

Myositis Overlaps

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What is Myositis Overlap?

- *When a patient has both myositis and another autoimmune disease*
- Autoimmune diseases occur when problems develop from the immune system attacking its own body
- Some autoimmune diseases that also occur in myositis patients include:
 - Rheumatoid arthritis
 - Lupus (systemic lupus erythematosus)
 - Scleroderma (systemic sclerosis)
 - Autoimmune thyroid disease (Hashimoto's or Grave's disease)
 - Sjogren's syndrome (dry eyes and dry mouth)
 - Vitiligo (loss of skin pigment)
 - Insulin-dependent diabetes mellitus
 - Multiple sclerosis

How Do You Tell when You have a Myositis Overlap and Does it Matter?

- It can be very difficult to tell if you have an overlap disease since the myositis syndromes (also called IIM) are systemic diseases with many organs involved
- Usually you would need to meet criteria not just for myositis but also for the other disease
- Sometimes the problems of the other disease can be more troublesome than the myositis
- Since most treatments are symptomatic, so it usually doesn't make much difference if you have an overlap or not in terms of your immediate treatment
- Muscle inflammation in overlap syndromes tends to be less severe but the other problems can be worse

IIM — SYSTEMIC MANIFESTATIONS

MUSCULOSKELETAL

- Weakness
- Muscle pain/tenderness
- Muscle atrophy
- Arthralgias
- Arthritis

GASTROINTESTINAL

- Dysphagia
- Reflux
- Dysmotility

CUTANEOUS

- Rashes
- Calcification



CARDIAC

- Arrhythmias
- Congestive failure

PULMONARY

- Atelectasis from muscle weakness
- Aspiration pneumonia
- Interstitial fibrosis

GENERAL

- Fever
- Fatigue
- Weight loss
- Raynaud's

Rheumatoid Arthritis – RA

- A chronic, systemic, inflammatory disease that chiefly affects the joints in the body
- About 1% of US population affected; more common in women (about 2:1), frequent onset age 40-50
- Because the disease is systemic, there are many extra-articular features of the disease as well
- Common problems include: *neuropathy, scleritis, lymphadenopathy, pericarditis, splenomegaly, arteritis, and rheumatoid nodules, renal, pulmonary, and heart involvement*

What to expect from a RA patient

- Stiffness in the morning
- Difficulty walking
- Difficulty using hands - dressing, using a pen, opening doors
- Fatigue
- Major therapies: Prednisone, NSAIDs, Methotrexate, Etanercept, DMARDs, other novel biologic agents
- Side effects of medications - prednisone, MTX, Etanercept, etc.
 - Cushingoid, easy bruising, hypertension, diabetes, prone to infection

Systemic Lupus Erythematosus - Lupus

- Chronic (lifelong) autoimmune inflammatory disease that causes the immune system to attack the body's own tissue and organs, including the joints, kidneys, heart, lungs, brain, blood, or skin
- About 0.1% of US population affected; more common in women (9:1 F:M), frequent onset age 20s and 40s
- Common problems include: Achy joints (arthralgia); fatigue; fevers of more than 100 degrees F.; arthritis (swollen joints); skin rashes; anemia and other low blood counts and kidney involvement

What to expect from a patient with SLE

- Fatigue
- Joint pain and swelling
- Skin rashes – malar, alopecia, oral ulcers, discoid, photosensitivity
- Some with severe disease
 - Kidney disease – hypertension, edema
 - Brain disease - headaches, depression, seizures
- Treatment: prednisone, hydroxychloroquine, cyclophosphamide, azathioprine, MMF, topical steroids, sun blockers and protective clothing

Systemic Sclerosis - SSc - Scleroderma

- A chronic multi-system connective tissue disease involving fibrous thickening and hardening
- About 0.01% of US population affected; more common in women, frequent onset age 30s and 40s
- Effects may either be visible (skin), or invisible (only internal organs such as the GI, lungs, heart or kidneys)
- Common problems include: Raynaud's Phenomenon (abnormal sensitivity to cold in the extremities); pain and stiffness of the joints; thickening of the skin, joint contractures; Sjogren's syndrome (dry mucus membranes); kidney (HTN), GI (reflux, constipation) and lung (ILD, fibrosis) involvement; Non-specific symptoms such as fatigue, generalized weakness, weight loss, and vague aching of muscles, joints and bones

What to expect with Scleroderma

- Signs and symptoms:
 - Cold intolerance
 - Difficulty with the GI tract
 - reflux, constipation, diarrhea
 - Shortness of breath
 - Dryness of the mouth and eyes
 - High blood pressure (incl hypertensive crisis)
- Treatments often ineffective: D-penicillamine; ACE inhibitors (renal); low dose prednisone and NSAIDs, cytoxan (pulmonary), prostaglandin inhibitors (Epoprostenol) for pulmonary hypertension
- Prognosis: variable depending on manifestations

What Causes Myositis Overlaps?

- *We don't know!*
- Some evidence for the role of genetics – more myositis overlaps in some families, HLA and other gene associations are the same for myositis as other autoimmune diseases
- Some evidence for the role of environmental triggers – viral and bacterial infections, drugs, UV light, stress
- Evidence that the immune system (T and B cells, autoantibodies and complement) is involved
- *So overall myositis overlaps seem to result from activation of the immune system in genetically susceptible individuals following exposure to specific environmental triggers*

How do you Treat Myositis Overlaps?

- Be sure the diagnosis is correct
- Determine the amount of disease activity (active inflammation) versus disease damage (scarring)
- For disease activity, assess the risk/benefit of different drugs that block the immune system
- For disease damage, use physical therapy and exercise approaches
- Develop, with the patient's input, a holistic individualized treatment plan taking into account expectations, manifestations, prognosis and risk factors for adverse events to therapies

What are the Major Unanswered Myositis Overlap Questions?

- Which of the current treatments work best in which patients ?
- How can we predict who needs more treatment from the beginning of disease and who doesn't?
- What mechanisms induce and sustain myositis overlaps?
- What new better therapies can block these mechanisms ?
- What are the genetic and environmental risk factors that lead to myositis overlaps?
- Can we prevent some forms of myositis overlaps?

What Can I do to Help Cure Myositis and Overlaps?

- Work with TMA to raise more funds for myositis awareness and research
- Educate family, friends, neighbors and others about myositis and encourage them to write Congress to increase funding for autoimmunity research generally and myositis research specifically
- Find out from TMA about ongoing research studies and consider participating in appropriate studies