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RHEUMATOLOGY ADVANCED PRACTICE PROVIDERS

Inaugural National Conference

December 3 – 5, 2020

VIRTUAL CONFERENCE

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Mixed Connective Tissue Disease/Myositis

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Faculty Disclosures

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- Consultant: Pfizer
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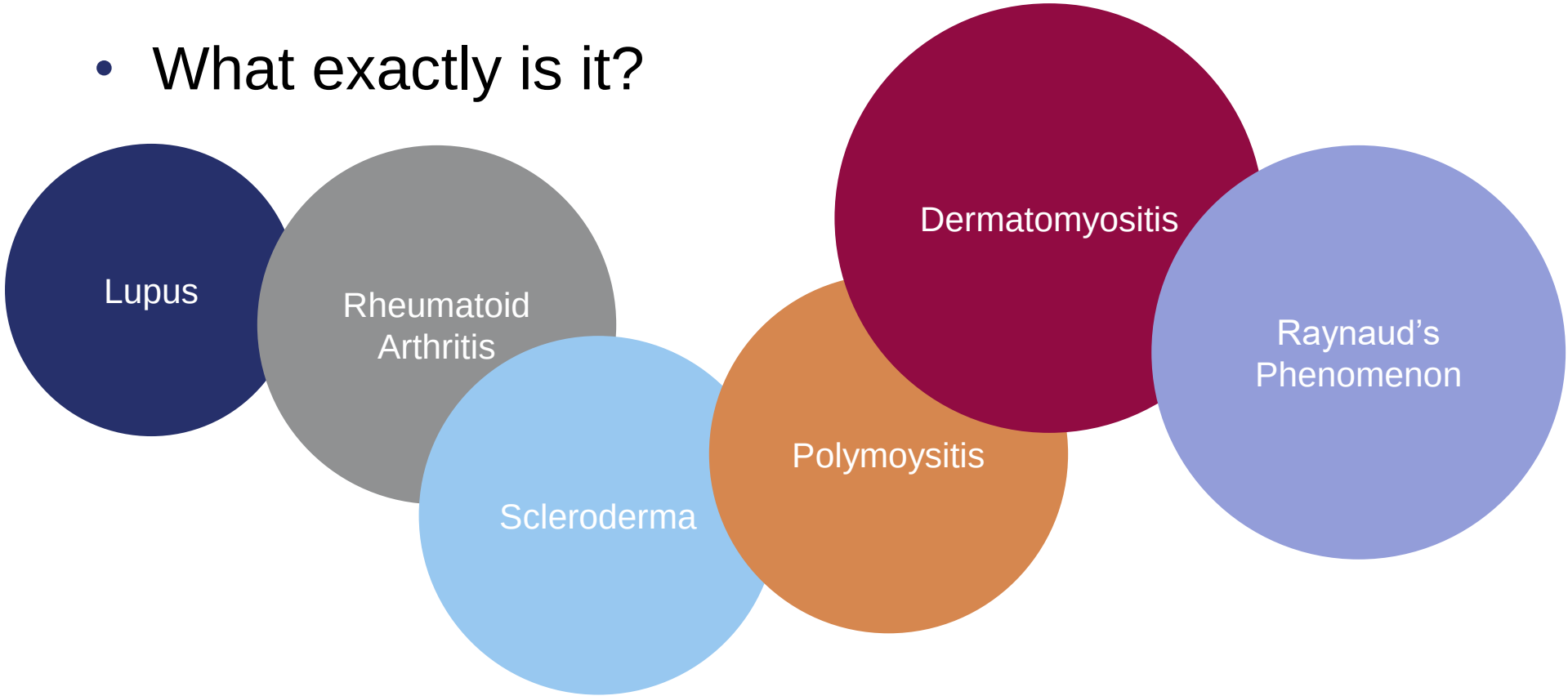
Mixed Connective Tissue Disease (MCTD)

- Introduction
- Epidemiology
- Symptoms
- Complications
- Diagnosis
- Management

MCTD

- What exactly is it?



MCTD

- By definition, a patient with MCTD has signs and symptoms of a combination of disorders
  - Primarily lupus, scleroderma, and polymyositis
- Patients often also have
  - Sjogren’s Syndrome
  - Rheumatoid arthritis
  - Raynaud’s phenomenon

MCTD

- Who gets it?
  - Women
  - 40-50 yo
- MCTD is rare
  - US incidence is ~1.9:100,000 adults per year

Causes

- MCTD is a known autoimmune disorder but specific cause is unknown
- With connective tissue disease, your immune system attacks the fibers that provide the framework and support your body
- ?? Family history
  - Role of genetics is unclear

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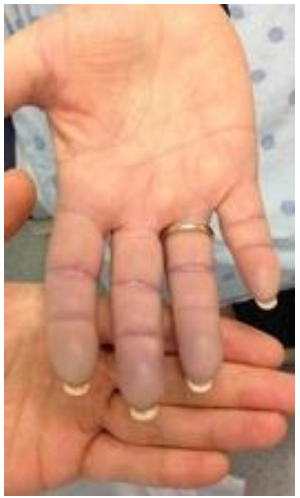
Symptoms

Symptoms

Raynaud’s phenomenon is often the earliest sign and can proceed other entities



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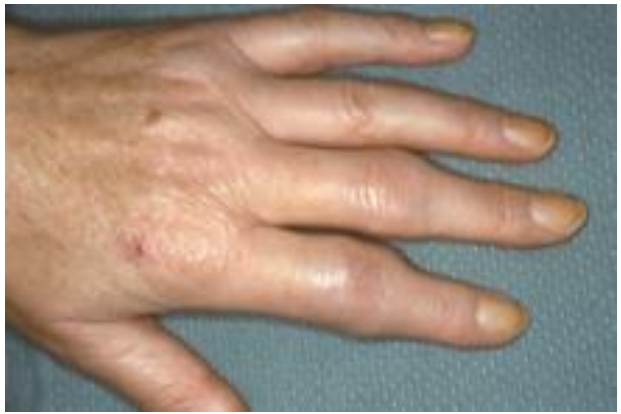
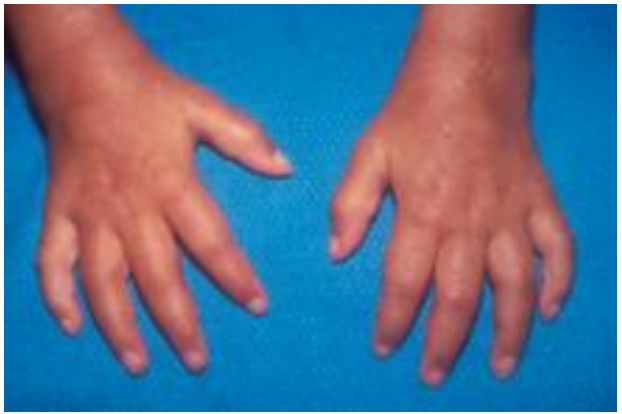
Symptoms (cont'd)

- Fatigue
- Malaise
- Arthralgias
- Low grade fever
- Skin changes
  - Lupus-like rash; tightening of the skin over the fingers
- Difficulty swallowing (solids), heartburn

Symptoms (cont'd)

- Swollen fingers and hands
- Inflammatory arthritis observed in 75% of patients





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Symptoms (cont'd)

- Muscle weakness
  - Clinical inflammatory myopathy
  - Histologically similar to polymyositis

Symptoms (cont'd)

- Shortness of breath
  - Includes interstitial lung disease and pulmonary hypertension
  - 75% of patients are affected

Symptoms (cont'd)

- Chest pain
  - 40% of patients have cardiac disease
  - Pericarditis is the most common variant

Symptoms (cont'd)

- Neurologic abnormalities are noted in approximately 10% of patients

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Complications

Complications

- Pulmonary hypertension
  - A major cause of death
- Interstitial lung disease
  - Lung scarring affects patient’s ability to breathe

Complications (cont'd)

- Kidney damage
  - Affects 25% of patients
  - Generally more mild than what we see with lupus nephritis
- Anemia
  - 75% of patients have iron deficiency anemia

Complications (cont'd)

- Tissue death
  - Patient's with severe Raynaud's can develop gangrene
- Hearing loss
- Nerve damage
  - Sjogren's syndrome can affect the trigeminal nerve and result in trigeminal nerve neuralgia

Complications (cont’d)

- Heart disease resulting from inflammation around the heart

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Diagnosis

Diagnosis

- Physical exam
  - Swollen hands and fingers
  - Raynaud’s
  - Lymphadenopathy, splenomegaly and hepatomegaly
  - Rash or hair loss
  - Dry mouth

Diagnosis (cont'd)

- Laboratory studies
  - +ANA and +RNP antibody
- Other labs possibly show
  - Elevated CK and/or aldolase
  - Elevated ESR
  - Anemia
  - Leukopenia

Diagnosis (cont'd)

- Imaging
  - Chest x-ray
  - X-rays of joints
  - Echocardiogram
  - PFTs

Diagnosis (cont'd)

- Less common imaging
  - High resolution CT scan
  - Angiogram
  - Right heart catherization

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What Is the Appropriate Management for These Patients?

Management

- Pharmacologic
  - Corticosteroids
  - Hydroxychloroquine
  - Calcium Channel Blockers
  - Proton Pump Inhibitors, bosentan or sildenafil
  - Azathrioprine, methotrexate, or mycophenalate

MCTD – Management

- Non-pharmacological management
  - NSAIDs
  - Smoking cessation
  - Reduce stress

MCTD – Management

- Monitoring
- PFTs and echocardiogram
- Labs
- Interprofessional team approach

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Myositis

Myositis

- Definition
- Epidemiology
- Symptoms
- Diagnosis
- Management

What Is Myositis?

- Disease of unknown cause
- Characteristic symptoms
  - Symmetric proximal muscle weakness
  - Elevated muscle enzymes (CPK, aldolase, transaminases, LDH)
  - Myopathic EMG abnormalities
  - Typical changes on muscle biopsy
  - Typical rash of dermatomyositis
- Polymyositis consists of definite 4 out of the 5 characteristic symptoms (probably 3 out of 5)
- Dermatomyositis consists of typical rash AND definite 3 out of the 4 other characteristic symptoms (probably 2 out of 4)

First Author First Initial et al. *Journal.* Year;Volume: First page-last page.

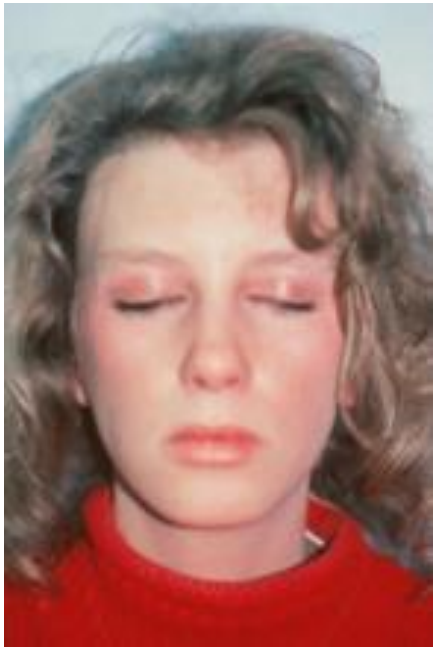
Epidemiology

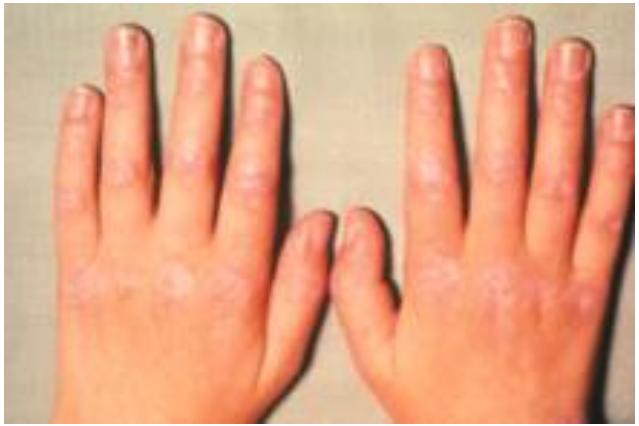
- Rare
  - 1:100,000
- Can occur in any age, including pediatric patients
  - Peak age is 40-50
- Women are twice as likely to be affected

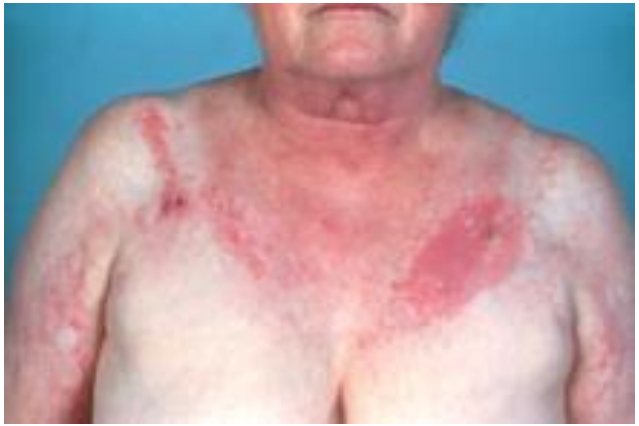
Symptoms

- Symmetric proximal muscle weakness
- Dysphagia, less often hoarseness
- Rash
  - Gottron’s papules
  - Heliotrope rash
  - Photosensitive – V sign
  - Mechanic’s hands

Symptoms (cont'd)







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Symptoms (cont'd)

- Dyspnea
  - Must monitor for interstitial lung disease
  - Can also be a result of ventilatory (diaphragmatic and intercostal) muscle weakness

Diagnosis

- Laboratory Studies
  - Muscle enzymes including CPK, aldolase
  - LFTs and LDH
  - Myositis specific antibodies, most commonly Jo 1

Diagnosis (cont'd)

- EMG is obtained to monitor the electrical activity in the muscles
- Muscle biopsy
- MRI
- Chest x-ray
- If needed:
  - High resolution CT
  - PFTs

Management

- Corticosteroids
- Methotrexate and azathioprine
- Less often
  - Mycophenelate
  - Rituximab
  - IVIg

Management (cont'd)

- Commonly linked to malignancy (especially dermatomyositis)
- Cancer screening is *vital*
  - Age-specific mammograms
  - PAPs
  - Colonoscopy

Conclusions

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Questions?