IDIOPATHIC INFLAMMATORY MYOSITIS

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IDIOPATHIC INFLAMMATORY MYOSITIS (IIM)

- I Primary Idiopathic Polymyositis
- II Primary Idiopathic Dermatomyositis
- III Dermatomyositis or Polymyositis associated with Malignancy
- IV Childhood Dermatomyositis or Polymyositis
- V Dermatomyositis or Polymyositis associated with Connective Tissue Diseases
- VI Inclusion Body myositis
- VII Miscellaneous, Eosinophilic myositis, myositis ossificans, focal myositis, infectious myositis, giant cell myositis
REVIEW- POLYMYOSITIS AND DERMATOMYOSITIS
SHARE MANY CLINICAL FEATURES:

<table>
<thead>
<tr>
<th>Polymyositis</th>
<th>Dermatomyositis</th>
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<tbody>
<tr>
<td>Symmetrical proximal muscle weakness - striated muscle</td>
<td>Polymyositis with differences:</td>
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<tr>
<td>Elevated muscle enzymes</td>
<td>specific dermatologic manifestations</td>
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<tr>
<td>Characteristic EMG and Muscle BIOPSY</td>
<td>Some distinct muscle biopsy features</td>
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<td>Can be associated with malignancies</td>
<td>Seen in children</td>
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<td>Rare 1-8 per 10^6</td>
<td>Black to white 3:1</td>
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<td>2 females to 1 male</td>
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<td>45-60 years of age</td>
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<td>Rare in children</td>
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<td>Black to white 5:1</td>
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INCLUSION BODY DISEASE- A DIFFERENT “ANIMAL”

- Slowly progressive myopathy with characteristic biopsy features
- Disease of older males > 50
- Asymmetric distal weakness
- Non responsive to therapy
- Some Neurologic features

CLASSIC SYMPTOMS OF IDIOPATHIC INFLAMMATORY MYOSITIS

- Slowly progressive weakness 3-6 months, muscle tenderness
- (IBM may progress over years)
- NO real precipitating event
- Is symmetrical
- Unable to rise from a chair, brushing hair
- Other symptoms
  - Swallowing difficulty, dysphonia, diaphragmatic weakness, cardiac symptoms
HISTORY

- Thorough history
- Illicit Drug history especially cocaine alcohol
- Medications OTC drugs
- Symptoms of endocrine disease
- FAMILY history of muscular dystrophy or metabolic myopathies
- Family history of autoimmune diseases

METICULOUS SKIN EXAM...

SKIN FEATURES OF DERMATOMYOSITIS

GOTTRON’S PAPULES—PATHOGNOMONIC FOR DERMATOMYOSITIS
DERMATOMYOSITIS - COMMON SKIN LESIONS

Autoimmune Pathogenesis: Distinct Diseases on Muscle Biopsy

Polymyositis - CD8 cell endomysial

Dermatomyositis - B cell, perimysial perivascular

Capillary Microscopy

Cuticular overgrowth
## MYOSITIS SPECIFIC AND MYOSITIS ASSOCIATED ANTIBODIES—A GROWING LIST

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<th>Antibody</th>
<th>Antigen (RNA synthetase)</th>
<th>Prevalence in IIM (%)</th>
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<td>PL-12</td>
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<td>Zo</td>
<td>Phenylalanyl</td>
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## MOST COMMONLY USED MYOSITIS ANTIBODIES

- Anti-Jo
- Anti-SRP
- Anti Mi-2
- Anti p155/140
- cAMD-140
- Anti PM-SCL
- Anti U1 RNP

- ILD, RP, Mechanic’s hands Arthritis
- Severe necrotizing PM
- Dermatomyositis
- Cancer associated DM
- Clinically amyopathic Dermatomyositis (ILD)
- Overlap CT with PM and Scleroderma features
- MCTD

## TRANSFER RNA SYNTHETASE SYNDROMES

- Specific amino acid Transfer RNA antibodies
CANCER ASSOCIATED MYOSITIS (CAM)

- Cancer rates are increased in DM-PM
  - DM, RR 2-4 fold increase, PM 1.5-2
- The majority of cancers are diagnosed in 1 year
  - Before or after diagnosis
  - DM: solid tumors, PM: lymphomas, Asians, N-P cancers
- Cancer risk returns to baseline after 5 years
- WHO gets CAM?
  - older (>45)
  - Men > women
  - Severe skin disease with Necrosis and ulcers
  - Rapid progressive weakness with dysphagia

64 AA female with cpk 380, rapid and progressive weakness, later found to have Endometrial cancer.

MYOSITIS MARKERS FOR MALIGNANCY

- Lack of autoantibodies favors malignancy
  - Anti Synthetase antibodies are protective
- Anti p-155/140 antibodies –associated with Cancer in DM
  - Several types TIF-1 alpha, TIF-1 beta, TIF-1 gamma
  - Regulate transcription, are involved in carcinogenesis
  - These autoantibodies are specific for DM, can occur in JDM
- http://omrf.org/research-faculty/core-facilities/myositis-testing/
- Meta-analysis of p-155 to diagnose CAM*
  - PPV 58%, NPV 95%

REMEMBER THIS CASE (1)

- 33 year old woman is referred for shoulder and hip-girdle weakness.
- Mild dyspnea on exertion for 3 months.
- Her medications are lisinopril 10mg po qd for mild HBP. Her sister has lupus.
- ON PE she has low grade fever, proximal muscle weakness; 3/5 both shoulders and hips.
- Swelling of the PIP’s and wrists (arthritis) with dry cracked finger tips.

CASE 1

- Laboratory studies:
  - Hgb 10.5g/dl (12-15g/dl)
  - ESR 30mm/hr (<20mm/hr)
  - AST 85 U/L
  - ALT 77 U/L
  - Creatinine normal
  - CPK 2540 (normal 30-135U/L)
  - Anti-Jo-1 antibodies positive

CASE 1 -- WHAT IS THE NEXT BEST STEP IN MANAGEMENT?

A. Computed tomography of the chest
B. Mammography
C. Ultrasonography of the liver
D. Magnetic resonance imaging of the thigh
E. ANA, SS-A, SS-B, ANTI-Sm, ANTI RNP
CASE 1 ANSWER IS A. COMPUTED TOMOGRAPHY OF THE CHEST

- Anti synthetase syndrome
  - Interstitial lung disease
  - Mechanic’s hands, fever
  - Raynaud’s, arthritis
  - Transfer RNA antibodies
    - Anti Jo-1
    - PL 7
    - PL 12
    - EJ
    - CJo

INTERSTITIAL LUNG DISEASE

BASIC WORKUP FOR IDIOPATHIC INFLAMMATORY MYOSITIS

- Creatine phosphokinase (CPK), aldolase,
- Liver function tests
  - LDH is more sensitive to muscle necrosis
- Myositis specific and associated antibodies, ANA
- EMG
- MRI - muscle inflammation vs atrophy. T2 weighted or STIR
- Plain x-rays - cutaneous or muscle calcifications
- Computerized tomography - ILD or cancer screen
- Urinalysis and urine myoglobin - Muscle biopsy (skin biopsy helpful)
- Other tests:
  - pulmonary function studies,
  - electrocardiography (ECG),
  - esophageal manometry or barium swallow
  - Total body CT

EMG FINDINGS IN IDIOPATHIC INFLAMMATORY MYOSITIS

Table 1 – Electromyographic findings in myositis

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<th>EMG signs</th>
<th>Normal</th>
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IMAGING IN POLYMYOSITIS AND DERMATOMYOSITIS

MRI  INFLAMMATION    VS
ATROPHY

KEEP IN MIND OTHER CAUSES OF MYOSITIS

- Viral and bacterial causes
- Systemic Vasculitis or CTD related elevations
- Endocrine causes such Hypothyroidism or Diabetes or Cushing’s disease
- Metabolic diseases
- Electrolyte abnormalities
- Inherited disorders of muscle MG or Dystrophies
- Myotoxins, Alcohol, cocaine, malathion, cimetidine
- Medication induced-- direct and immune mediated cpkemias- statins, colchicine,
DIAGNOSTIC CLUES TO LEAD YOU AWAY FROM INFLAMMATORY MYOSITIS

- Episodic weakness related to activity or fasting (MG or metabolic myopathies)
- Asymmetric or unilateral weakness suggests neurologic disorder
- Facial or ocular weakness occurs in MG rarely in IIM
- Hypertrophy or early muscle atrophy
- Neuropathy or fasciculations or cramping
- Family history of muscle disorder
- No Family history of autoimmunity
- NO fever, rashes, arthritis or other CTD symptoms, no capillary nail bed changes
- No myositis specific antibodies
- Enzymes <2x or >100x normal
- MRI normal or only atrophic
- No response to therapy

ANOTHER CASE (2)

- An Air Force Colonel curbsides about a 18 y o recruit
  - Good athlete but noted dark urine after games
  - CPK is measures at 132,000 ALT is 1240 LDH is 6412
  - Recruit is hydrated and returns to normal
- Further find out
  - Recruit has no weakness, has not been ill, has no delays in development and denies drugs
  - Musculoskeletal exam normal
  - But CPK is 402 (<350), AST, ALT, LDH normal
  - Urine no myoglobin

CASE (2) YOUR ADVICE IS...

- A. Discharge from the military
- B. See a neurologist or rheumatologist
- C. Get a muscle biopsy
- D. Try to reproduce the rhabdomyolysis again
CASE 2  ANSWER IS D. TRY TO REPRODUCE THE RHABDOMYOLYSIS AGAIN

- Forearm ischemic exercise test
- Exercise the forearm and measure the ammonia and lactate
- Normal — ammonia and lactate rise
- IF only the ammonia rises, the patient lacks the enzyme myophosphorylase
- McArdle’s Disease — Myophosphorylase deficiency
- Valuable tool in the diagnosis of metabolic myopathies
  - Glycogen storage diseases
  - Myoadenylate deaminase deficiency
  - Clinical muscle examination may be un-revealing
  - Muscle strength is often normal
  - Muscle enzymes may only be elevated during symptomatic periods
  - Electromyograms are frequently normal or demonstrate unspecific changes
  - Immunohistochemistry, biochemical assays, and molecular analysis will allow a definitive diagnosis

MCARDLE’S DISEASE - GLYCOGEN STORAGE V

- Most common - Autosomal recessive (PYGM gene)
- Usually presents in ten years with ex after exercise
- “Second wind” phenomenon
- CAN PRESENT LATER IN LIFE
- 50% have myoglobinuria
- Muscle biopsy will lack the enzyme
- Renal failure from rhabdomyolysis
- 1 in 100000
- Avoid isometric exercise, B6 and sucrose
- CONSIDER metabolic myopathies

C is McArdle’s showing no myophosphorylase staining
F is control

REMEMBER THIS CASE 3

- An 80 woman with HBP, Hyperlipidemia is admitted to hospital for progressive SOB and weakness. Despite diuresis for her CHF, her SOB and weakness and muscle cramping progressed.
- On PE her RR 20/min, pulse 120/min, BP 132/90, lungs crackles, Skin normal, Muscle testing 3/5, neck flexors shoulder abductors/flexors, hip flexors.
- Lab shows
  - cpk 3510 (<200 U/L)
  - AST 335 (<40 U/L)
  - Urine myoglobin 8620 (<40 mg/mL).
CASE 3 CONTINUED.

- ANTI Jo-1, SRP, ANA were negative.
- An EMG showed myopathy but not myositis
- Muscle biopsy shows:
  - Muscle necrosis
  - No inflammatory infiltrate

Arrows show necrotic muscle fibers
But no inflammatory infiltrate

CASE 3 AND THE NEXT STEP IS...

- A. Give prednisone 1mg/kg for Transfer RNA Synthetase syndrome
- B. Stop Statin
- C. Give IVIG for Inclusion Body Muscle disease
- D. Give prednisone and Methotrexate for Polymyositis
- E. Stop Statin and give prednisolone 1mg/kg

CASE 3 ANSWER IS …D. STOP STATIN AND GIVE PREDNISOLONE

- Diagnosis: Statin induced Necrotizing myositis –
  - Another and new form of immune mediated statin induced myopathy
  - Antibody against the 3-hydroxy-3-methylglutaryl-coenzyme A reductase (HMGCR) protein, which is up-regulated in regenerating muscle fibers.
- Stopping Statin alone for this form is not enough, must give prednisone or other immunosuppressive
- People at risk for statin myopathy
  - Older
  - Muscle pain with statins, unexplained cramping
  - Simvastatin, Atorvastatin more likely
  - Average onset 6 months (range 1 week-4 years) less likely in patient on statins for years
REMINDER DRUGS THAT CAUSE MYOSITIS

- Direct Toxic Myopathy
  - Alcohol
  - Hydroxychloroquine
  - Colchicine
  - Cocaine—most common cause of illicit drug-induced medical problems in ER
  - Retroviral drugs
- Immune mediated myopathy/myositis
  - D-penicillamine
  - Hydralazine
  - Procainamide
  - Dilantin
  - Ace inhibitors
  - Statins — rhabdomyolysis and severe muscle inflammation
  - Interferon alpha

ONE INTERESTING QUESTION — IS HOW MUCH WORKUP FOR CANCER SHOULD BE DONE IN INFLAMMATORY MYOSITIS

<table>
<thead>
<tr>
<th>Conventional</th>
<th>Extensive*</th>
</tr>
</thead>
<tbody>
<tr>
<td>H&amp;P</td>
<td>CT-chest, abdomen, pelvis</td>
</tr>
<tr>
<td>CBC, CMP, ESR, UA</td>
<td>Endoscopy/colonoscopy</td>
</tr>
<tr>
<td>Fecal Occult blood</td>
<td>Bronchoscopy</td>
</tr>
<tr>
<td>Chest xray</td>
<td>Bone Marrow</td>
</tr>
<tr>
<td>AGE appropriate cancer screening</td>
<td>Serum tumor markers</td>
</tr>
<tr>
<td></td>
<td>PET imaging*</td>
</tr>
</tbody>
</table>


GOALS OF IIM THERAPY

- Reducing muscle inflammation with medications
- Decreasing weakness — rehabilitation*
- Preventing muscle atrophy
- Minimizing medication side effects such as steroid myopathy
- Avoid the sun

* when cpk <1000
TREATMENTS FOR DERMATOMYOSITIS AND POLYMYOSITIS

- Corticosteroids - only FDA approved drugs for Dermatomyositis
- Limited clinical trials
- Hydroxychloroquine
- Methotrexate/azathioprine
- Mycophenolate mofetil
- IVIG - make sure no IgA deficiency exists
- TNF inhibitors, rituximab, cyclosporine, tacrolimus and cyclophosphamide

SUMMARY

- Idiopathic inflammatory myositis are rare autoimmune diseases – women 2:1, blacks 3:5:1
- Insidious symmetric proximal muscle weakness including neck flexors but sparing facial and ocular muscles
- Gottron’s papules are pathognomonic for Dermatomyositis
- MSA denote subtypes of IIM
  - Transfer RNA synthetase, anti SRP, anti Mi-2

SUMMARY

- Cancer is associated with PM and DM
  - Anti p155/140 antibody useful in DM signalling cancer
  - Keep looking for 3 years, less likely after 5 years
- Amyopathic Dermatomyositis does occur
- Not all CPK elevations are Idiopathic Inflammatory Myositis
- Always consider metabolic, infectious, toxins, medications, and dystrophies
- Statins – IM but non inflammatory necrotizing myopathy
SUMMARY

- Do appropriate testing
  - CPK, aldolase, liver enzymes, EMG MRI ultrasound Biopsy
  - Use the myositis specific antibodies (Transfer RNA’s, Anti Mi-2, Anti-SRP)
- Most IIM responds to therapy (exception is Inclusion Body Myositis)
  - Corticosteroids Methotrexate -Beware steroid myopathy
  - Keep in mind the GOALS of therapy
- Use Consultants - Rheumatology Neurology Pathology
  - Dermatology and Rehabilitation

Thank you for your attention!
Any Questions?