renal
  - Focal glomerulonephropathy
  - Drug toxicity from treatment *
- Vasculitis
  - Distal infarcts, ulcerations, gangrene
- CNS
  - Generally spared
Systemic Sclerosis - Scleroderma

• Definition:
  – Disease process characterized by progressive fibrosis, vascular abnormalities and inflammatory processes that can be manifested quite locally or diffusely systematically with organ system involvement

• Etiology:
  – Poorly understood
Systemic Sclerosis

• Clinical Presentation:
  – Often initial signs are a thickening, hardening of the skin, usually fingers, hands and face and Raynaud phenomenon
  – Female > Male
  – African-Americans tend to have worse prognosis due to greater likelihood of having a more severe diffuse form
Systemic Sclerosis

• Skin
  – Sclerodactyly
  – Telangiectasias
  – Digital ulcers
  – Calcinosis
  – Raynaud phenomenon
In scleroderma, the abnormal build-up of fibrous tissue in the skin can cause the skin to tighten so severely that the fingers curl and lose their mobility.
Telangiectasia

Dilation of small vessels and capillaries cause flat red marks to appear on the skin.
Systemic Sclerosis

- **Pulmonary**
  - Most common cause of death
  - Interstitial lung disease
    - Alveolitis leading to eventual fibrosis
  - Pulmonary vascular disease
    - Pulmonary hypertension
- **Cardiac**
  - Pericarditis w/wo effusion
  - Myocardial fibrosis
    - Resultant ventricular dysfunction with diminished cardiac output
  - Dysrhythmias
    - Fibrosis of conduction system resulting in sudden death
Systemic Sclerosis

• Renal
  – Renal crisis
    • Acute onset of renal failure
    • Proteinuria and microscopic hematuria
    • Abrupt onset of hypertensive emergency

• Gastrointestinal
  – Hypomobility
    • Esophageal dysmotility / GERD
    • Pseudo-obstruction, constipation
    • Teleangectasias with bleeding
    • Pneumatosis intestinalis
Systemic Sclerosis

Treatment

- Generally immunosuppressive therapy
- Based on specific organ system involved
- Emergently
  - Pulmonary decompensation
  - Cardiac – effusion, failure, dysrhythmia
  - Renal crisis
    - ACE inhibitor is first line antihypertensive agent – Captopril
    - Captopril + Ca channel blocker
    - Angiotensin receptor blocker for those who can not tolerate ACE inhibitor
Systemic Lupus Erythematosus

• Definition:
  – Chronic autoimmune disease characterized by presence of autoantibodies with multi-organ system involvement

• Etiology:
  – Genetic predisposition and nebulous factors combine to alter immune cell function resulting in production of autoantigens and thereby auto-antibodies with systemic consequences.
SLE

• Clinical Presentation:
  – Female > Male 10 : 1
  – Typical presentation 21 – 45 yrs of age
  – African American > Caucasians

• Constitutional
  • Wt loss, fever, myalgias, arthralgias
  • Fatigue often the most debilitating

• Skin
  • Butterfly malar rash – may be fleeting
  • Oral and nasal ulcerations
SLE

• Pulmonary
  – Pleurisy, effusion, interstitial lung disease, pulmonary hypertension
  – “lupus lung” – alveolar hemorrhage

• Cardiac
  – Pericarditis *, effusion
  – Increased risk for CAD

• Renal
  – Lupus nephritis
    • Elevated creatinine, proteinuria, hypertension
SLE

- **Neurologic**
  - Cognitive defects, cephalgia, seizures, peripheral neuropathies (stocking / glove), psychosis, stroke (antiphospholipid antibody syndrome)

- **Musculoskeletal**
  - Arthritis, atrophy, tendon rupture
SLE

• Treatment:
  – Immunosuppression
    • Glucocorticoids
    • Methotrexate, cyclophosphamide, azathioprine, mycophenolate, rituximab

• Causes of death
  – Early deaths – first few years
    – Active lupus (cardiac, renal, CNS dz)
    – Infection due to immunosuppression
  – Late deaths
    – Chronic effects of lupus (ESRD, CAD)
    – Infection, malignancy
Polymyalgia Rheumatica

• Definition:
  – Rheumatic condition frequently associated with giant cell (temporal) arteritis

• Etiology:
  – Genetic predisposition
  – Relatively common
  – 50% of pts with GCA will develop PMR
  – 15% of pts with PMR will develop GCA
Polymyalgia Rheumatica

• Clinical Presentation:
  – Age 50 or older at onset
  – Bilateral aching and morning stiffness which lasts 30 min or more for 1 month or more involving at least 2 of 3 areas
    • Neck or torso
    • Shoulders or proximal arms
    • Hips or proximal thighs
  – ESR = 40 mm/hr or greater
Polymyalgia rheumatica
Giant Cell Arteritis

Source Undetermined
Polymyalgia Rheumatica

- treatment:
  - Prednisone 10 – 20 mg / day
    - (compared to 60 mg / day dose for GCA)
  - Rapid response is characteristic (often after first dose)
  - Relapse is commonly seen requiring increase in prednisone
Polymyositis / Dermatomyositis

• Definition:
  – Idiopathic inflammatory myopathies

• Etiology:
  – Genetic component with presumed environmental triggers
  – Peak incidence between 40 – 50 yrs of age
  – Female : Male 2 : 1
Polymyositis / Dermatomyositis

• Clinical Presentation:
  – Muscle weakness
    • Onset is insidious
    • Gradually worsening over months
    • Typically symmetric and proximal
  – Myalgias / muscle tenderness occurs in 25-50% but is mild compared to PMR or fibromyalgia
  – Dermatologic findings in DM
    • Gottron’s sign
      – Erythematous, often scaly exanthem occurring symmetrically over MCP and IP joints and/or over extensor surfaces of elbows and knees resembling psoriasis
Polymyositis / Dermatomyositis

- Heliotrope rash
  - Violaceous eruption on upper eyelids, often with swelling

- Shawl sign and V sign
  - Diffuse flat erythematous lesion over chest and shoulders (shawl sign) or over anterior neck and chest (V sign)

- Erythroderma
  - Extensive areas of skin redness (malar, forehead)

- Mechanic’s hands (DM or PM)
  - Rough, cracking skin at tips and lateral aspects of fingers with irregular dirty appearing lines
Polymyositis / Dermatomyositis

- Diagnostic testing
  - Elevated CK, LDH, aldolase, aminotransferases
- Increased incidence of malignancy associated with dermatomyositis

- Treatment:
  - Glucocorticoid regimen
    - initiate with high doses for several months to establish disease control
    - Slow taper to lowest effective dose for 9 – 12 months
  - Glucocorticoid-sparing regimen
    - Azathoprine / methotrexate
Vasculitides

• Large vessel
  – Takayasu arteritis
  – Giant cell (temporal) arteritis

• Medium vessel
  – Polyarteritis nodosa
  – Kawasaki disease

• Small vessel
  – Churg-Strauss arteritis
  – Wegener’s granulomatosis
  – Henoch-Schönlein purpura