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Acquired Inflammatory Myopathies

Bassam A. Bassam, MD, FAAN
University of South Alabama

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Disclosures

- **Argenx Pharma Speaker Bureau member**
  (No relation to, or conflict with this presentation)

Objectives

Acquired Inflammatory Myopathies

- Review of clinical features of inflammatory myopathy.
- List similarities and differences of inflammatory myopathy subtypes.
- Discuss the pathological features of inflammatory myopathy.
- Review management and prognosis of inflammatory myopathy

Case 1.

50 Y/O W/F complains of upper limbs and neck weakness for 2 years, with no known inciting event. She has mild swallowing difficulty. Her weakness is constant, more notable with reaching for objects above her shoulders. She attributed her symptoms to an MVA whiplash. She has mild neck ache, but denies arms pain or sensory disturbances, diplopia or ptosis.

Negative PMHx and FMHx positive for CAD

Neurological examination; Normal cognitive functions and CNs exam.
4/5 weakness of neck flexion, and 4/5 weakness of arms abduction.
No lower limbs muscle weakness, mild difficulty to arise from a chair unassisted.
DTR's are symmetric +2, no pathological reflexes. Normal coordination and Gait.

A most likely differential Diagnosis is;

- Motor neuron disease
- Polymyositis
- Toxic myopathy
- Inclusion body myositis
- Cervical radiculopathy
- Myasthenia Gravis

Polymyositis

CBC, T4, TSH, B12, folate were normal. Negative AchR antibodies. Normal brain MRI. Cervical MRI; mild degenerative changes.

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Next step of diagnostic study should be?

A. Ice-Pack test
B. Serum muscle enzymes
C. Electrodiagnostic study
D. Single Fiber EMG
E. B and C

B and C

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Electrodiagnostic Study;

- **Motor conduction study;** median, ulnar & fibular nerve all normal.
- **RMNS;** of ulnar & spinal accessory facial nerve at 3Hz no decrement.
- **Sensory conduction study;** median, ulnar, sural nerve all normal.
- **NEMG ;** Increased IA, Fib & PW, early recruitment and polyphasic short duration small amplitude MUP's in proximal muscles.
- **Muscle enzymes;** serum CPK 1686 mcg/L, serum aldolase 14.5 U/L

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The electrodiagnostic study findings are most consistent with;

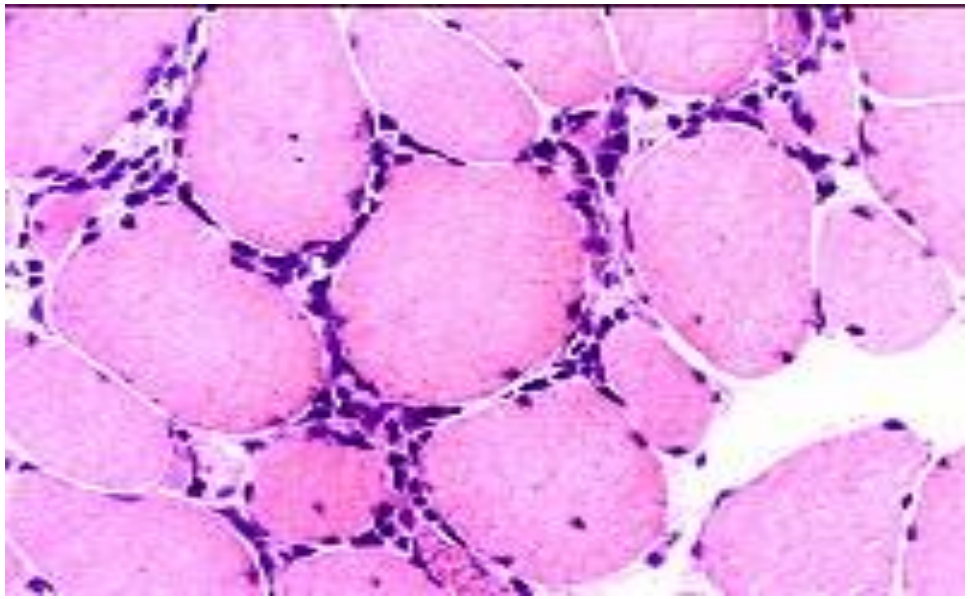
- Myotonic dystrophy
- Axonal motor neuropathy/neuronopathy
- Inflammatory myopathy
- Myasthenia Gravis

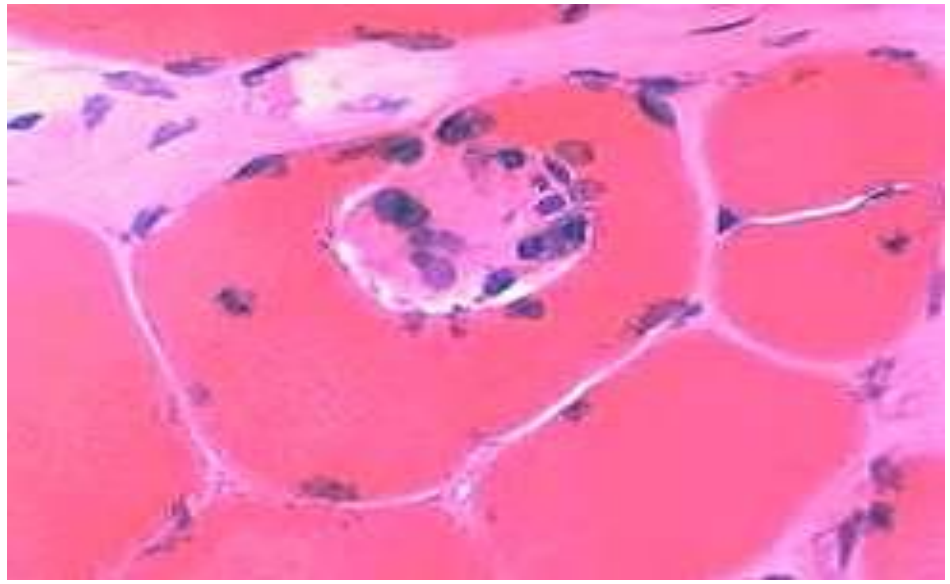
Inflammatory myopathy

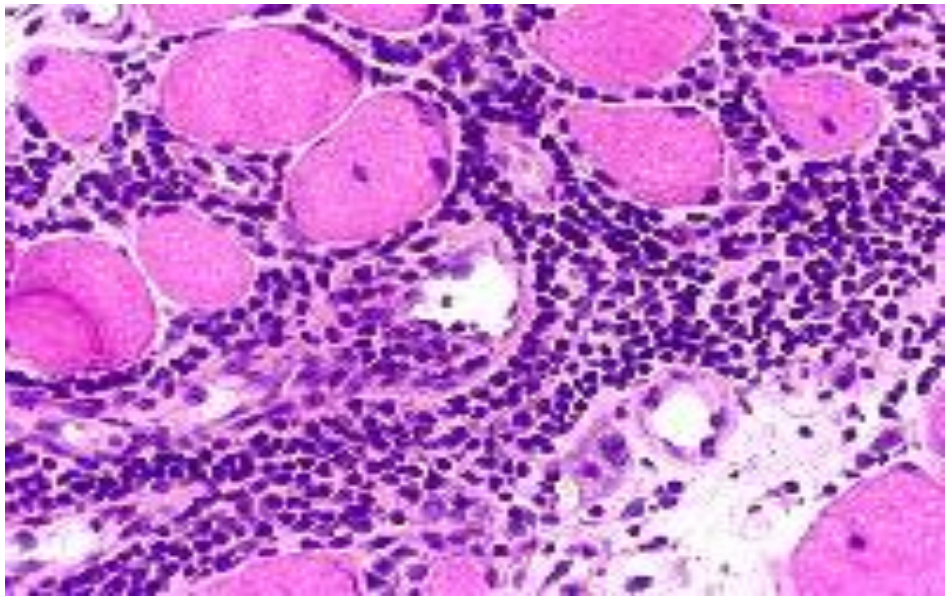
A specific diagnosis can best be achieved by;

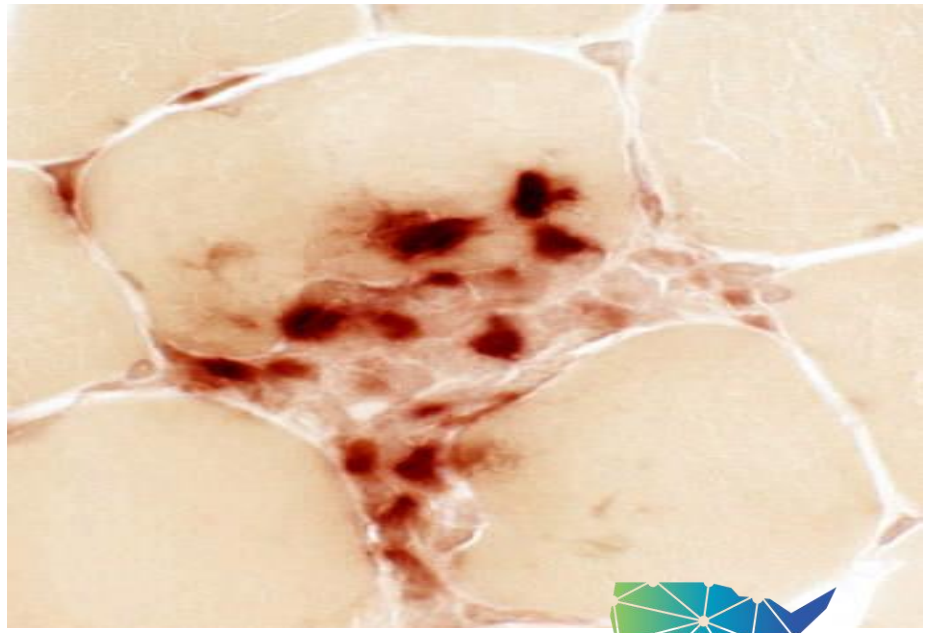
- Muscle ultrasound
- Muscle MRI
- Muscle biopsy
- Genetic DNA test
- Muscle and nerve biopsy

Muscle Biopsy









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Case 2

- A 56 Y/O male with 6 years history of progressive, painless weakness and muscle atrophy of forearms, hands and lower limbs. Gradually he became unable to rise from a squat or to climb stairs and his grip strength decreased. He developed increasing swallowing difficulty, but denies shocking.
  He has no sensory complaints, rash, cardiac or pulmonary symptoms.
- **PMHx;** Hypertension, DM-II and prior ETOH abuse.
- **FMHx;** noncontributory.

- **Examination;** Normal mentation and cranial nerves.

  Motor strength; biceps and triceps 4+/5, wrist flexors 4/5, fingers long flexors 3+/5, hip flexors 4+/5, quadriceps 3+/5, and TA 3+/5 on MRC scale.
  Normal sensation and DTR's, except **absent knee reflex bilaterally.**
  He has difficulty arising from the chair and has a mild waddling gait.

Initial Differential Diagnosis may include all below, except?

A. Motor neuron disease
B. Polymyositis
C. ETOH induced myopathy
D. Inclusion body myositis
E. Diabetic polyradiculopathy
F. Sporadic Limb Girdle Muscular Dystrophy

Diabetic Polyradiculopathy

CBC, serum chemistry, TSH, B12, folate, SPEP, ESR, heavy metals screen, Lyme disease antibodies & CSF studies all were normal.
CPK 682 IU/L, negative anti-GM1 & specific myositis autoantibodies.

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- **Next step of diagnostic study should be?**
  - Muscle ultrasound
  - Muscle biopsy
  - Electrodiagnostic study
  - Muscle MRI
  - Ischemic exercise test

Electrodiagnostic Study

• Nerve conduction;

Sensory;

Normal study of right median and bilateral ulnar nerve, mildly reduced SNAP amplitude of sural nerve bilaterally.

Motor;

Border line CMAP amplitude of bilateral fibular nerve normal right median and ulnar motor conduction study.

Needle EMG;

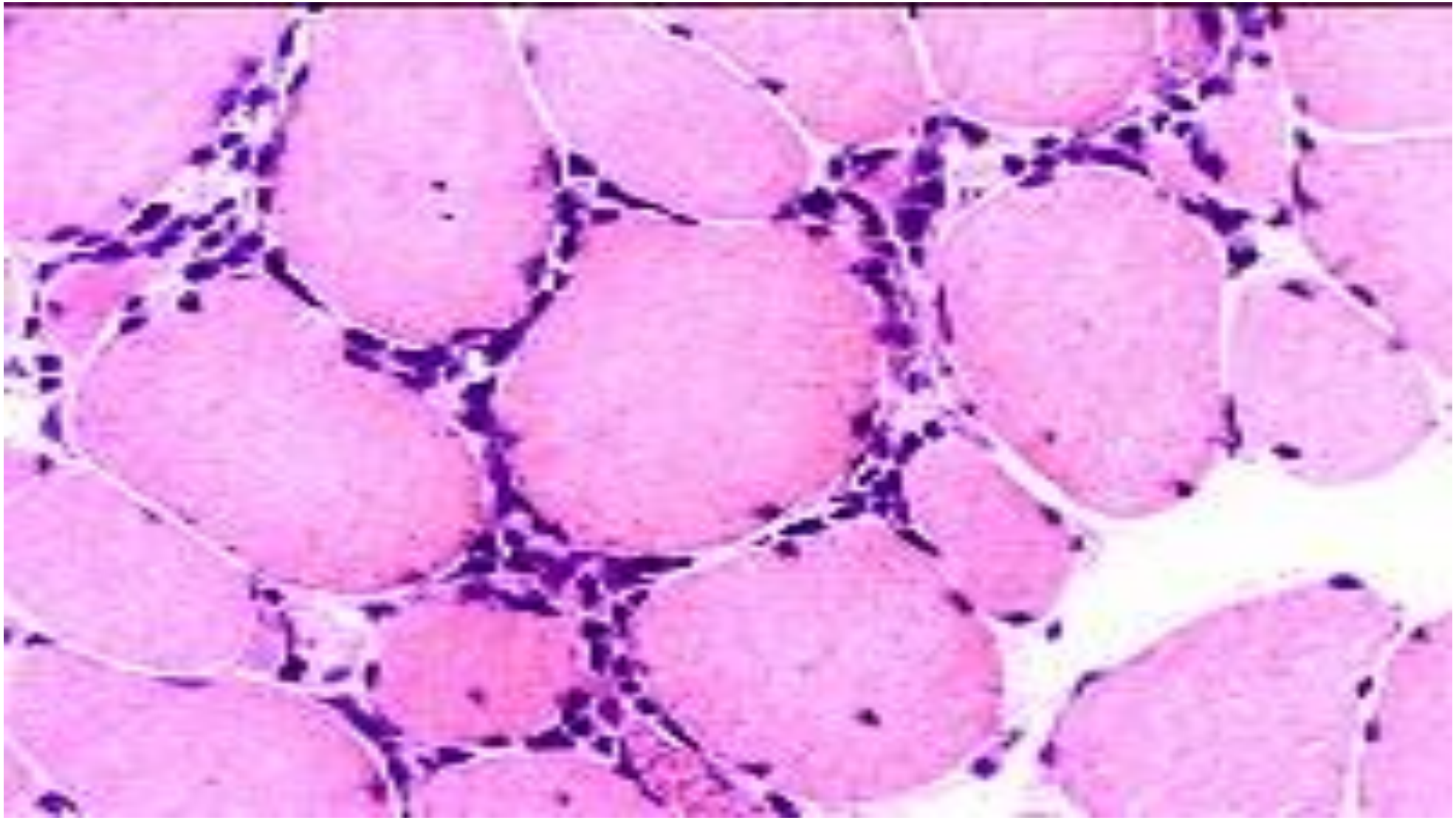
Increased IA, Fib/Pw and brief, small, abundant motor-unit action potentials (BSAP) most prominent in FCU, FDP, RF, TA. Few large MUPs in TA and FDI.

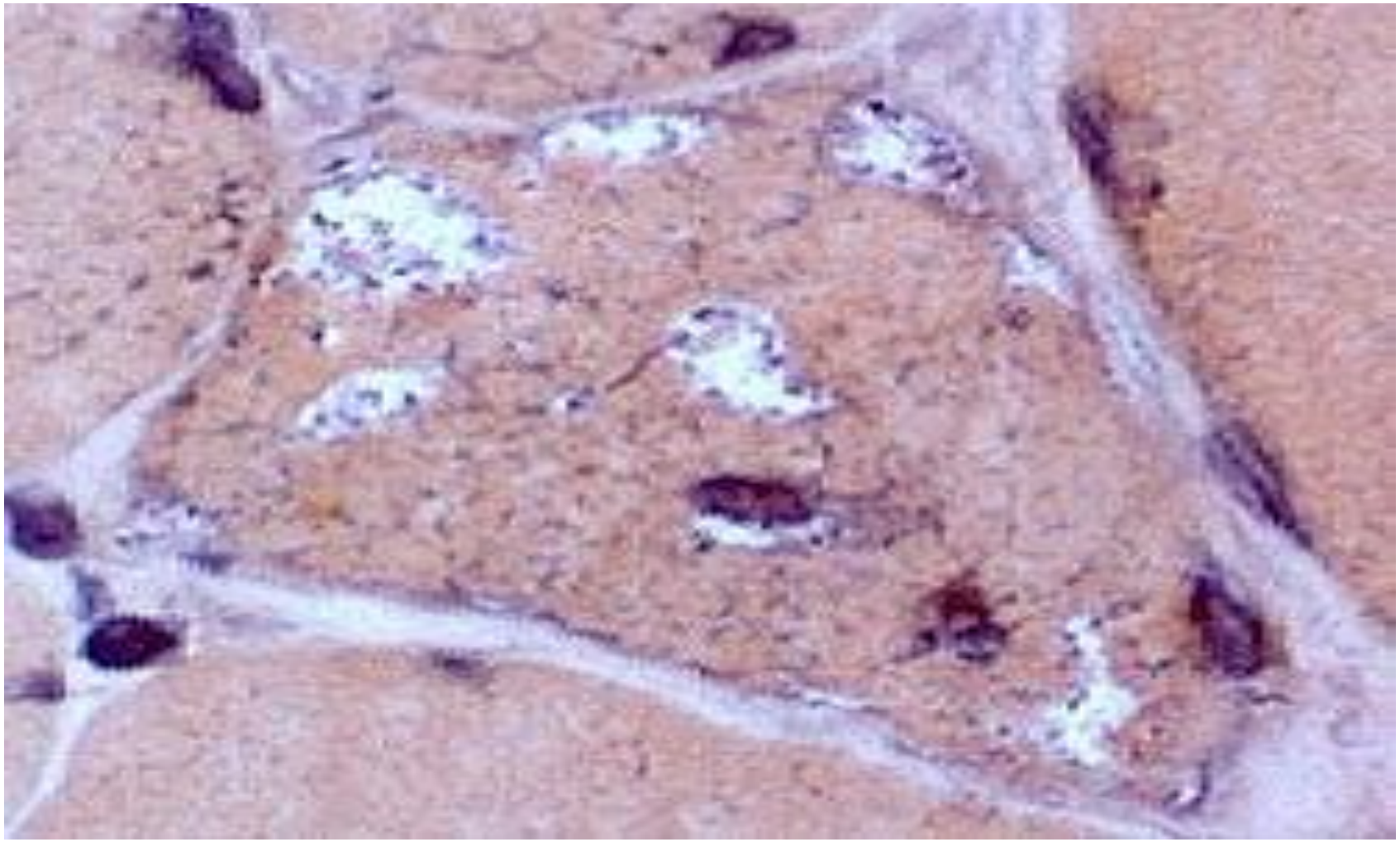
Muscle	Fb/PW	Fas	Amp.	Dur.	Poly Sm	Poly Lg	Rec.
Deltoid	+	0	↓	↓	++	0	early
Biceps	+++	0	↓	↓	+++	+	early
FCU	+++	0	↓	↓	+++	0	early
Fst. DI	+	0	↓	↓	++	+	early
Ext dig.	++	0	↓	↓	++	0	early
Gast.	++	0	↓	↓	+	0	normal
TA	+++	0	↓	↓	+++	+	early
Rec. F	+++	0	↓	↓	+++	0	early
Psoas	++	0	↓	↓	++	0	early
Ab.Hall L	+	0	N	N	0	0	normal

A specific diagnosis can best be achieved by;

- Muscle ultrasound
- Muscle MRI
- Muscle biopsy
- Genetic DNA test
- Muscle and nerve biopsy

Muscle Biopsy





The biopsy findings is consistent with diagnosis of;

- Mitochondrial myopathy
- Polymyositis
- Dermatomyositis
- Inclusion body myopathy
- Limb girdle muscular dystrophy
- Necrotizing myopathy

Inclusion body myopathy (IBM)

Acquired Inflammatory Myopathies

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LEARNING OBJECTIVES

- Review of clinical features of inflammatory myopathy.
- List similarities and differences of inflammatory myopathy subtypes.
- Discuss the pathological features of inflammatory myopathy.
- Review management and prognosis of inflammatory myopathy.

Acquired Inflammatory Myopathies

- **Polymyositis (PM)**
- **Dermatomyositis (DM)**
- **Inclusion Body Myositis (IBM)**
- **Necrotizing myopathy (NM)**

  - Familiarity with the clinical characteristics allows early accurate diagnosis
  - PM, DM and NM are eminently treatable
  - Ultimate outcome depends on early diagnosis & treatment
  - Several different myopathic or neurogenic disorders may resemble PM, DM, NM or IBM

General Features;

- PM & DM are characterized by subacute (weeks to months), progressive proximal weakness
- NM has an acute or subacute proximal weakness, more rapid and more severe course
- Bimodal age distribution of DM (5-14 years & ADM), PM 45-65 years.
- PM incidence in USA is 5 -10/million/year, women > men (2 to 1), black> white
- IBM progress slowly (average 6 years), male predominance, white > black
- IBM is most common IMs in patients over age 50 years (>30% of IMs cases referred to a tertiary care center)[22]

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Weakness Pattern;

- ***Proximal & neck flexors weakness is common in all***
- ***Distal and asymmetric weakness in IBM***
  (wrist & finger flexors, quadriceps and ankle dorsiflexion)
- ***Dysphagia occurs in 30% of DM & PM & in 60% of IBM cases***
  (very common in scleroderma)
- ***Respiratory muscle involvement is rare***
  (seen in advanced PM and DM)
- ***Extraocular muscles are unaffected & facial weakness is very rare***
  (orbital myositis)
- ***In IBM shoulder abductors are stronger than forearm muscles, and hip flexors are stronger than quadriceps***
- ***Higher incidence of PN in IBM compared to DM , PM & NM***

Polymyositis Clinical Features;

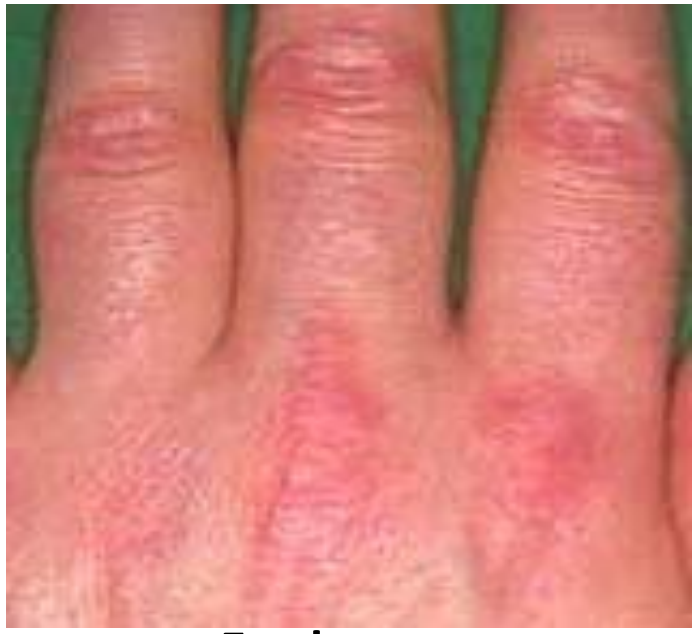
- Muscle weakness
  - Symmetric proximal > distal
  - Selective regions of weakness; dysphagia, neck flexors, hip flexors>quadriceps
- Onset age usually > 40 years
- Progression usually over weeks or months
- Usually progressive and disabling if not treated
- Muscle pain and tenderness are seen in 30% of PM & DM, more common with associated CTD & anti-Jo-1 autoantibody

Skin lesions in Dermatomyositis:

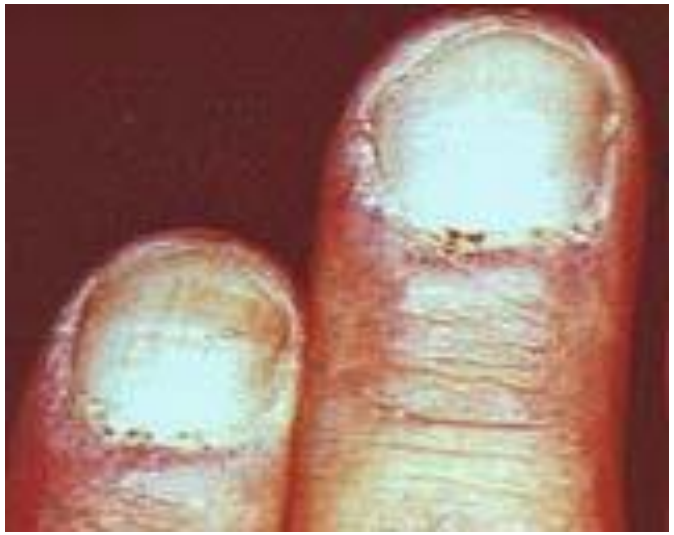
- **Often precedes the weakness onset, and may continue after the weakness improves**
- **Occurs in light-sensitive areas and is erythematous, edematous and sometimes pruritic**
- **Characteristically occurs over the knuckles, knees, elbows, anterior chest, back and shoulders (shawl sign), molar regions and bridge of the nose**

  - ***Heliotrope rash;*** purplish discoloration and periorbital edema at the upper eyelids
  - ***Gottron's sign;*** symmetric, scaling, erythematous, violaceous raised papules over the knuckles, and interphalangeal joints.
  - ***Mechanic's hands;*** raised scaly dry on extensor surfaces of finger joints
  - ***Nailfold lesions;*** dilated capillary loops at the base of the fingernails and irregular thickened cuticles

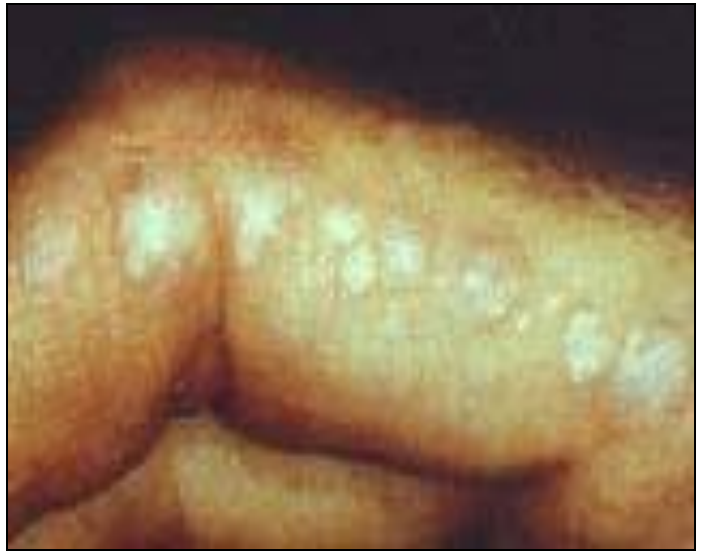
Heliotrope rash



Erythema



Nailfold lesions



Gottron's papules

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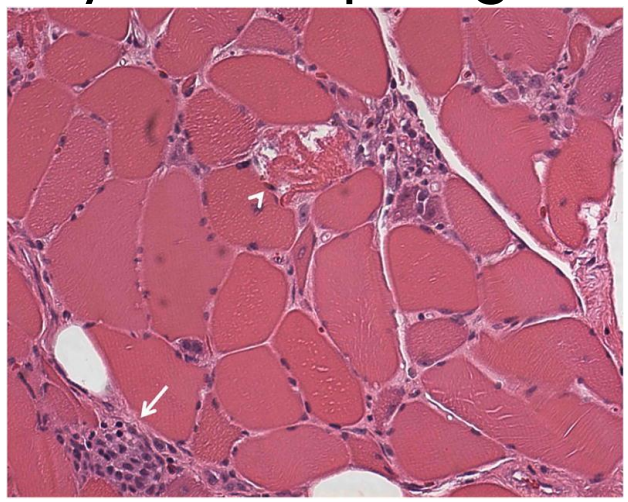
Necrotizing Myopathy (NM)

- An immune mediate myopathy with acute/subacute onset of proximal arms and legs weakness
- More rapid and severe course, and very high CK level than PM & DM
- Anti-HMGCR autoantibody is identified in up to 60% of patients, with a varying degree of exposure to statin (30 – 60% of these)
- 10-20% has anti-SRP antibodies; more severe and can be associated with cardiomyopathy, interstitial lung disease (ILD) & dysphagia.
- Muscle biopsy; necrotic fibers invaded by macrophages, some inflammatory T-cells
  no primary inflammatory process.



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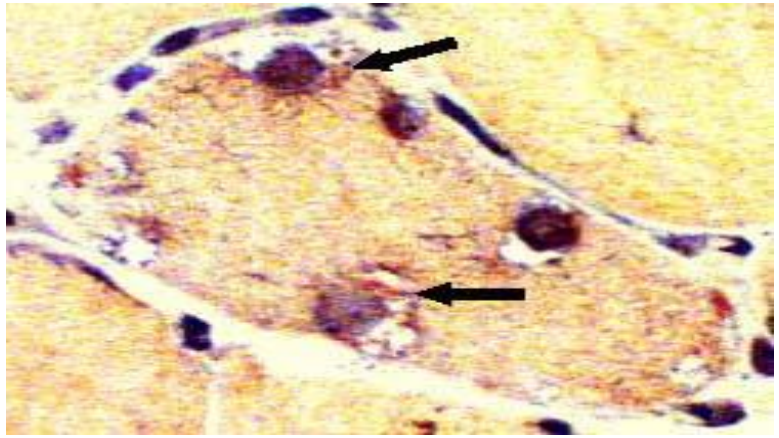
Inclusion Body Myositis (IBM)

- Older age of onset, male > female, frequent in North America
- Painless, slow progression of proximal and distal weakness (wrist & fingers flexors, quadriceps frequently severely involved). Most pts. remain ambulatory
- Rimmed vacuoles sarcoplasmic inclusions bodies, along with other features of inflammatory myopathy
- Poorly responsive to corticosteroids and other immunosuppressive therapy



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Associated Disorders:

- **Cardiac**
- **Pulmonary**
- **Esophageal paresis ;** Upper 1/3 with muscle weakness
- **Malignancy;** 3 - 4% in PM, and 25% in adult DM (TIF-1 and < NXP2 antibodies)
- **Connective tissue disorder (overlap myositis)**
- **Thyrotoxicosis** (rare)
  - CK is usually low, and may resolve with thyrotoxicosis treatment

(TIF-1; human transcriptional intermediary factor. NXP2; nuclear matrix protein 2)

Associated with PM, DM & NM;

- **Lungs**; In PM and DM due to pulmonary and none pulmonary causes, less in NM
  - Respiratory muscle weakness, CHF, chronic aspiration, lung infections, medication toxicity.
  - Associated ILD (5-10% radiological & ≥ 30% by pulmonary function tests). **Anti-Jo-1 autoantibody is present in > 50% in ILD associated cases**

- **Cardiac;** May be associated with poor prognosis
  - Myocarditis, CHF, pericarditis and valvular disease are uncommon
  - ECG abnormalities in up to 40% of patients.
  - Frequent association with anti-SRP autoantibody in severe PM and NM
  - Abnormal vasoconstriction, angina associated & Raynaud's

- **Inflammatory signs** in severe PM and DM with collagen vascular disease (fever, high C-reactive protein, high ESR)

- **Calcinosis** in muscle & subcutaneous tissue in late childhood DM

Association With Malignancy

Risk of malignant disease

- More often develop within ≤3 years of diagnosis, and less thereafter
- Mainly in adults, higher with increasing age in all forms, except IBM
- Higher incidence in White, Northern Europeans or Australians.
- Stronger association in adult DM (25%) than in PM 3.4% (75% of ADM with anti-TIF-1 & 37% of ADM with NXP2 antibody and in NM cases with anti-HMGCR antibody)
- In DM specific associations in order of decrease frequency; ovarian, lung, pancreatic, non-Hodgkin lymphoma, stomach, colorectal
- In PM specific associations in order of decreased frequency ; non-Hodgkin lymphoma, lung, bladder and hematological

(TIF-1; human transcriptional intermediary factor. NXP2; nuclear matrix protein 2)

Associated Connective Tissue Disorders

- Seen in patients with PM & DM (overlap syndromes)
  (scleroderma 5 -17%, SLE <10%, RA 13%, Sjögren's and MCTD)
- Usually present with arthralgia and arthritis with less severe muscle disease
- IBM rarely can be associated with Sjögren's syn, or RA
- Usually more responsive to treatment
- Associated malignancy is rare

MYOSITIS SPECIFIC ANTIBODIES (MSAs)

- **Found in patients with PM, DM and NM,**
  - high sensitivity and specificity
  - heterogeneous, each occurring in only a small proportion of patients, and defines a specific subgroup of patients.
  - directed against cytoplasmic ribonucleoprotein involved in the process of protein synthesis, and nuclear antigen

  - **Antibody class IgG;**
    - **antisynthetases (anti-tRNA)**
    - **anti-signal recognition particle (anti-SRP)**
    - **anti-nuclear helicase (anti-Mi-2)**
    - **anti-HMGCR antibody (3-hydroxy-3-methylglutaryl-coenzyme A reductase)**
    - **anti-TIF1-y and anti-NXP2 antibody**

MYOSITIS SPECIFIC ANTIBODIES (MSAs)

- **Found in NM**
  - anti-HMGCR antibody (3-hydroxy-3-methylglutaryl-coenzyme A reductase)

- **Found in IBM**
  - cN1A autoantibody directed against the 5'-citosolic nucleotidase1A was identified in the sera of sIBM
  - widely variable sensitivity 37% -50 and specificity (87%-100%).

- **Found in myositis associated with CTD**
  - none specific overlap antibodies
  - in myositis and scleroderma, RA, SLE, MCTD, Sjögren's Syndrome

Association with Autoantibodies

Myositis-specific autoantibodies (MSAs)

Autoantibodies:	Clinical Features:
• ***Antisynthetase (anti-tRNA) mostly anti-Jo-1***	20 - 25% of PM and DM, acute onset ILD, nonerosive arthritis, Raynaud's, frequent relapse during prednisone
• ***Anti-signal recognition particle (anti-SRP)***	5% of PM, acute onset, myalgias, frequent myocarditis, resistance to prednisone, poor prognosis (25% 5-year survival). 15% in NM with more severe weakness, ILD and cardiomyopathy
• ***Anti-nuclear helicase (Anti-Mi-2)***	5 -10% of DM, acute onset with florid rash, good response to immunosuppressive therapy.
• ***Anti-HMGCR antibody***	in ≥ 50% of NM, varying degree of exposure to statin and incidence of malignancy
• ***Anti-cNA1***	in ≥ 37% of sIBM, more severe dysphagia

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ASS

OM

DM

NM

IBM

Auto-antibody	Frequency	Typical clinical features
Anti-tRNA: Jo-1, PL-7, PL-12, HA (YRS/Tyr), OJ, KS, ZO, EJ	anti-tRNA: 30% in myositis Jo-1: 15-20% in myositis PL-7 and PL-12: each 3-4% All others <2%.	Higher rate of ILD and mortality in PL-7/PL-12 than Jo-1
Anti-SS-A/Ro52/Ro60 SS-B/La	SS-A: up to 19% in myositis, 25% in OM, SS-B: 7% in myositis, 12% in OM Ro52 often together with anti-synthetase, e.g. 56-72% of Jo-1.	Association with Sjögren's syndr., SLE and systemic sclerosis. Ro52 more common in myositis than Ro60; both occur in CTD. Ro52 and Jo-1-double positive: high rate of malignancies, poorer prognosis.
U-snRNP	up to 10% of myositis	Associated with CTD, SLE and systemic sclerosis. Often good prognosis.
PM/Scl	~8-10% of myositis	Associated with systemic sclerosis. Often severe disease course and insufficient treatment response.
Ku	up to 20-30% in OM	Associated with systemic sclerosis, SLE and CTD. High rate of ILD, which does not respond well to glucocorticosteroids.
Mi-2	5-10% in DM	Classical DM
MDA5	15-30% in DM	Often amyopathic DM, often ILD.
TIF-1α/β/γ	~20% in DM	Malignancy common (75%). Most common in JDM–without tumor.
NXP-2	10-15% in DM	Malignancy frequent (37.5%). Second most common antibody in JDM–without malignancy, but often calcinosis.
SAE	2-8% in DM	Often amyopathic and with ILD.
SRP	5% in myositis	Often severe with muscle atrophy, ILD and dysphagia. Often basic immunosuppressive treatment regimen not sufficient.
HMGCR	5-8% in myositis	High frequency of malignancy.
cN1A	~30% in IBM	Sjögren or SLE positive by 20-30%, even without muscle symptoms. In IBM: more severe disease course, dysphagia and higher mortality.

Schmidt J. J Neuromuscular Dis. 5(2); 2018, 109-129

Laboratory and Diagnostic Features

- Routine hematologic, chemistry and radiology are generally normal unless there is an underlying neoplasm or CTD (elevated ESR)
- Elevated serum CK in ≥ 90% of cases, normal in ≤ 6%
  (5 – 50 folds of normal in PM, DM & NM, and less in IBM 3–10 folds)
- CK is most reliable and correlates with clinical course
- Other enzymes (LDH, transaminases, aldolase) in active myositis
- MRI parameters to assess pattern of affected muscles, current inflammation and fibrosis
- Ultrasound can help to identify affected muscles suitable for biopsy.

Electrodiagnostic Features

Motor and sensory nerve conduction studies are typically normal

NEMG Examination;

Signs of membrane irritability (fibrillations; positive waves, CRD), *most frequent in paraspinal muscles.*
Myopathic MUAP's
Early recruitment of MUAP's

Frequency of typical findings

Most common in acute DM, PM & NM
In the chronic stages; a mix of high amplitude, long duration, polyphasic MUAP's; sparse or lack of spontaneous activity

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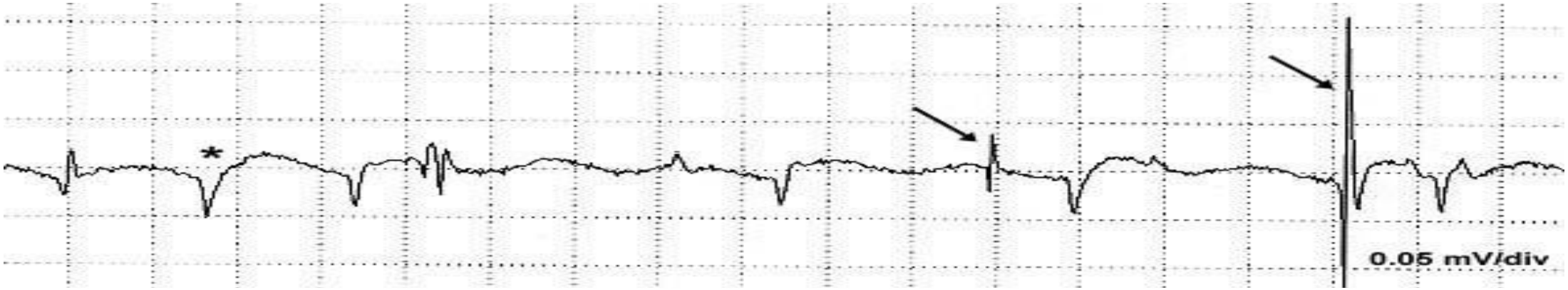
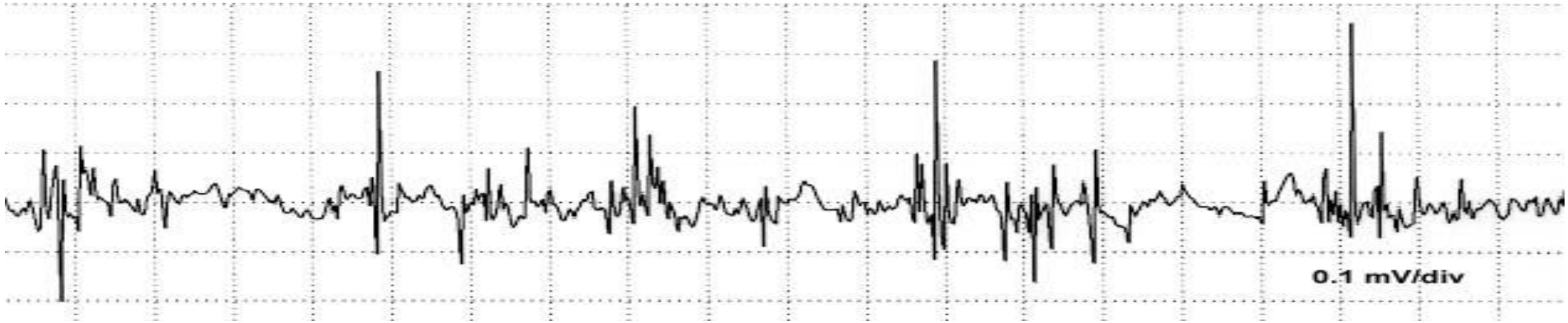
- 

Spontaneous activity (Fibrillations and positive sharp waves)



Short duration, small amplitude, polyphasic MUAP's

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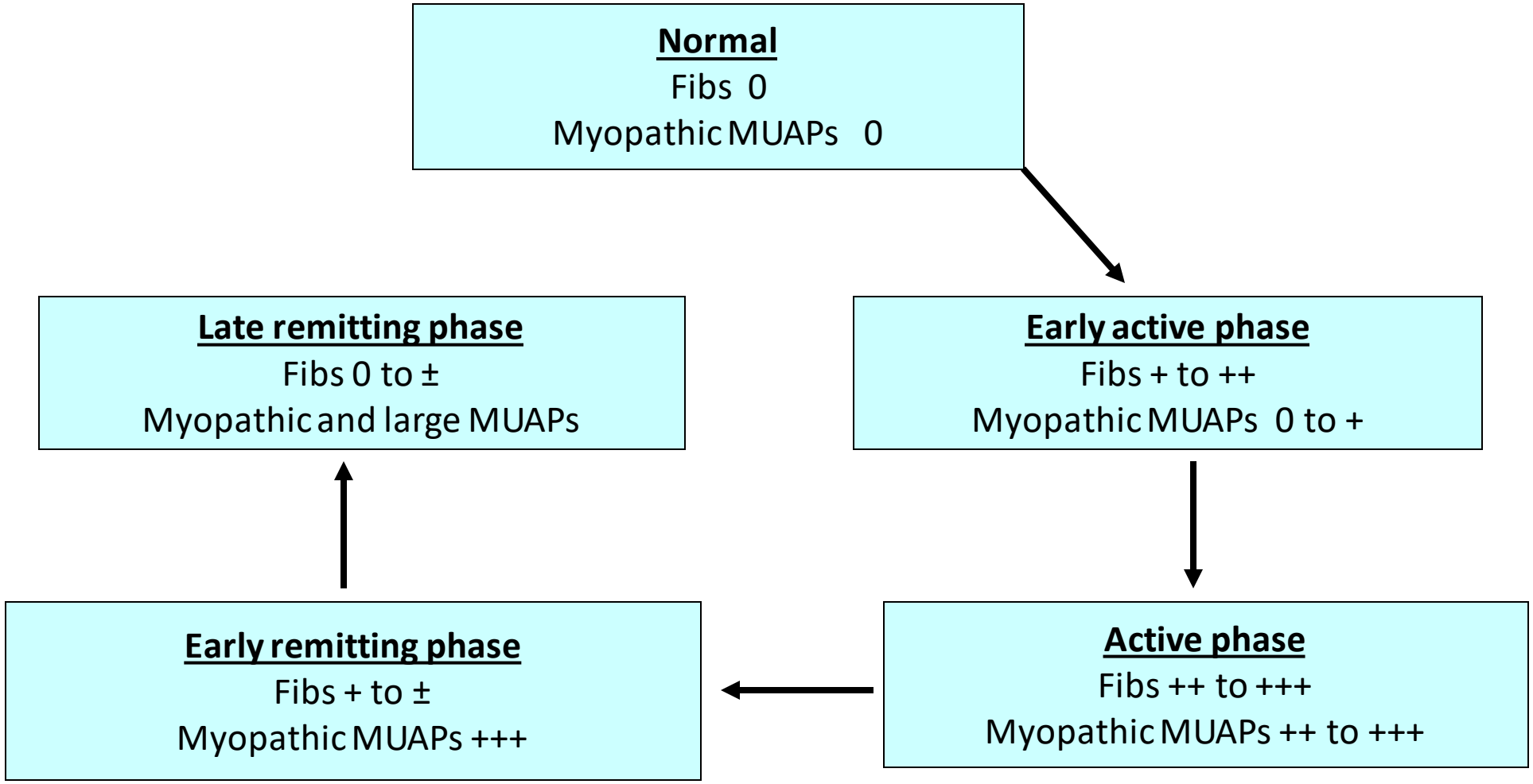

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PATHOGENESIS:

DM;

- Humeral mediated process involving B cells and helper T cells infiltrate
- Deposition of MAC and complement in intramuscular microvasculature, leading to necrosis and thrombosis of capillaries, arterioles and venules in the periphery of the fascicle

PM;

- Cell-mediated immune response directed against muscle fibers, CD8 cytotoxic and suppressor T cells, and macrophages infiltrate
- Associated pro-inflammatory cytokines and chemokines destroy healthy muscle fibers

NM;

- Autoantibody-induced complement dependent muscle fiber necrosis

IBM;

- Immune and degenerative mechanisms (invasion of non-necrotic fibers by T cells, and Congo-red-amyloid positive inclusions)
- Filamentous inclusions resembling myxovirus nucleocapsids *(virus etiology never proven)*

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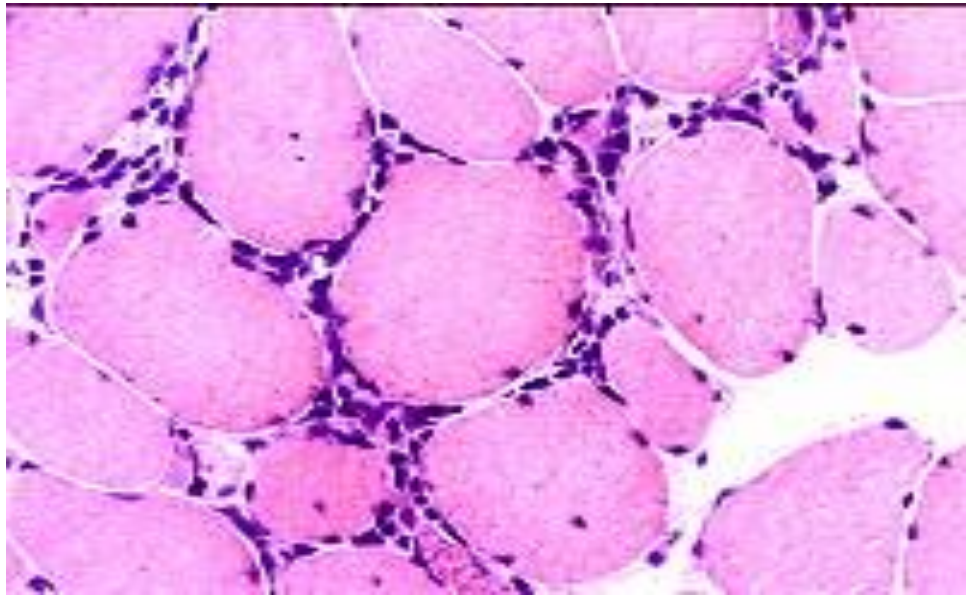
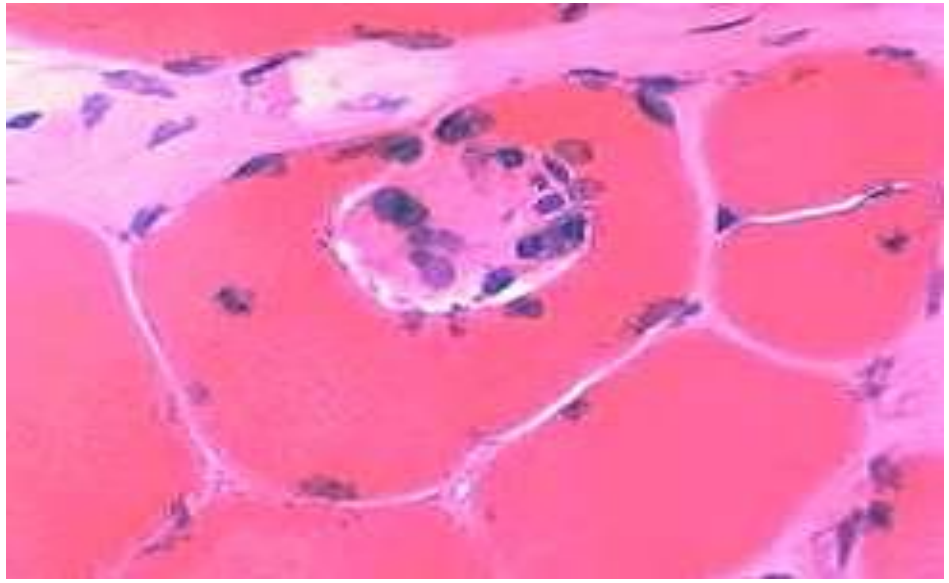
Pathology (Muscle Biopsy)

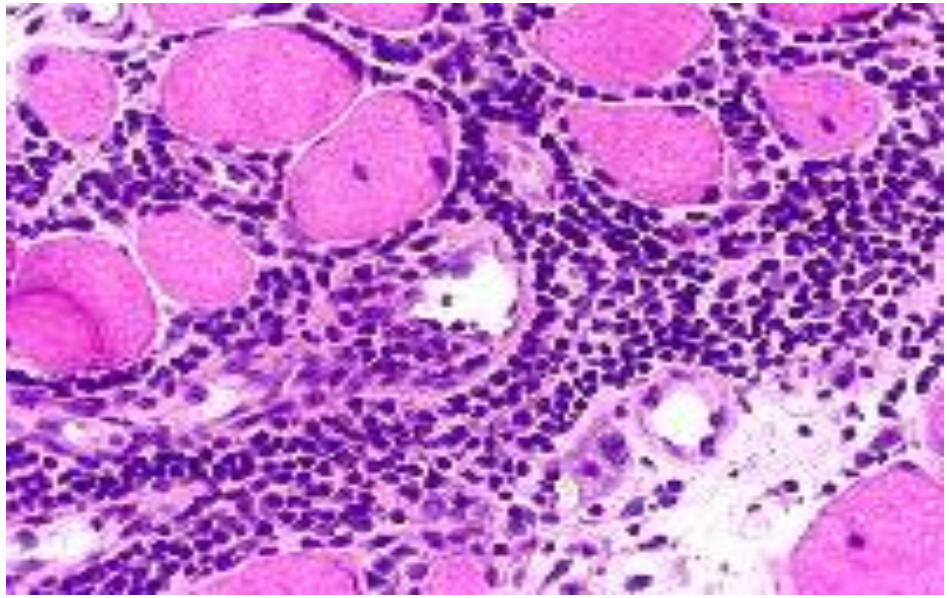
- Is the diagnostic tool of great importance in suspected IMs.
- Excludes other causes & reveals unique distinctive features
- Characteristic features include; mononuclear cell inflammation, muscle fiber necrosis, and regeneration.
- Necrosis, regeneration and inflammation may also be found in muscular dystrophies, and chronic neurogenic atrophies.
- Muscle biopsy is normal or only mildly abnormal in ≤12.5% in otherwise typical PM or DM (Bohan et al.)

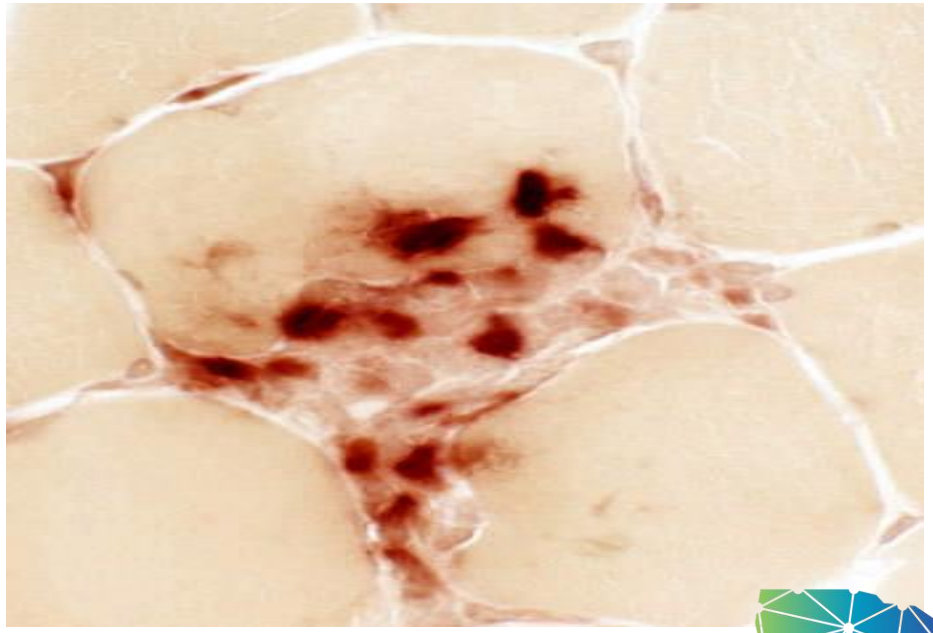
Muscle biopsy findings in PM, DM, NM and IBM

Histopathology	Distinguishing features
• Mononuclear cell inflammation	• In PM & IBM is endomysial; in DM is perivascular and interfascicular
• Scattered necrotic muscle fibers invaded by macrophages	• In NM
• Fibers degeneration and regeneration and vacuoles	• In PM & IBM is scattered. In DM is perifascicular
• Fiber size variation	• IBM; small fibers cluster and rimmed vacuoles
• Oxidative enzymes staining alterations	• In DM perifascicular small fibers
• Capillary damage	• Characteristic of DM, no alterations in PM or IBM
• Muscle inclusions	• In IBM intracytoplasmic and intranuclear filamentous inclusions, amyloid deposit, mitochondrial abnormality in some IBM cases









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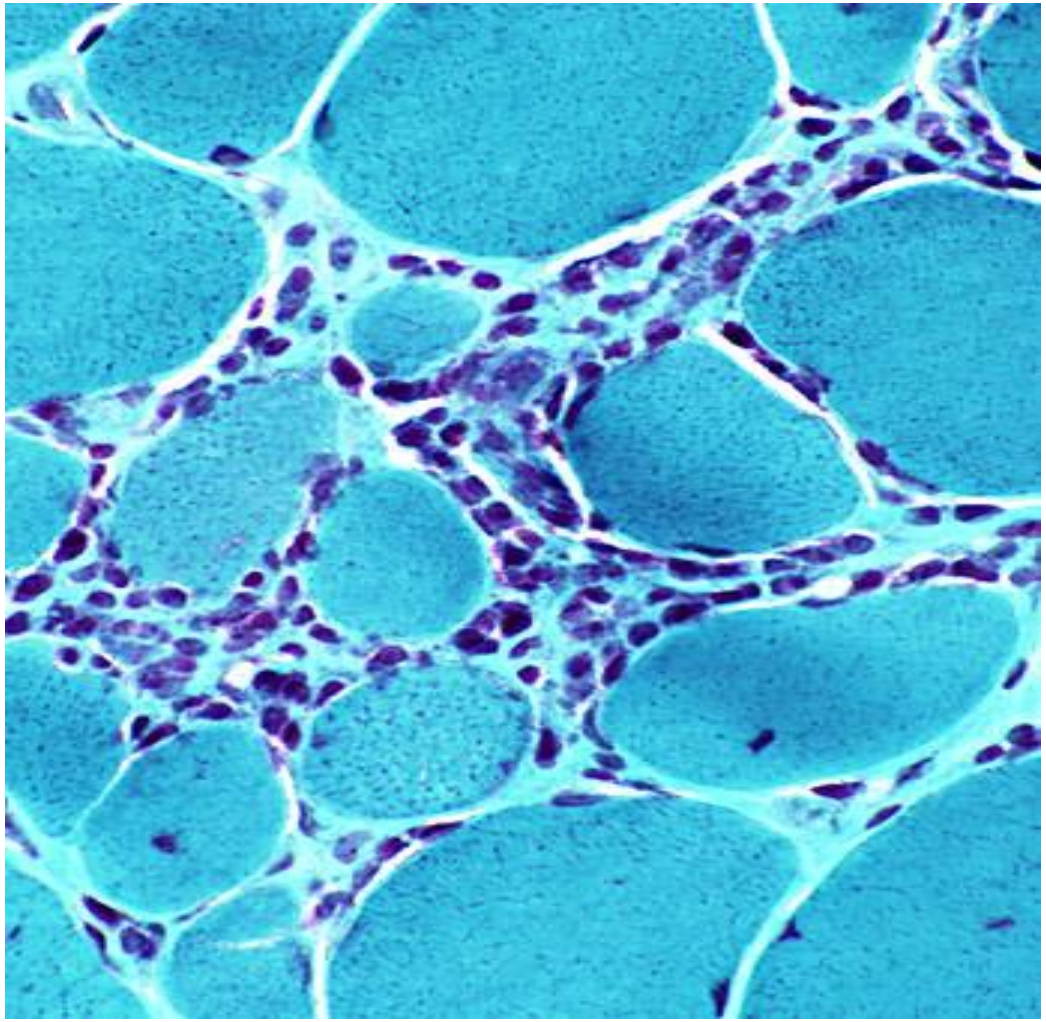
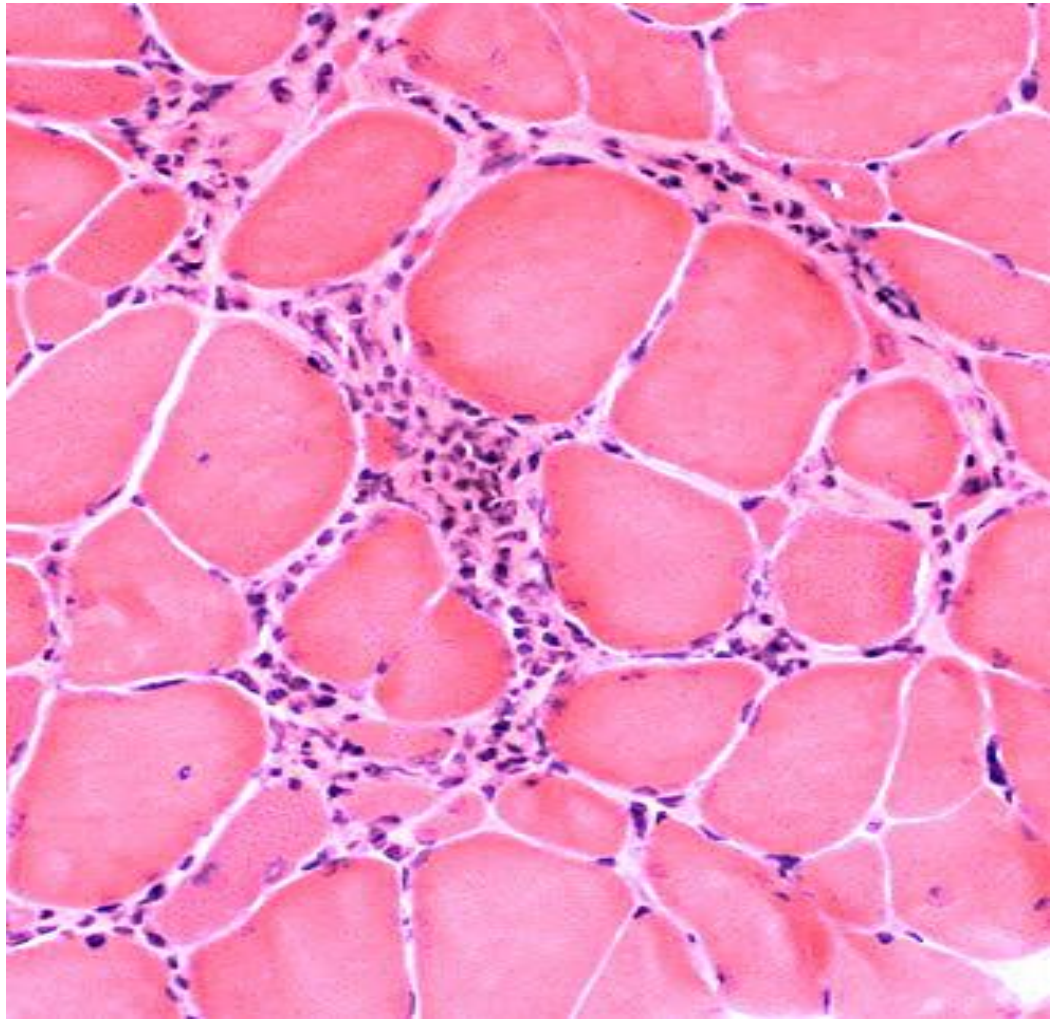
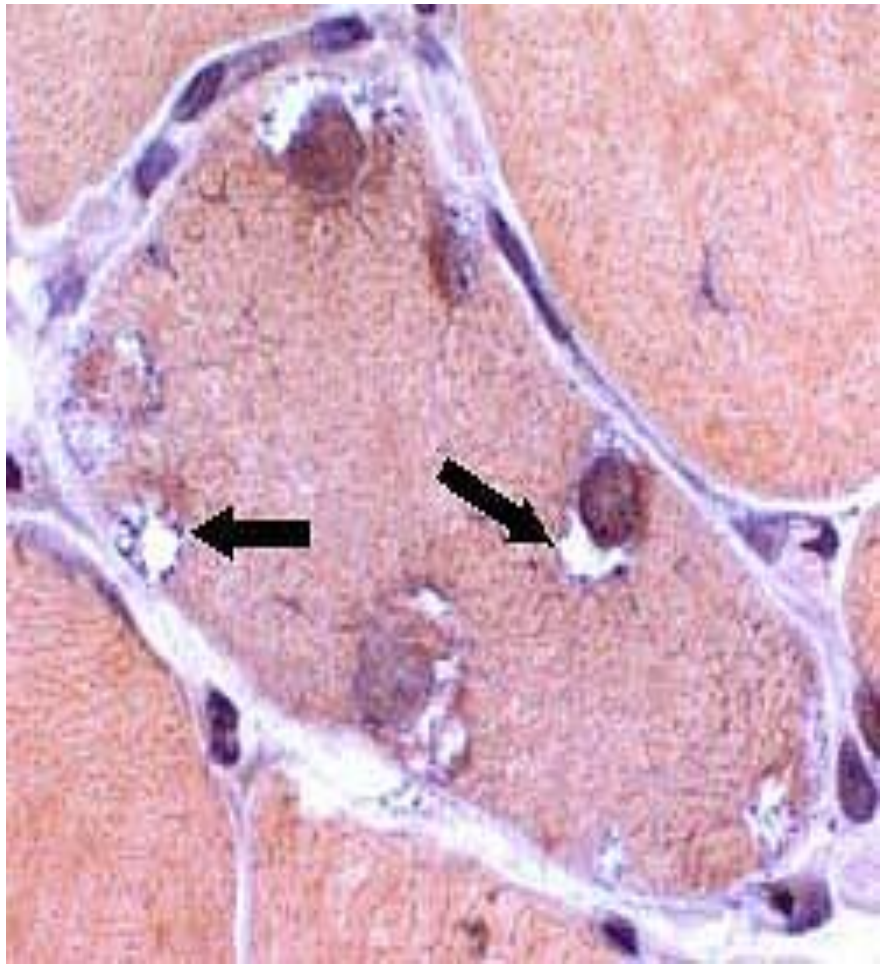
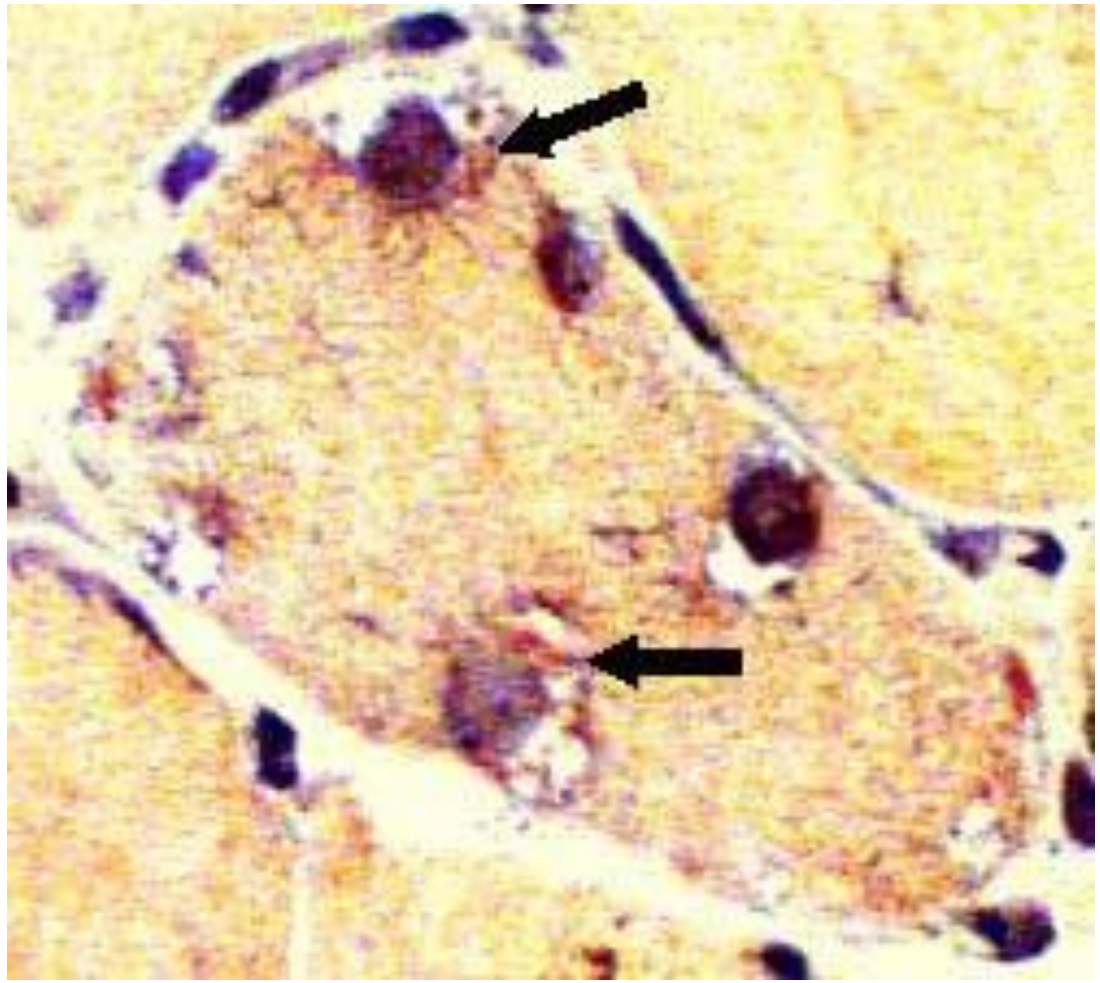
INFLAMMATION: Predominantly Endomysial









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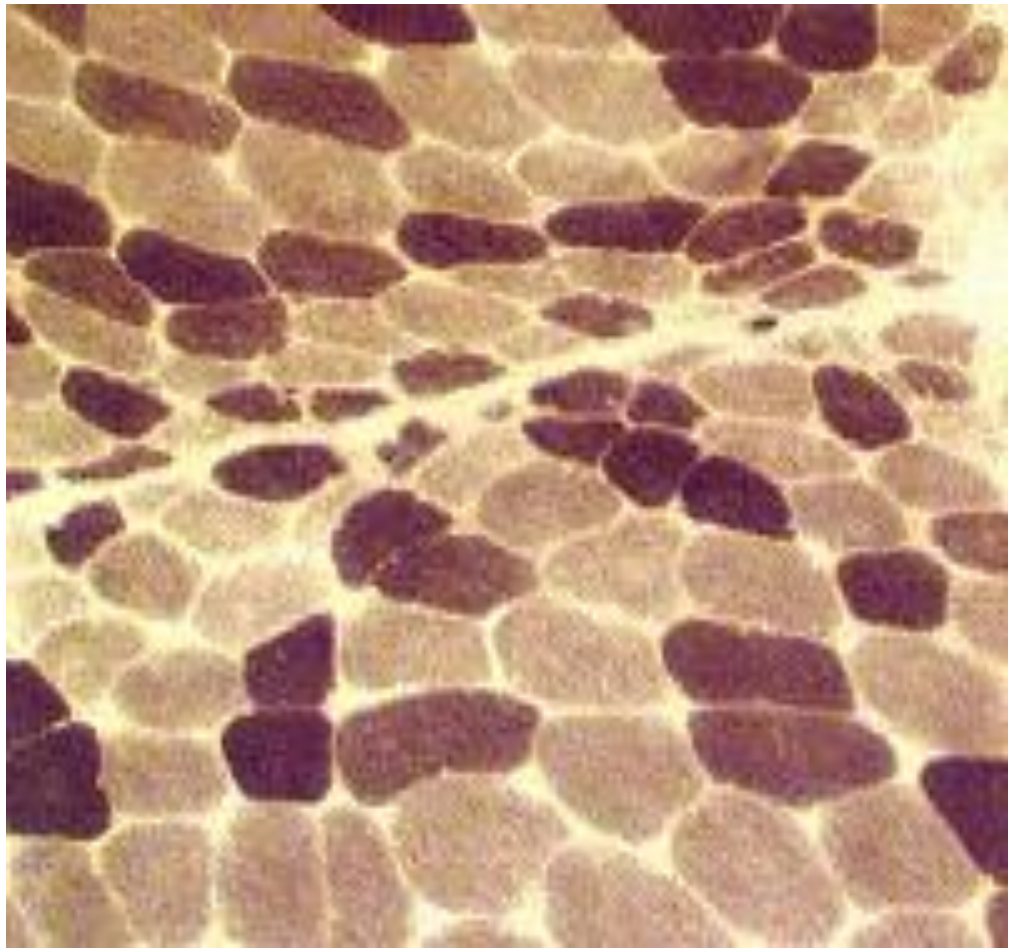

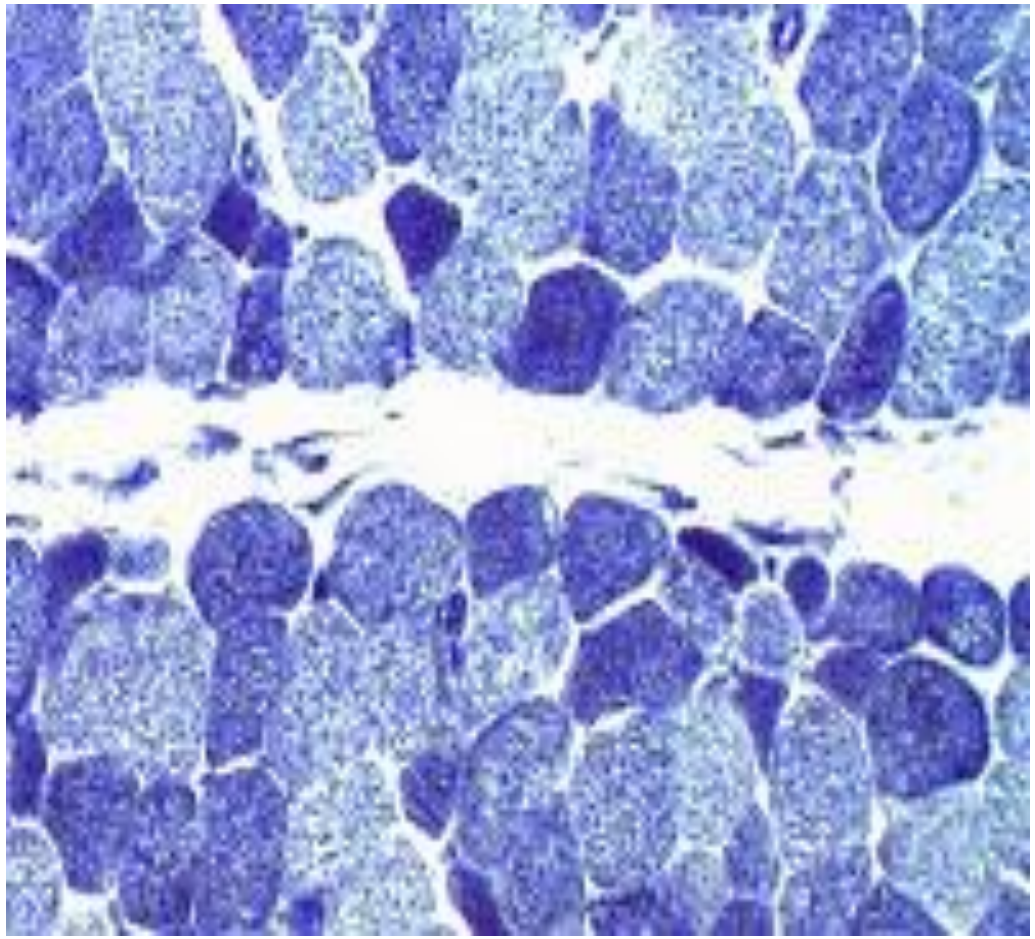

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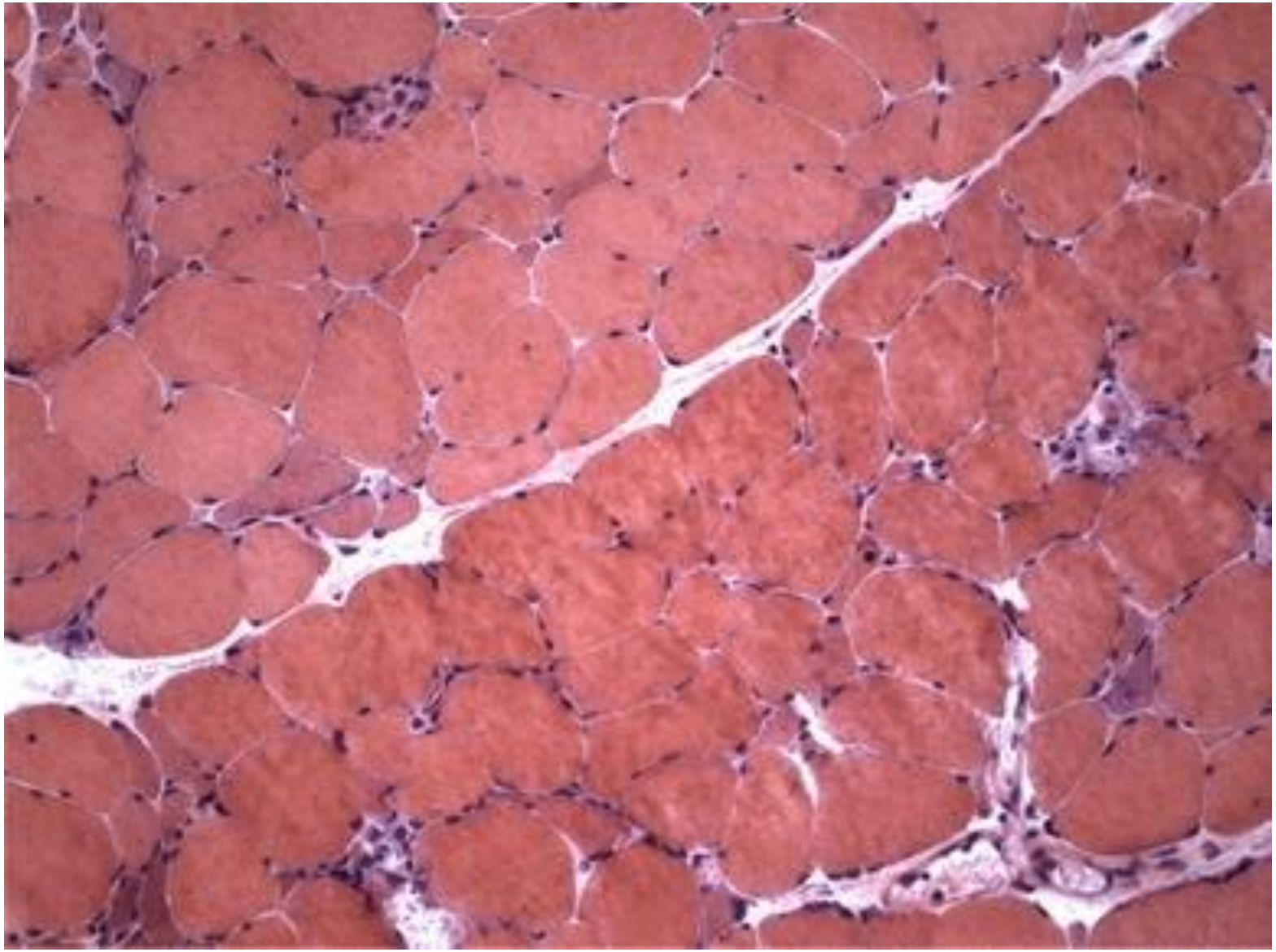

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TREATMENT

- The natural history of untreated PM and IBM has not been formally studied
- Improvement and ultimate favorable outcome depend on early diagnosis and prompt initiation of immunotherapy
- IBM is therapy resistant
- Spontaneous recovery or improvement seen only in very small number of patients
- Patients with subtle weakness may be closely followed without specific treatment
- Associated disorders of the lungs, heart, CTD, or neoplasms are potentially treatable.

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Treatment options

- **Prednisone**
  Oral 1 mg/kg per day, then taper to every other day
  Alternative; Solumedrol 500 - 1000 mg/day for 3 – 5 days
  or Dexamethasone 40 mg/day for 4 days monthly (both have fewer side effects)
  Latency before benefit is 4 to 8 weeks
  Tapering slowly based on clinical improvement
  Maintenance dose of 5 – 15 mg is often necessary

- **Steroids sparing agents;**
  - **Azathioprine**
    50 mg/day, then increase weekly up to 200 mg/day,
    (lower dose in low TPMT enzyme activity)
    Latency before benefit: 4 to 6 months
  - **Methotrexeat**
    7.5 to 22.5 mg/week, with folic acid 5 -10 mg next day
    Latency before benefit: 2 to 4 months
  - **Mycophenolate**
    500 mg BID up to 2000 3000 mg/day in divided dose
    Latency before benefit up to 2- 4 months

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- **Alternative or add-on treatment ;**
  - **IVIG**
    1 -2 gram/kg/BW over 2 – 3days course every 3- 6 weeks
    Latency before benefit 2 to 3 courses
    Individual dose established over several cycles, clinical effect and tolerance
- **Treatment escalation;**
  - **Cyclosporine**
    Starting dose: 2.5 mg/kg bid
    Latency before benefit: 2 to 4 months
  - **Rituximab or Cyclophosaphamide**
- **IBM**

  No currently effective treatment based on several clinical trials
  Steroids, immunosuppressive drugs, IVIg all failed to show benefit.
  Ongoing clinical trials (arimoclomol, pioglitazone, rapamycin, rAAV1 virus follistatin gene transfer.
  Symptomatic treatment and swallowing management.

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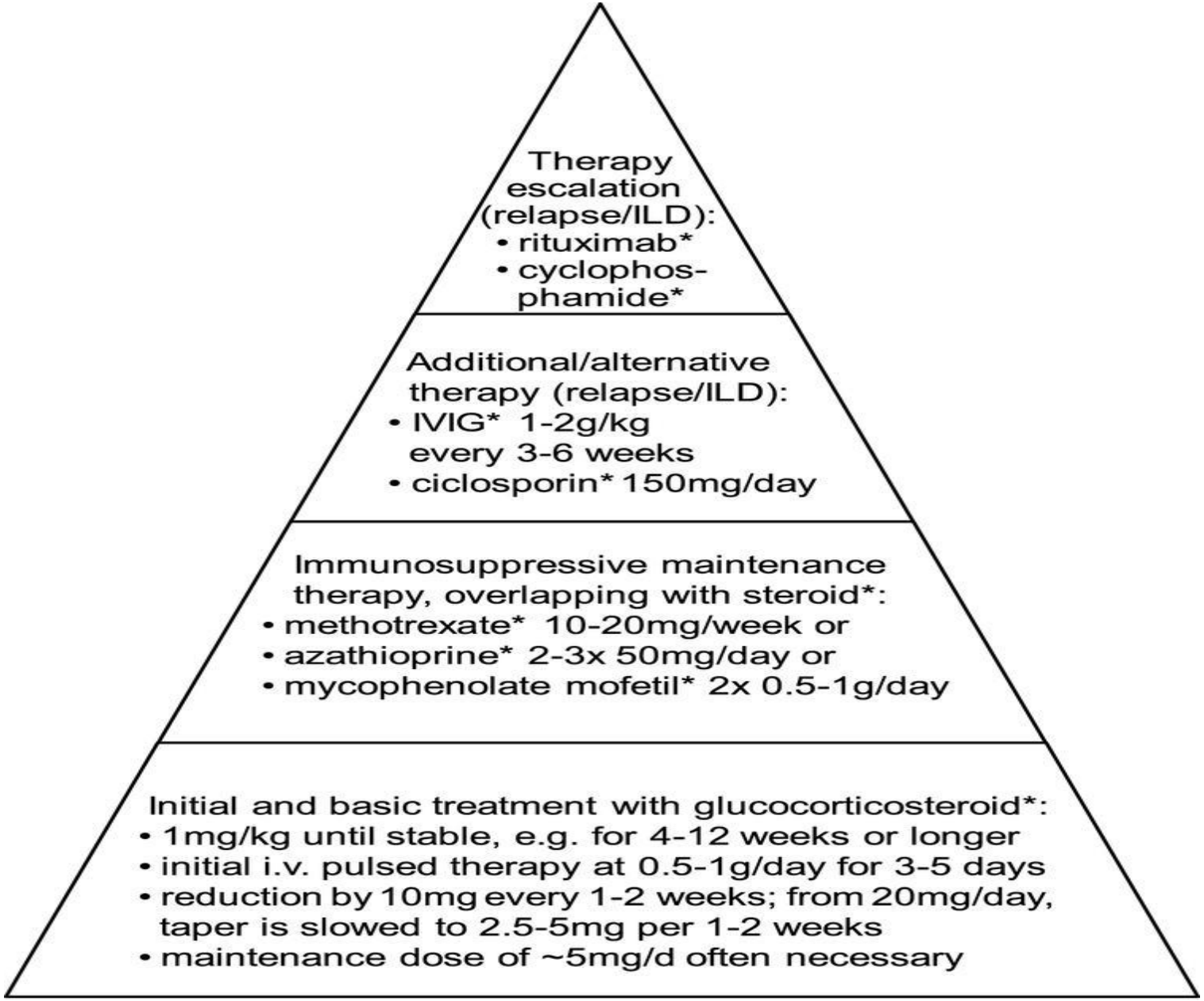

*Individual dosing to be adapted for each case; further information in the text.

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Corticosteroids Treatment Issues;

- Optimal dose …
- When to begin dose reduction …
- How long to continue therapy …
- The role of alternate-day dose …
- Ways of assessing response …
- Steroid resistant patients …
- Second-line and third-line therapy …

➢ **1mg/kg per day 4 X 6 weeks.**
Then taper to 1mg/kg every other day over 10 weeks.
Then by 5mg every 3 weeks, until 40mg every other day.
Then by 2.5 mg every 2 weeks to the lowest maintenance d

➢ Features predicting response;

- Proximal pattern of weakness, with onset less than a year
- Association with myalgias, DM skin rash, CTD
- Serum CK very high
- Muscle biopsy features characteristic of DM, active PM & NM.

➢ Features predicting resistance;

- Chronic > 18 months focal or asymmetric pattern of weakness
- Normal or low serum CK
- Anti-signal recognition particle or anti-Jo-1 MSAs positive
- Muscle biopsy features characteristic of IBM, or burned-out PM

Supportive and Symptomatic Therapy

- Management of impaired swallowing secondary to functional stenosis of upper esophagus sphincter options include;
    - cricopharyngeal myotomy,
    - pharyngeo-balloon dilation
    - repeated botulinum injections (favored option)
- Regular screening for aspiration using fiberoptic swallowing test (FEES)
- Screening for malignancy initial in ADM and subtypes of auto-antibody
- Involved skin protection from sunlight in DM with topical treatment.
- Interdisciplinary management of ILD and cardiac involvement.
- Patients should receive physiotherapy from beginning of the disease

Prognosis and Outcome;

- Weakness severity, CK elevation and biopsy abnormality at onset do not correlate with outcome
- Prognosis is worse with cardiac involvement, ILD
- PM or DM and associated malignancy have higher mortality
- Patients with anti-Jo-1 and anti-SRP antibodies have a poorer prognosis, and less response to corticosteroids
- Diagnosis delay for ≥18 months have less favorable response
- PM or DM without unfavorable factors show 70-80% good response
- Most patients require to remain on low drug maintenance dose
- Relapses were noted in 60% of pts. over 13 years, most frequent during s drug maintenance stage
- **IBM responds poorly, if at all to treatment**

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THANK YOU

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