Inflammatory Myopathies

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Objectives

- Recognized general classes of muscle disease in order to identify treatable disorders
- Understand the evolution of the classification of Idiopathic Inflammatory Myopathies (IIMs) and the implications on diagnostic workup
- Utilize the clinico-sero-histopatholigic features to accurately diagnose IIMs
- Identify potential initial therapeutic options based on the subclass of IIM

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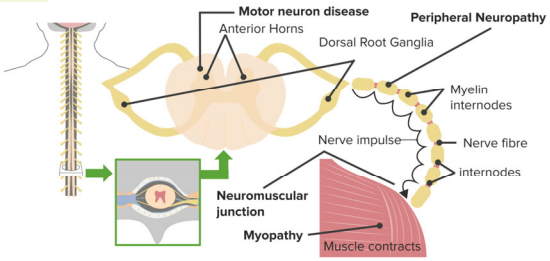
Localization



Disorders along this path are in the differential diagnosis and often misdiagnosed including disorders of:

- Motor neuron
- Myelination and Axon (nutrition)
- Neuromuscular junction disorder

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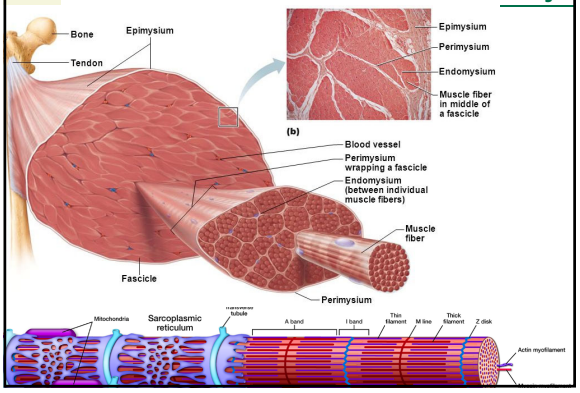
Muscle Anatomy



Myopathy – Disease of Muscle

INHERITED

- Muscular dystrophy – progressive disease due to malformation in muscle proteins (Duchenne, Becker, Limb Girdle, Facioscapulohumeral, Emery-Dreifuss , myofibrillar)
- Congenital – nonprogressive disease, often presenting at birth, distinct from muscular dystrophy (nemaline rod, centronuclear, central core)
- Myotonic – Dystrophic and non-Dystrophic (channelopathies: myotonia congenita, paramyotonia congenita, Potassium aggravated myotonia and periodic paralysis)
- Metabolic
  - Metabolism, Storage/Accumulation – glycogen (acid maltase, phosphorylase, debrancher deficiencies), lipid (carnitine), amyloidosis
  - Endocrine – thyroid, parathyroid, adrenal, pituitary, diabetes
- Mitochondrial – progressive external ophthalmoplegia, Kearns-Sayer

ACQUIRED

- Infectious – HIV, HTLV, Toxoplasmosis, Trichinosis
- Toxic – statin, alcohol, amiodarone, colchicine, antivirals, checkpoint inhibitors
- **Inflammatory – Dermatomyositis, Anti-Synthetase, Immune-Mediated Necrotizing Myopathy (IMNM), Inclusion Body Myositis,** +/-Polymyositis

Evolution of Classifications

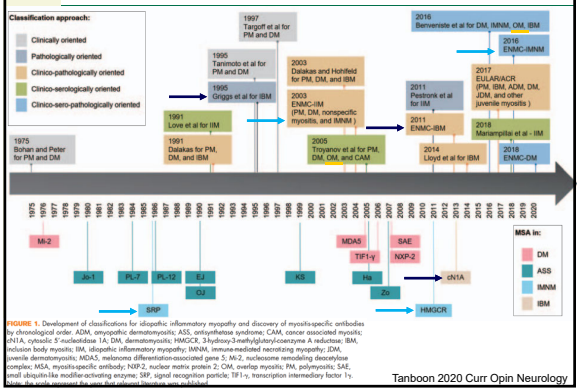


FIGURE 1. Development of classifications for idiopathic inflammatory myopathy and discovery of myositis-specific antibodies by chronological order. ADM, amyopathic dermatomyositis; ASS, antisynthetase syndrome; CAM, cancer associated myositis; cN1A, cytosolic 5′-nucleotidase 1A; DM, dermatomyositis; HMGCR, 3-hydroxy-3-methylglutaryl-coenzyme A reductase; IBM, inclusion body myositis; IIM, idiopathic inflammatory myopathy; IMNM, immune-mediated necrotizing myopathy; JDM, juvenile dermatomyositis; MDA5, melanoma differentiation-associated gene 5; Mi-2, nucleosome remodeling deacetylase complex; MSA, myositis-specific antibody; NXP-2, nuclear matrix protein 2; OM, overlap myositis; PM, polymyositis; SAE, small ubiquitin-like modifier-activating enzyme; SRP, signal recognition particle; TIF1-γ, transcription intermediary factor 1γ.

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History and Exam

- Motor predominant symptoms
  - Sensory symptoms mild with comorbidities or absent
- Pattern of weakness (proximal +/-distal, asymmetry, facial)
  - Difficulty rising from a chair, combing hair
  - Common clues and complaints:
    - Dysphagia
    - Shortness of breath
    - Cramps and myalgia
    - Exertional fatigue
  - Flags for inherited disorders
    - Rhabdomyolysis – tea-colored urine
    - Eye movement abnormalities
    - Developmental delay
    - Reportedly "less athletic" growing up
    - Family history

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Workup

- **HISTORY!!!**
- Exam
- Initial laboratory testing
  - **Creatine Kinase (CK),** aldolase, CMP, +/-GGT, Vitamins (D, B1), ESR, CRP, ANA, Endocrine (TFTs, PTH, Cortisol), Urine Drug Screen
    - CK can be normal, mildly elevated or significantly elevated (clue)
- Nerve conduction studies / Electromyography
    - Reduced motor amplitudes
    - Myopathic +/- mixed neurogenic
    - Insertional activity - Irritable versus non-irritable myopathies

Source: Zahid H. Bajwa, R. Joshua Wootton, Carol A. Warfield: *Principles and Practice of Pain Medicine*, 3rd Edition
www.accessanesthesiology.com

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Myopathies with Irritability on EMG

INHERITED

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- Congenital – nonprogressive disease, often presenting at birth, distinct from muscular dystrophy (nemaline rod, centronuclear, central core)
- Myotonic – Dystrophic and non-Dystrophic (channelopathies: myotonia congenita, paramyotonia congenita, Potassium aggravated myotonia and periodic paralysis)
- Metabolic
  - Metabolism, Storage/Accumulation – glycogen (acid maltase, phosphorylase, debrancher deficiencies), lipid (carnitine), amyloidosis
  - Endocrine – thyroid, parathyroid, adrenal (HyperK+ PP), pituitary, diabetes
- +/-Mitochondrial – progressive external ophthalmoplegia, Kearns-Sayer

ACQUIRED

- Infectious – HIV, HTLV, Toxoplasmosis, Trichinosis, **Critical illness myopathy
- Toxic – statin, alcohol, amiodarone, colchicine, antivirals, checkpoint inhibitors
- **Inflammatory – Dermatomyositis, Inclusion Body Myositis, Anti-Synthetase, Immune-Mediated Necrotizing Myopathy (IMNM),** +/-Polymyositis

Workup

- More extensive laboratory testing
  - Rheumatologic, infections studies, electrophoresis, immunofixation
  - **Myositis antibody panels**
    - ELISA/radioimmunoassay (**Mayo**) vs. Western Blot (WashU, Quest, ARUP)
    - Myositis Specific Antibodies (MSA) - Specific but lower sensitivity
    - Myositis Associated Antibodies (MAA) – Seen in overlap syndromes with connective tissue disease as with lupus, scleroderma, and rheumatoid arthritis
    - **Add-on MSA antibodies:**
      - **Anti-HMGCR** – with high CK (necrotizing)
      - **Anti-cN1A (NT5C1A)** – indolent (IBM – cytoplasmic 5'-nucleotidase)
  - +/- Genetic Panels and metabolic profiles
    - Free (Comprehensive NM Disorders) versus ~$250 self-pay focused panels
- Muscle Biopsy - If necessary, preferably prior to treatment
- CT chest abdomen pelvis with contrast
- RF Esophogram, EKG, Echo, PFTs

Antibodies – MSA and MAA

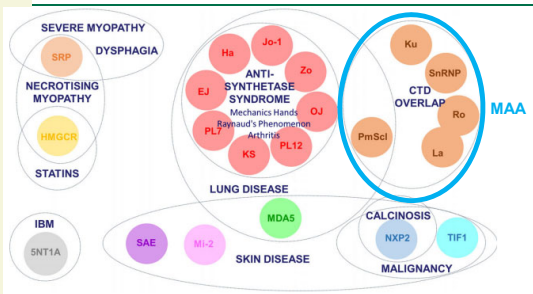

Fig. 2 *Myositis autoantibodies and their key clinical associations. IBM, inclusion body myositis; CTD, connective tissue disease; SRP, signal recognition particle; HMGCR, 3-hydroxy-3-methylglutaryl-coenzyme A reductase; TIF1, transcription intermediary factor 1; NXP2, nuclear matrix protein 2; MDA5, melanoma differentiation-associated gene 5; SAE, small ubiquitin-like modifier activating enzyme; 5NT1A, cytosolic 5'nucleotidase 1A; Mi-2, nucleosome-remodelling deacetyalse complex; Jo-1, histidyl tRNA synthetase; PL7, threonyl tRNA synthetase; PL12, alanyl tRNA synthetase; OJ, isoleucyl tRNA synthetases; EJ, glycyl tRNA synthetase; KS, asparaginyl tRNA sythetase; Zo, phenylalanyl tRNA synthetase, Ha; tyrosyl tRNA synthetase; snRNP, small nuclear ribonucleic protein.*

Betteridge 2016 J Int Medicine

Myositis Antibody Frequency

Patients with inflammatory myopathies	$n = 207$
Negative for anti-ribonucleoprotein (RNP) autoantibodies	108 (52%)
Positive for anti-RNP autoantibodies	99 (48%)
Anti-signal recognition particle (SRP)	41 (20%)
Anti-aminoacyl transfer RNA synthetase (ARS)	23 (11%)
Anti-OJ	8 (4%)
Anti-Jo-1	7 (3%)
Anti-EJ	4 (2%)
Anti-PL-7	3 (1%)
Anti-PL-12	1 (0.5%)
Anti-KS	0 (0%)
Anti-Ku	10 (5%)
Anti-U1RNP and anti-Sm	14 (7%)
Anti-U2RNP	1 (0.5%)
Anti-U5RNP	1 (0.5%)
Anti-ribosome	2 (1%)
Anti-Th/To	1 (0.5%)
Anti-SS-A/Ro and anti-SS-B/La	27 (13%)
Coexistence of two autoantibodies	20 (10%)
Coexistence of three autoantibodies	3 (1%)

Suzuki 2014 J Immunology

ANA Patterns

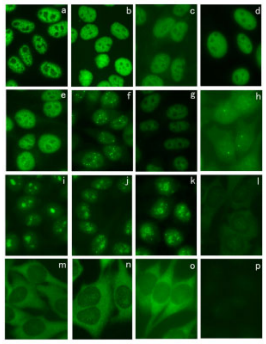

Fig. 3.
Immunofluorescence antinuclear antibodies using sera from patients with PM/DM. HEp-2 ANA slides were stained using sera from patients with PM/DM. **a** Anti-U1RNP, **b** anti-Mi-2, **c** anti-TIF1γ/α, **d** anti-TIF1β, **e** anti-SAE, **f**, **g** anti-MJ/NXP-2, **h** anti-SMN, **i**, **j** anti-PM-Scl, **k** anti-U3RNP, **l** anti-Jo-1, **m** anti-PL-7, **n** anti-PL-12, **m** anti-SRP, **p** anti-MDA5

Satoh 2018 Clin Rev Allergy Immunology

Evolution of Classifications Dermatomyositis (DM)

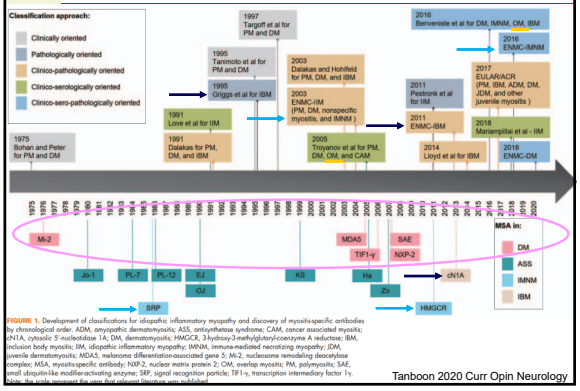

FIGURE 1. Development of classifications for idiopathic inflammatory myopathy and discovery of myositis-specific antibodies by chronological order. ADM, amyopathic dermatomyositis; ASS, antisynthetase syndrome; CAM, cancer associated myositis; cN1A, cytosolic 5′-nucleotidase 1A; DM, dermatomyositis; HMGCR, 3-hydroxy-3-methylglutaryl-coenzyme A reductase; IBM, inclusion body myositis; IIM, idiopathic inflammatory myopathy; IMNM, immune-mediated necrotizing myopathy; JDM, juvenile dermatomyositis; MDA5, melanoma differentiation-associated gene 5; M-2, nucleosome remodeling deacetylase complex; MSA, myositis-specific antibody; NXP-2, nuclear matrix protein 2; OM, overlap myositis; PM, polymyositis; SAE, small ubiquitin-like modifier-activating enzyme; SRP, signal recognition particle; TIF1-γ, transcription intermediary factor 1γ. [Note, the circle represent the year that relevant literature was published]

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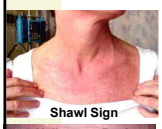
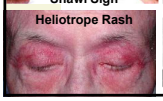
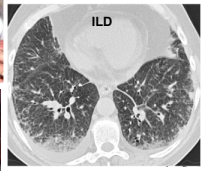
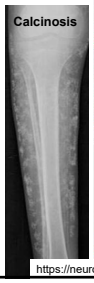
Dermatomyositis (DM)

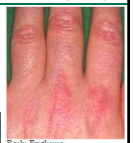
- Weeks-months, symmetric proximal & axial muscle weakness, spares face
- Female : Male 2:1, any age of onset
- Skin, subcutaneous involvement, +/-ulcers
- Interstitial lung disease (ILD)
- CK: **normal** – 50xULN

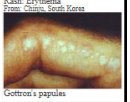

Shawl Sign

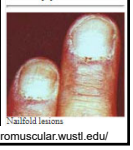

Heliotrope Rash






Rash: Erythema
From: China, South Korea


Gottron's papules


Nailfold lesions

https://neuromuscular.wustl.edu/

Dermatomyositis Associated Antibodies

- TIF1 gamma (Transcriptional Intermediary Factor 1-gamma)
  - >50% **cancer** (pancreas, stomach, intestinal, lymphoma) in 3 years
  - Severe skin disease, subcutaneous edema, calcinosis, ILD, dysphagia
- NXP2 (Nuclear matriX Protein 2)
  - Mostly Juvenile DM (15%), associated with **cancer** in adults
  - Severe onset, calcinosis with subcutaneous edema
- Mi-2
  - Acute severe rash, weakness with high CK, responds well to therapy
- **MDA 5** (Melanoma Differentiation Associate protein)
  - Can be **AM**yopathic with ulcerating skin lesions
  - Rapidly progressive **ILD with high mortality**
- SAE1 (small ubiquitin-like modifier Activating Enzyme)
  - Dysphagia common, later onset myositis
- Antibody negative – classic DM

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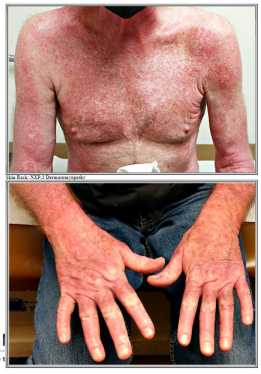
Skin Changes



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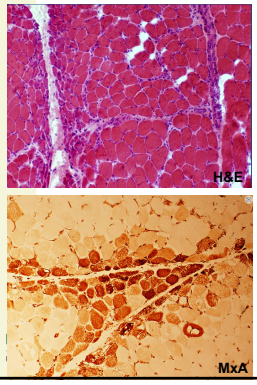
Dermatomyositis Histopathology



- Perifascicular atrophy (PFA)
- Reduced capillary density
- Type 1 Interferon pathway
  - Myxovirus resistance protein A (MxA) positivity
  - CD4+ infiltrates
- Complement activation (C5b-9)
  - Sarcolemma
  - Capillaries

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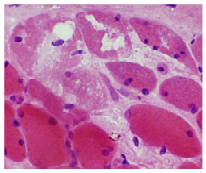
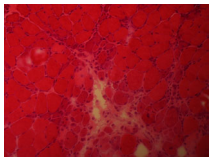
Dermatomyositis Histopathology

- Classic, SAE1 and antibody negative DM
  - Perifascicular atrophy
  - MxA positivity
- TIF1 gamma
  - Punched out vacuoles
  - Mild perivascular lymphocytic infiltrates
- NXP2
  - Microinfarction
- Mi-2
  - Perifascicular necrosis
  - Perimysial connective tissue fragmentation
  - Clusters of B cells
- MDA 5
  - No PFA / normal or nonspecific
  - Scattered sarcoplasmic MxA staining





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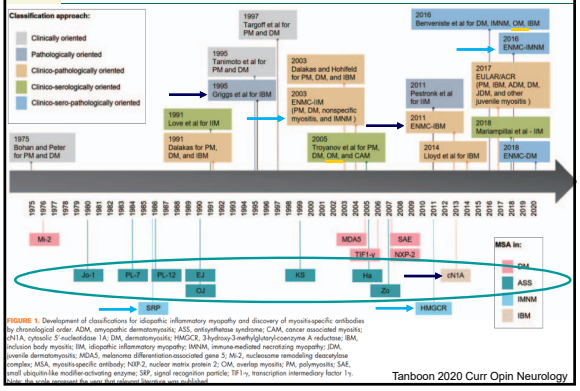
Anti-synthetase syndrome (ASS/ASyS)



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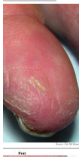
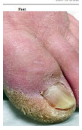
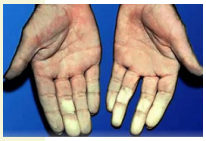
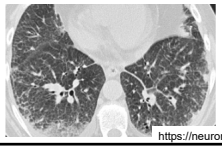
Anti-Synthetase Syndrome

- Weeks-months, symmetric proximal & axial muscle weakness, spares face
- Female : Male 2:1, average age of onset 50
- Extra muscular manifestations:
  - Raynaud's
  - Interstitial lung disease (ILD)
  - Mechanics hands
  - Arthritis
  - Fever









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Anti-Synthetase Syndrome

- Amino acid charging enzymes - Aminoacyl tRNA synthetase subtypes:
  - Jo-1 - Myositis > ILD, classic ASyS syndrome
    - White predominance
    - Responds well to therapy
  - PL-7 & 12 - ILD > myositis
    - Milder than Jo-1
    - More severe in black
    - PL-12 associated with malignancy
  - OJ - Severe myositis early, ILD, skin lesions
  - Ks - ILD, myositis in ~25%, skin lesion
    - Japanese predominance
  - Ha and Zo – Myositis, ILD (Rare)

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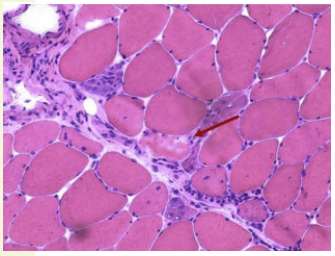
Anti-Synthetase Histopathology



- Perifascicular atrophy
- Myofiber necrosis
- Regenerating myofibers
- Perimysial connective tissue fragmentation

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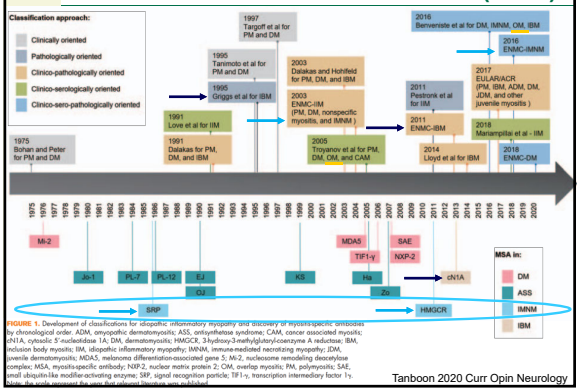
Immune Mediated Necrotizing Myositis (IMNM)



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Immune Mediated Necrotizing Myositis

- Weeks- months of proximal lower > upper limb weakness +/- atrophy
- Female : Male 2:1
- CK >1,000 (Average 6000-9000)
- Subtypes:
  - Anti-HMGCR **self-limited statin toxicity will not have antibodies
    - 2/3 statin exposure
    - Severe in younger (18mo), milder in elderly
    - Minimal extra-muscular manifestations
  - Anti-SRP (signal recognition protein)
    - Acute, aggressive, CK >10,000
    - Poor response to treatment
    - +/- Dilated cardiomyopathy and ILD
  - Antibody negative – cancer association

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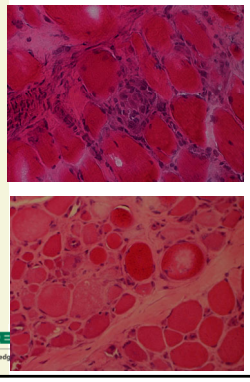
IMNM Histopathology



- Myofiber necrosis with phagocytosis
- Myofiber regeneration
- Little to no inflammation
- Increased endomysial connective tissue

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Inclusion Body Myositis (IBM)

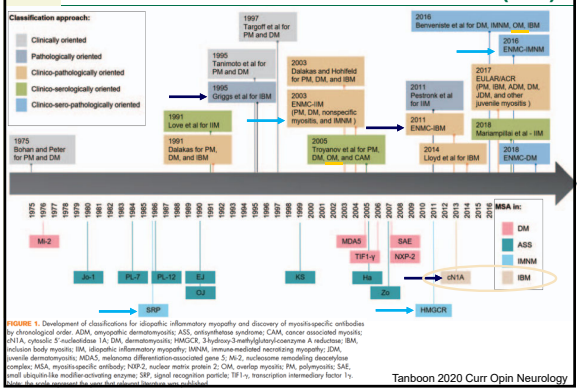
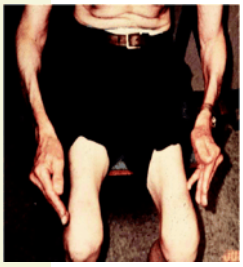


FIGURE 1. Development of classifications for idiopathic inflammatory myopathy and discovery of myositis-specific antibodies by chronological order. ADM, amyopathic dermatomyositis; ASS, antisynthetase syndrome; CAM, cancer associated myositis; cN1A, cytosolic 5′-nucleotidase 1A; DM, dermatomyositis; HMGCR, 3-hydroxy-3-methylglutaryl-coenzyme A reductase; IBM, inclusion body myositis; IIM, idiopathic inflammatory myopathy; IMNM, immune-mediated necrotizing myopathy; JDM, juvenile dermatomyositis; MDA5, melanoma differentiation-associated gene 5; Mi-2, nucleosome remodeling deacetylase complex; MSA, myositis-specific antibody; NXP-2, nuclear matrix protein 2; OM, overlap myositis; PM, polymyositis; SAE, small ubiquitin-like modifier-activating enzyme; SRP, signal recognition particle; TIF1-γ, transcription intermediary factor 1γ.

Tanboon 2020 Curr Opin Neurology

Inclusion Body Myositis



- Most common acquired IIM over 50 years of age
- Male : Female 3:1
- Indolent, progressive
- Weakness of:
  - Knee extension > hip flexion
  - Finger flexion > shoulder abduction
  - Dysphagia in 1/3
  - +/- Orbicularis oculi
- Pathogenicity: degenerative and inflammatory signaling
- **Fails to respond to immune-modulating therapies**

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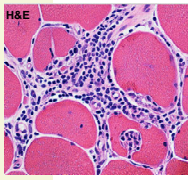
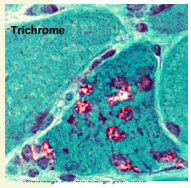
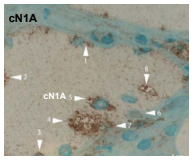
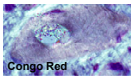
Sakai, K 2015 Neuromuscul Disord

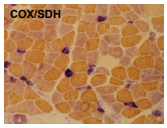
IBM Histopathology







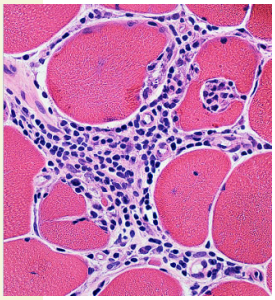




- CD8+ T cells surround and invade
- Rimmed vacuoles
- Anti-cytoplasmic 5'-Neucleotidase-1A (anti-cN-1A or NT5c1A)
- Inclusions: amyloid, p62, tubulofilaments
- Mitochondrial pathology

Kramp 2016 AutoImmun Highlights
https://neuromuscular.wustl.edu/

IBM Histopathology



- **On biopsy, can:**
  - **PM be diagnosed?**
  - **IBM be excluded?**

NO

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https://neuromuscular.wustl.edu/

Antibodies – MSA and MAA

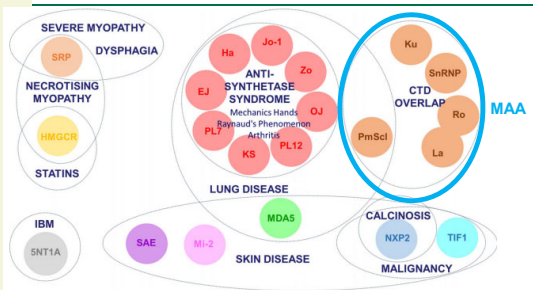

Fig. 2 *Myositis autoantibodies and their key clinical associations. IBM, inclusion body myositis; CTD, connective tissue disease; SRP, signal recognition particle; HMGCR, 3-hydroxy-3-methylglutaryl-coenzyme A reductase; TIF1, transcription intermediary factor 1; NXP2, nuclear matrix protein 2; MDA5, melanoma differentiation-associated gene 5; SAE, small ubiquitin-like modifier activating enzyme; SNT1A, cytosolic 5′nucleotidase 1A; Mi-2, nucleosome-remodelling deacetylase complex; Jo-1, histidyl tRNA synthetase; PL7, threonyl tRNA synthetase; PL12, alanyl tRNA synthetase; OJ, isoleucyl tRNA synthetases; EJ, glycyl tRNA synthetase; KS, asparaginyl tRNA sythetase; Zo, phenylalanyl tRNA synthetase, Ha; tyrosyl tRNA synthetase; snRNP, small nuclear ribonucleic protein.*

Betteridge 2016 J Int Medicine

Myositis Associated Antibodies (MAAs)

- SSA/Ro-60 – Most associated with IIMs, not known to cause weakness
- PM SCL – Myositis, sclerodermatous skin changes, DM rash, +/- ILD
- Myositis with mixed connective tissue disease
  - U1 RNP
  - Ro-52 – Commonly occurs with other antisynthetase antibodies
  - Ku – Commonly occurs with other MSAs

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McHugh 2018 Nature
Menendez 2013 Sci World Journ

Mainstay of IIM Treatment

- ****IBM – currently no recommended treatment**
- Treat associated diseases and underlying cancer
- **High-dose corticosteroids**
  - Reduces morbidity and improve strength
  - Clinical improvement within days to months
    - DM: 58-100% partially respond, 30-66% respond completely
  - **Prevent side effects**: Infection, hypertension, hyperglycemia, weight gain/type 2 muscle atrophy, cataracts, osteoporosis
- Regimen:
  - IV solumedrol 1g x3-5d if requiring hospitalization
  - Prednisone 0.75-1.5mg/kg/day up to 60mg/day **maintained until significant clinical improvement and CK normalizes**
  - Taper **no faster than 10mg per 1-3 months to 20mg daily**, then 2.5-5mg/month as tolerated

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Steroid Sparing Agents

- **Common Side effects**: Diarrhea, nausea, fever, cytopenia, malignancy (lymphoma, skin), leukoencephalopathy, infection reactivation
- **Vaccinations**:
  - Provide at least 2 weeks prior to starting or re-vaccinating
  - May be less effective
  - Avoid live vaccinations
- Azathioprine (Imuran)
  - 50mg/d, 2-3mg/kg (incr 50mg Q1-2mo); onset 9-24 months
  - Flu-like (*do not rechallenge), hepatic, bone marrow failure (TPMT)
  - CBC, CMP Qwk x1mo, Qmo x 1year, then Q6mo; Dermatology Qyr
  - Consider in women in a **child-bearing age**
- Mycophenolate mofetil (CellCept)
  - 500mg BID, 1.5g BID (incr 500mg BID Q2-4wks); onset 6-24 months
  - Teratogenicity (2 forms of birth control)
  - CBC Qwk x1mo, Qmo x 1year, then Q6mo; Dermatology QYr
  - May be better with **interstitial lung disease** (ILD)

Next Line Agents

- Methotrexate
  - 5mg/wk, 15-60mg/wk (incr 2.5-5mg/wk, IM >20mg); Onset month
  - Stomatitis (folic acid), renal, hepatic, teratogenicity, ILD (avoid)
  - CBC, CMP Q3mo; Liver US >1g cumulative dose; Chest x-ray and PFTs;
  - Consider when **refractory to prednisone**
- Intravenous immunoglobulin (IVIG)
  - 2g over 3-5d, 1g Q3-6wk; onset 2 days – 2 weeks;
  - Infusion reaction, renal failure, thrombosis, hemolytic anemia, headache
  - CBC, CMP with infusions or Q3mo
  - Consider early and/or as monotherapy with **anti-HMGCR** antibodies
- Rituximab
  - 1g day1 and 15, then ~Q6mo PRN; Onset 2-6 weeks
  - Infusion reaction, Hepatitis B reactivation (GI referral)
  - Consider early with **anti-SRP** and **anti-synthetase with severe ILD**
- Other: Tacrolimus, cyclosporine, cyclophosphamide, PLEX
- Upcoming: JAK, interferon, & complement inhibitors, FcRN blockade

Supportive Measures

- Therapies
  - Physical and Occupational with **assistive device assessment**
  - Speech/Swallow Therapy
- Health with steroids
  - DEXA scan, Vitamin D, Calcium, PPI, +/-Alendronate
  - Bactrim DS MWF (prednisone > 20mg +/- steroid-sparing agent)
- Referrals
  - Dermatology – skin cancer
  - Ophthalmology – cataract, retinopathy
  - Rheumatology – skin and joint involvement
  - Pulmonology – ILD
  - Cardiology – cardiomyopathy
  - Endocrinology – glucose control and bone health
  - Gastroenterologist – esophageal dilation +/- myotomy
  - PCP, Dietician, Weight Loss Clinic
  - Hematology/Oncology – with concern for cancer

Questions?

Thank You!

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