

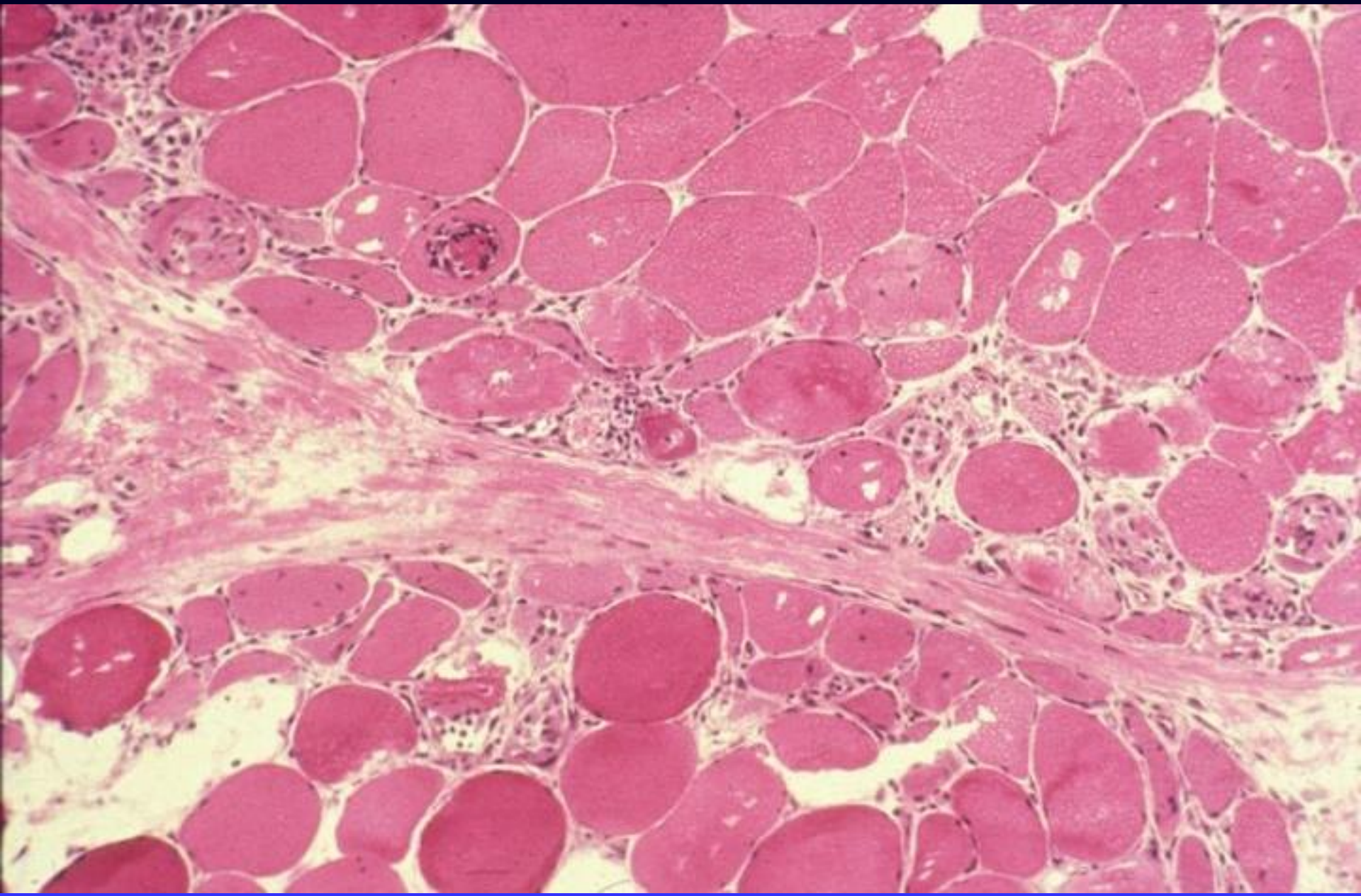
Myositis 101

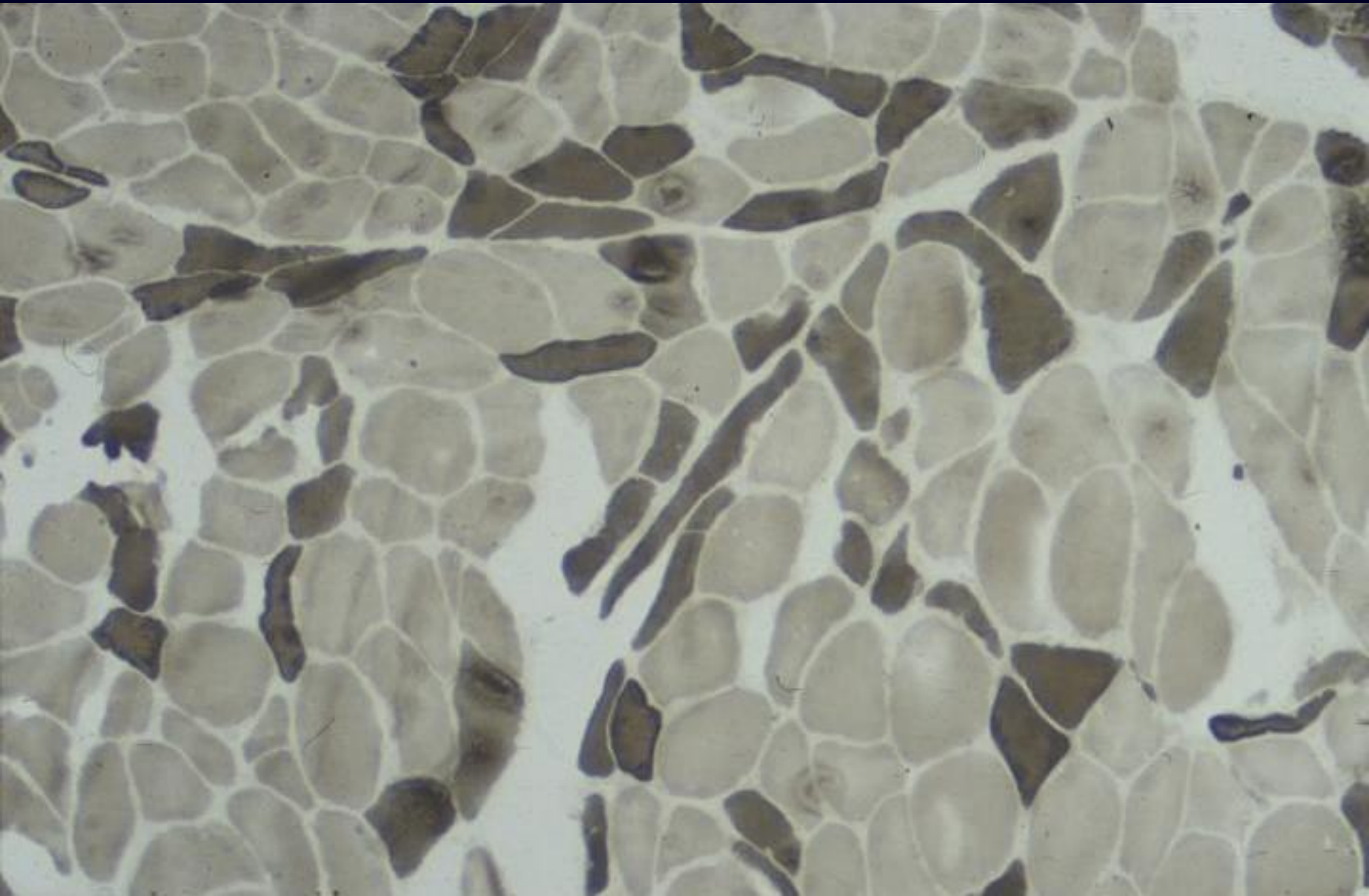
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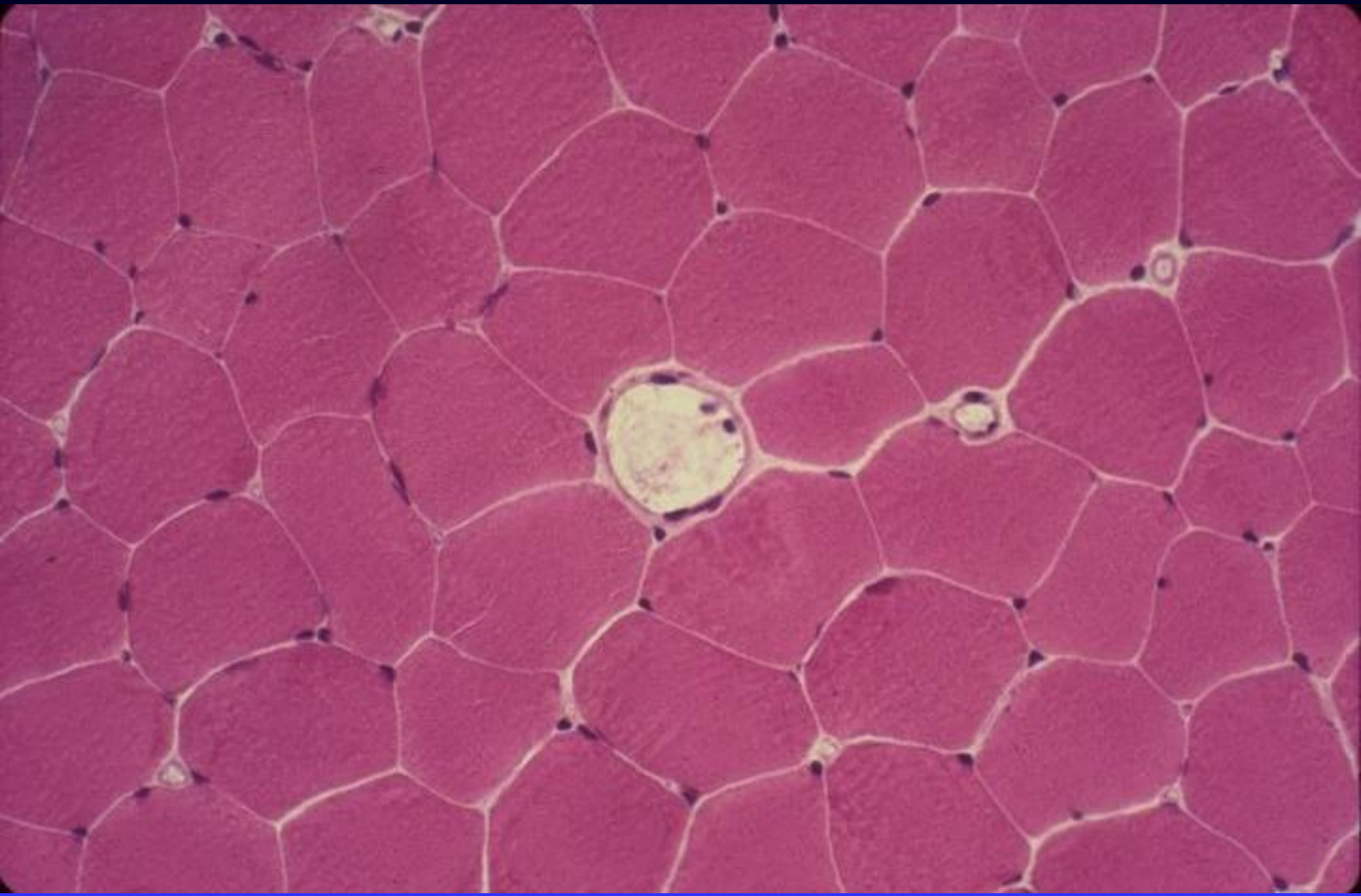
Criteria for Defining Polymyositis

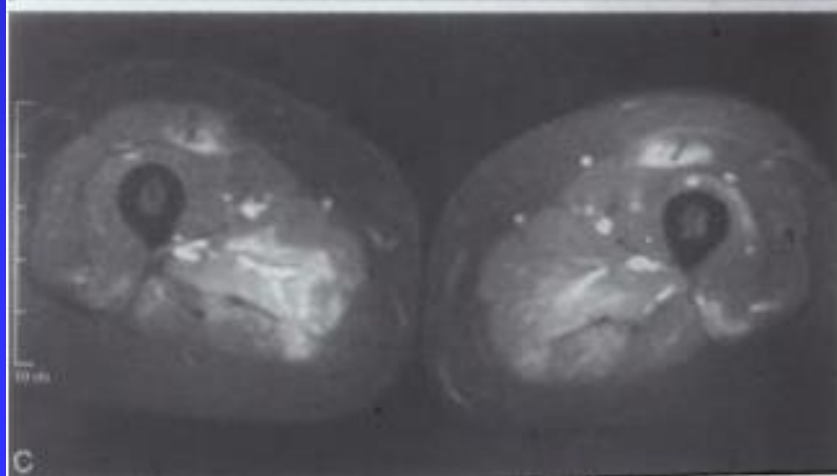
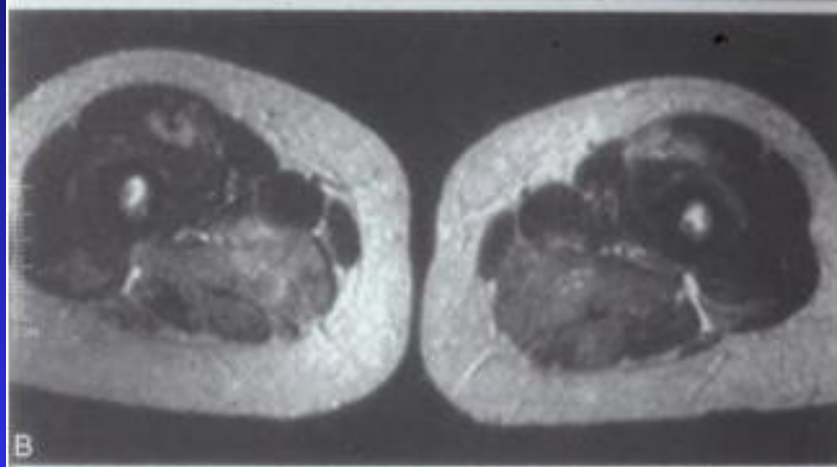
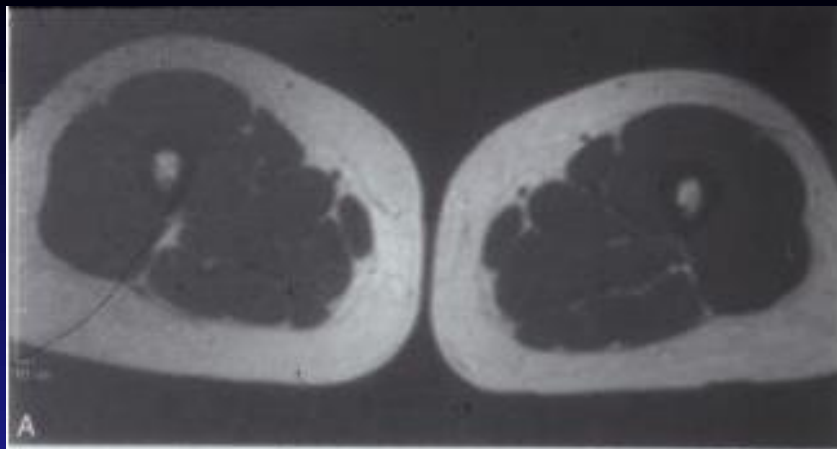
1. Symmetrical weakness of limb-girdle muscles and anterior neck flexors.
2. Muscle biopsy evidence of necrosis of Type I and II fibers, phagocytosis, regeneration, variation in fiber type with inflammatory exudation.
3. Elevation in serum or skeletal-muscle enzymes.
4. Electromyographic triad of short, small, polyphasia motor units, fibrillations and sharp waves; and bizarre, repetitive discharges.
5. Dermatologic features.

*Nonsuppurative Inflammation of
Muscle*

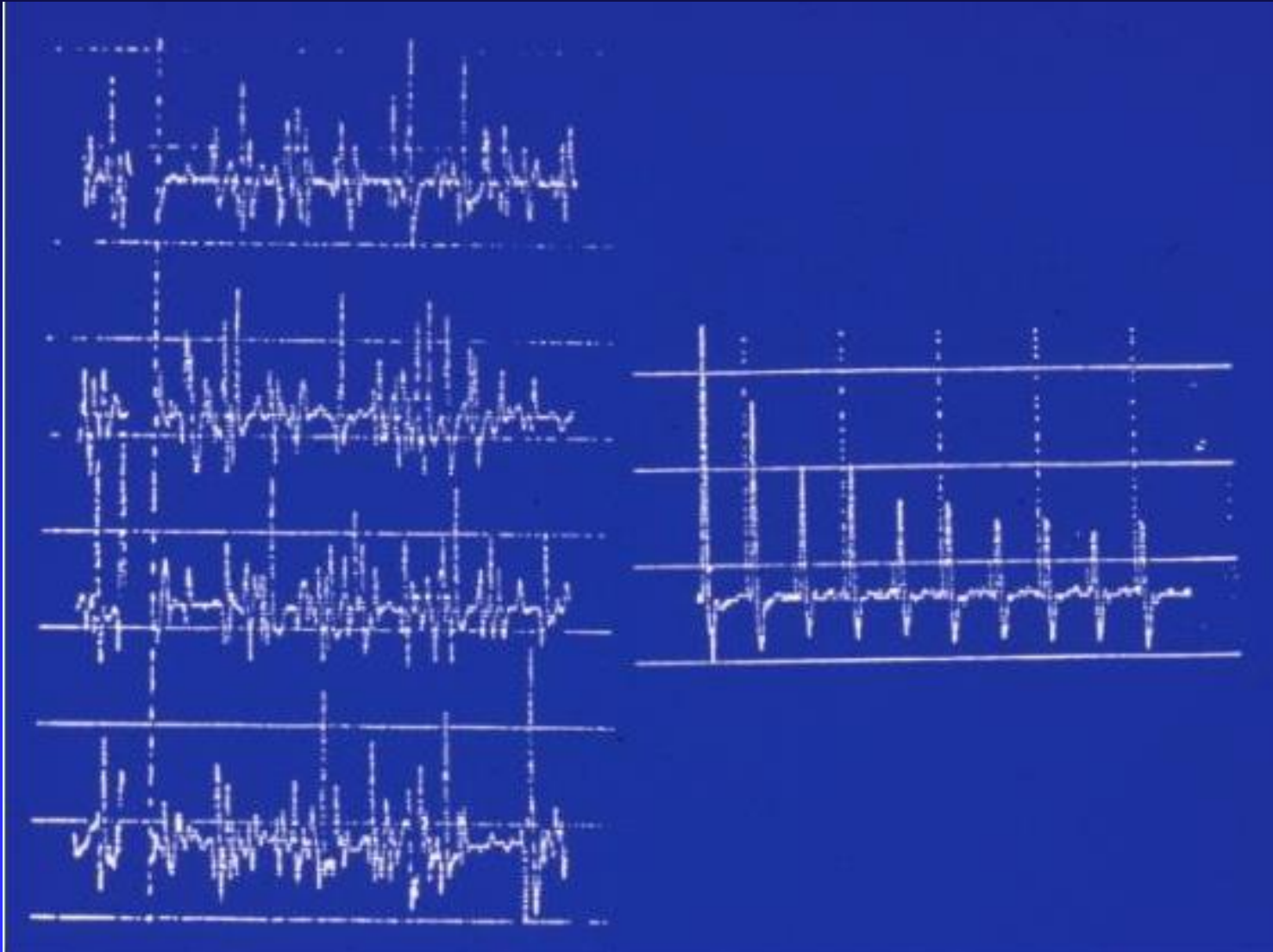








Electrodiagnostic Testing



Electrophysiologic Changes in Inflammatory Myopathy

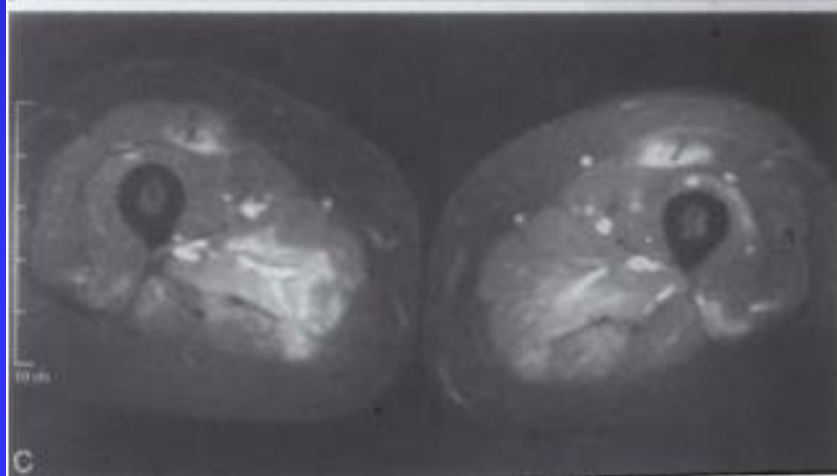
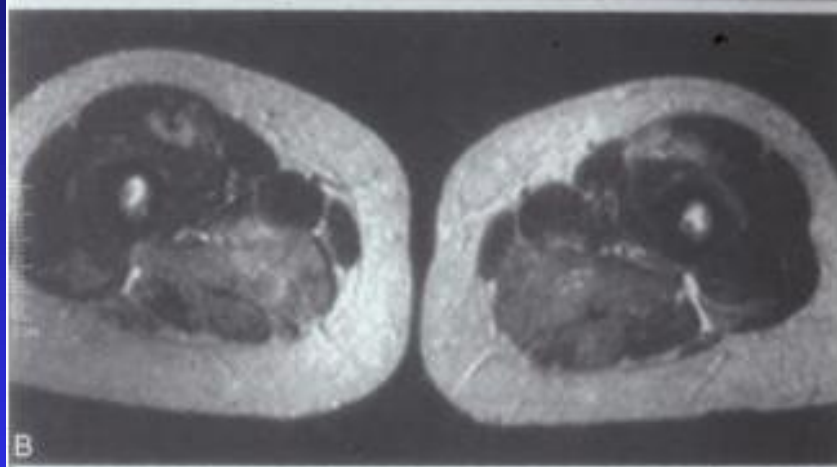
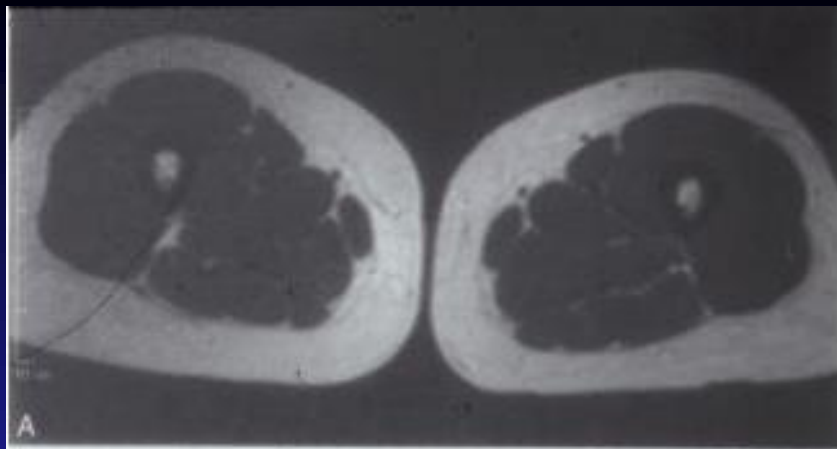
- Fibrillation at rest
- Increased insertional activity
- Bizarre high-frequency repetitive discharges
- Polyphasic potentials of short duration and low amplitude
- Spontaneous and positive sharp waves

EMG and Inflammatory Myopathies

- About 40% will have the classic triad
- EMGs are entirely normal in 10%
- Abnormalities may be limited to paraspinous muscles
- Neuropathic findings may also be seen in
 - ◆ Inclusion body myositis
 - ◆ Myositis with anti-SRP antibodies
 - ◆ Myositis and malignancy

EMG and Nerve Conduction

- Differentiate myopathic and neuropathic disorders and further localize the lesion.
- Identify appropriate site for biopsy.



Muscle Enzymes in IIM

- CPK, aldolase, AST, ALT, and LDH
- None of these enzymes may correlate well with disease activity
- Patients with an IIM may become completely asymptomatic, but continue to have elevated enzymes

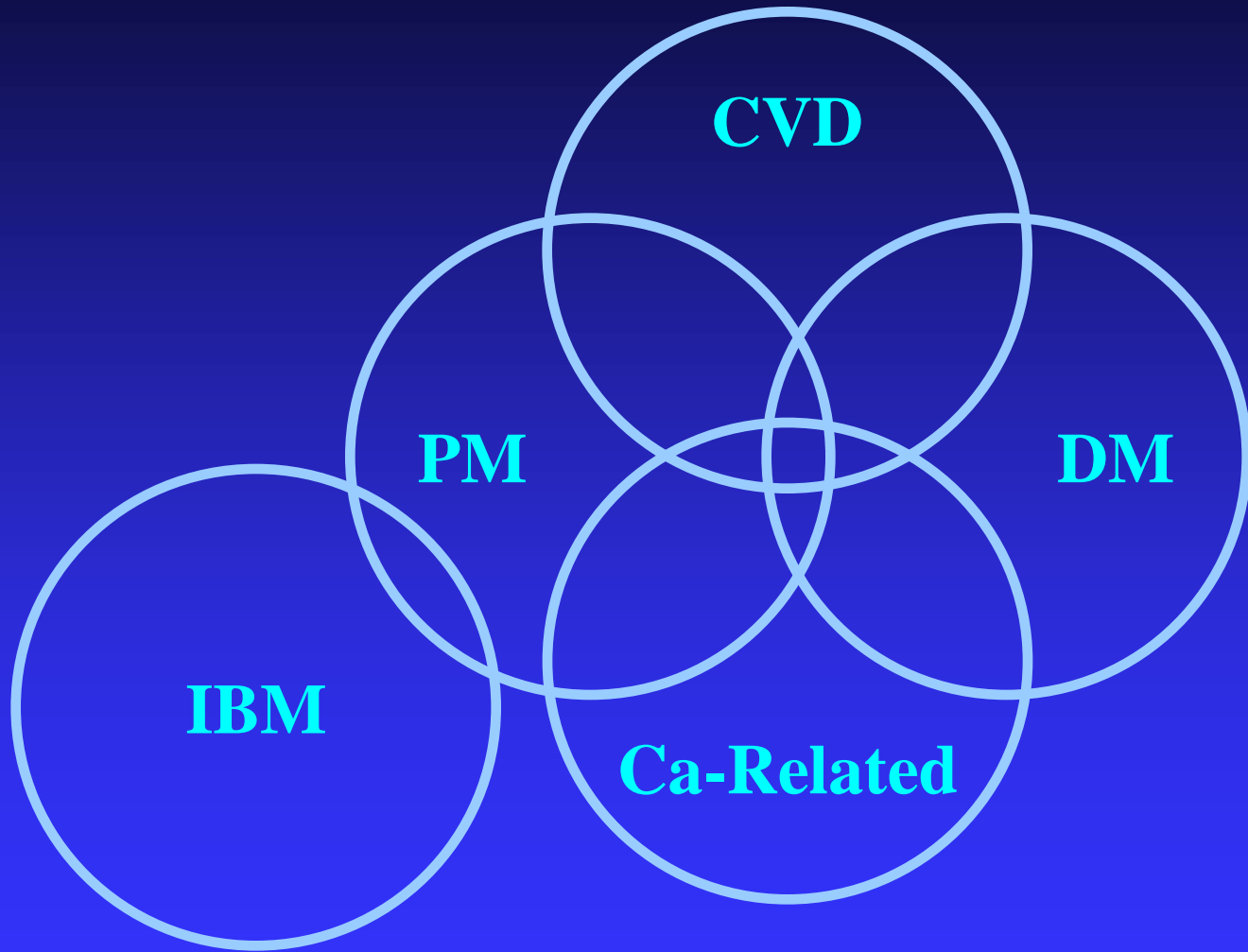
Not all High CK Levels are the due to Polymyositis! Other causes include:

- Racial differences
- Trauma
- Exercise
- Drugs/Toxins
- Carrier-states
- Pre-disease
- Benign (cause unknown)

Although the criteria are nonspecific, when occurring together, and without other explanations, they allow the diagnosis of an idiopathic inflammatory myopathy,

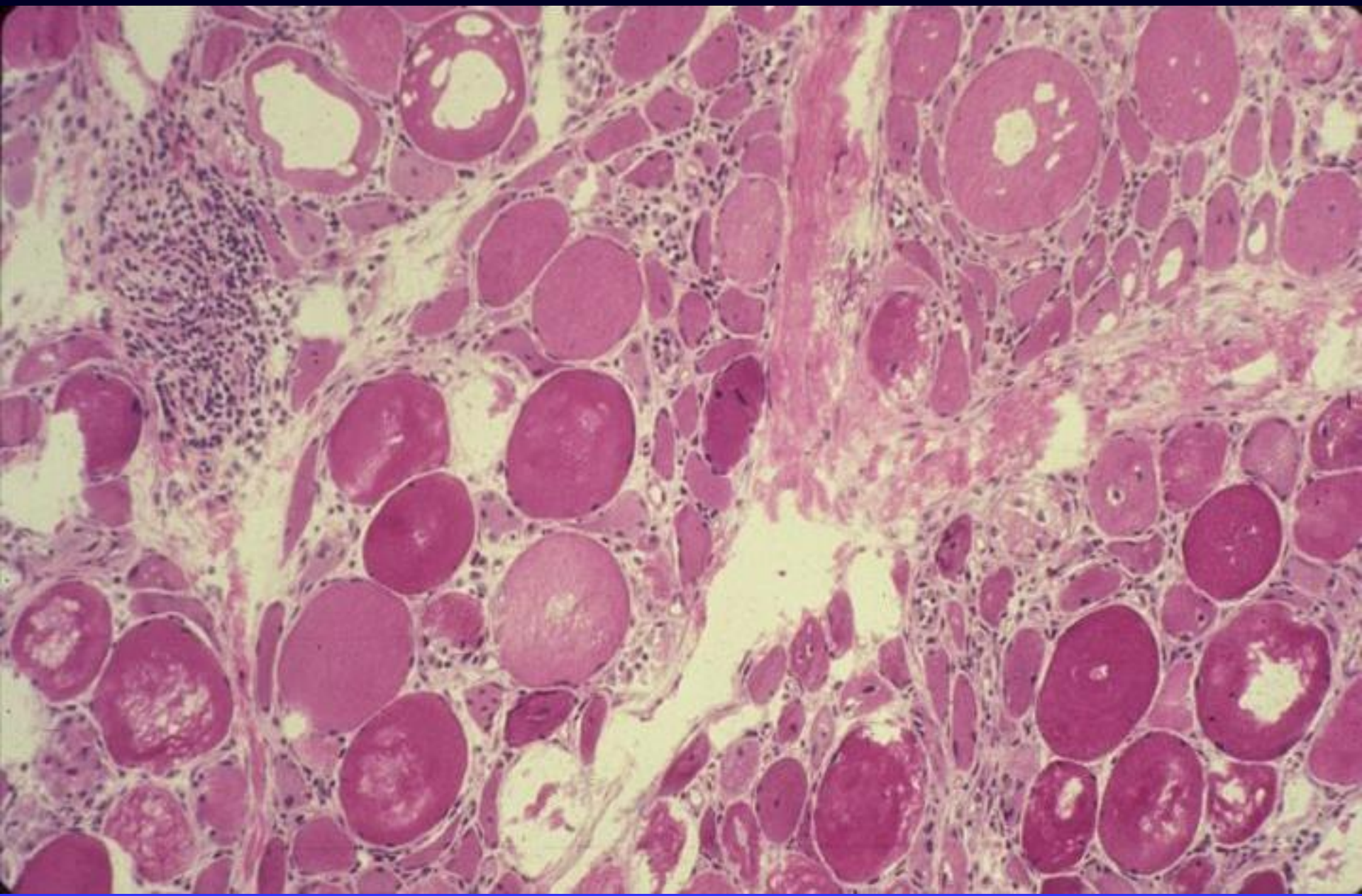
Idiopathic Inflammatory Myopathies

- Polymyositis
- Dermatomyositis
- Myositis with associated Collagen Vascular Disease
- Myositis with Malignancy
- Inclusion Body Myositis



Polymyositis

- Proximal muscle weakness
- Elevated CPK
- Myopathic EMG
- Inflammation on histology



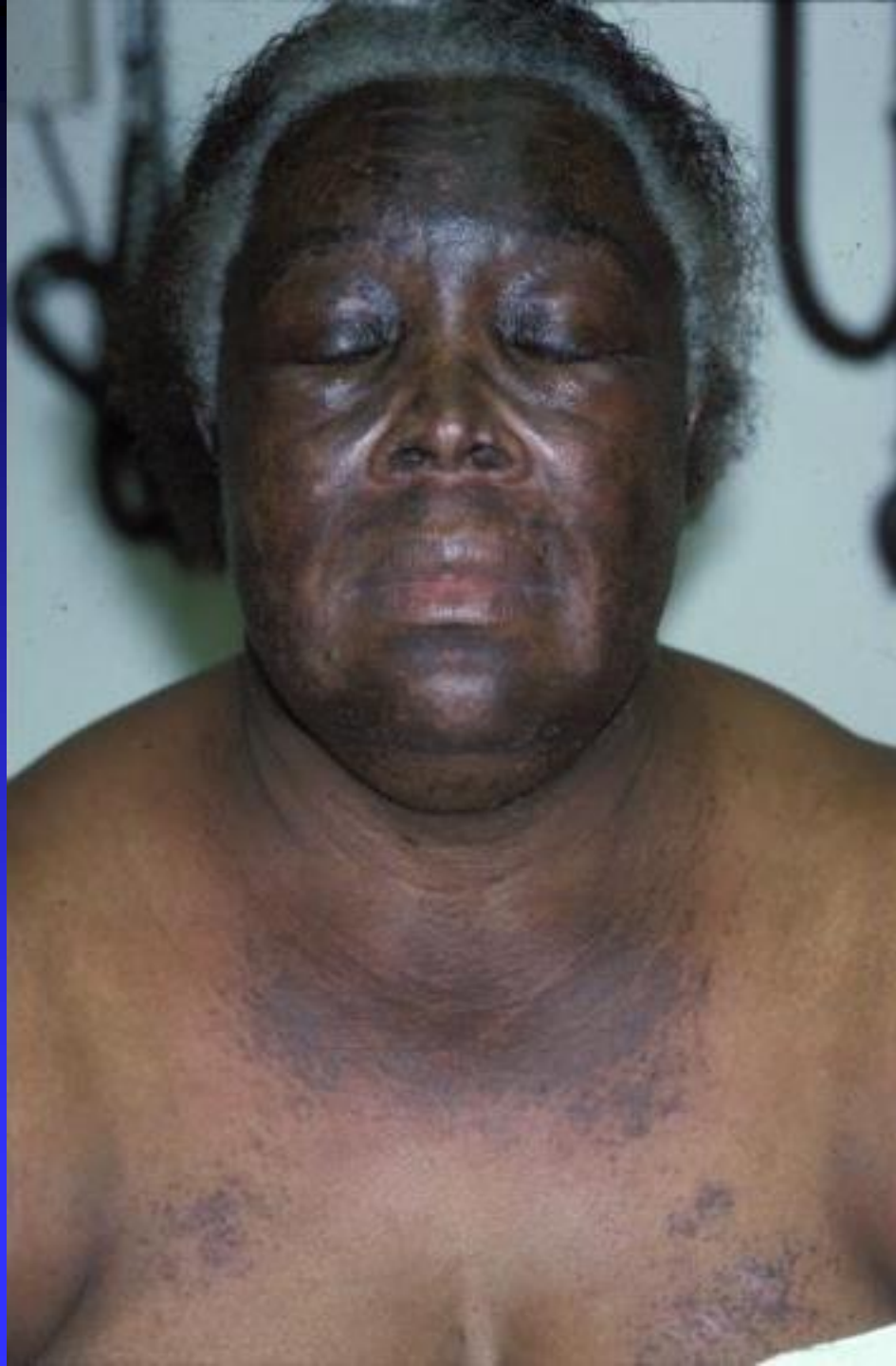
Dermatomyositis

- Polymyositis plus rash
- A different disease
- Different diseases



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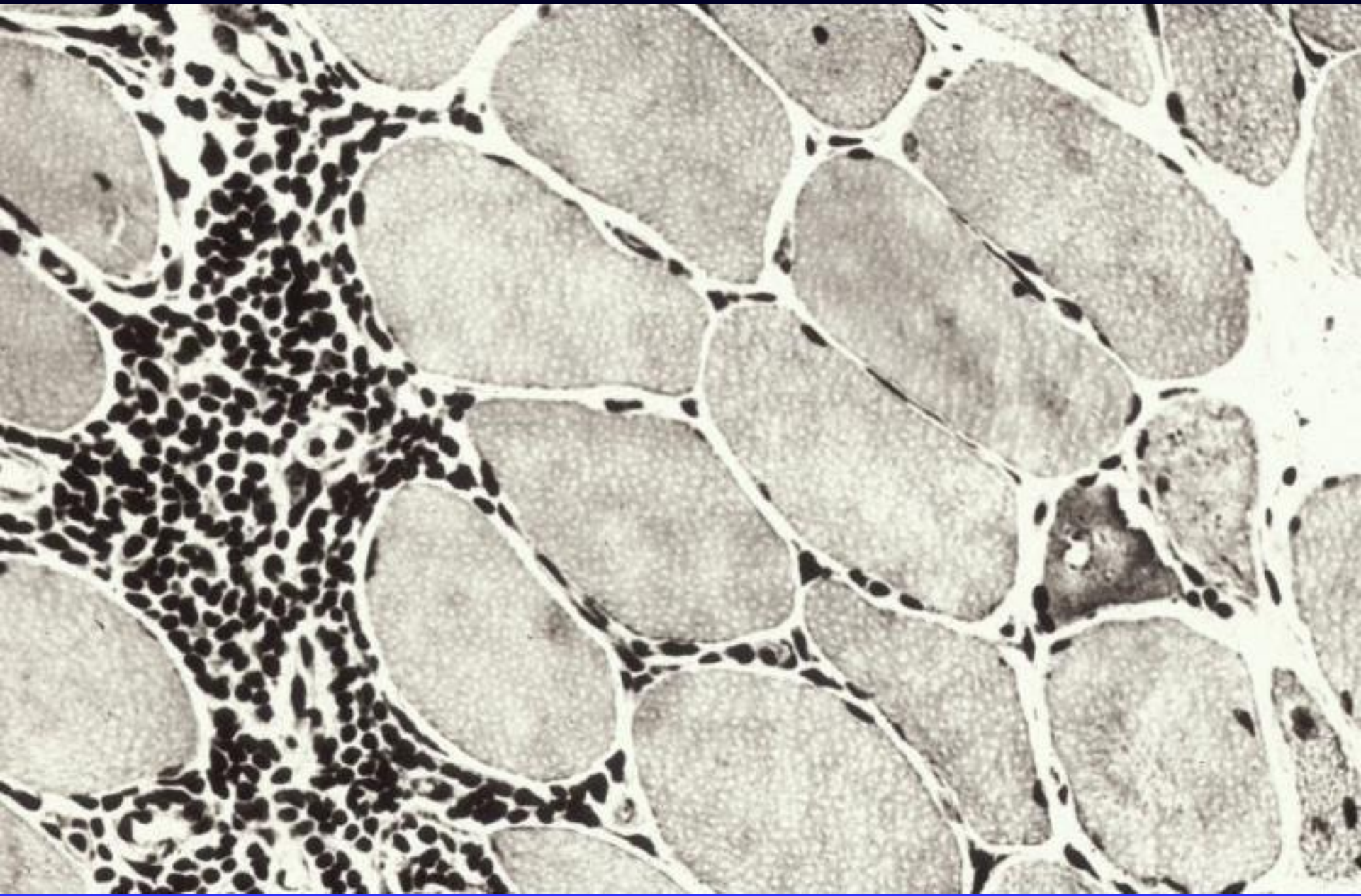


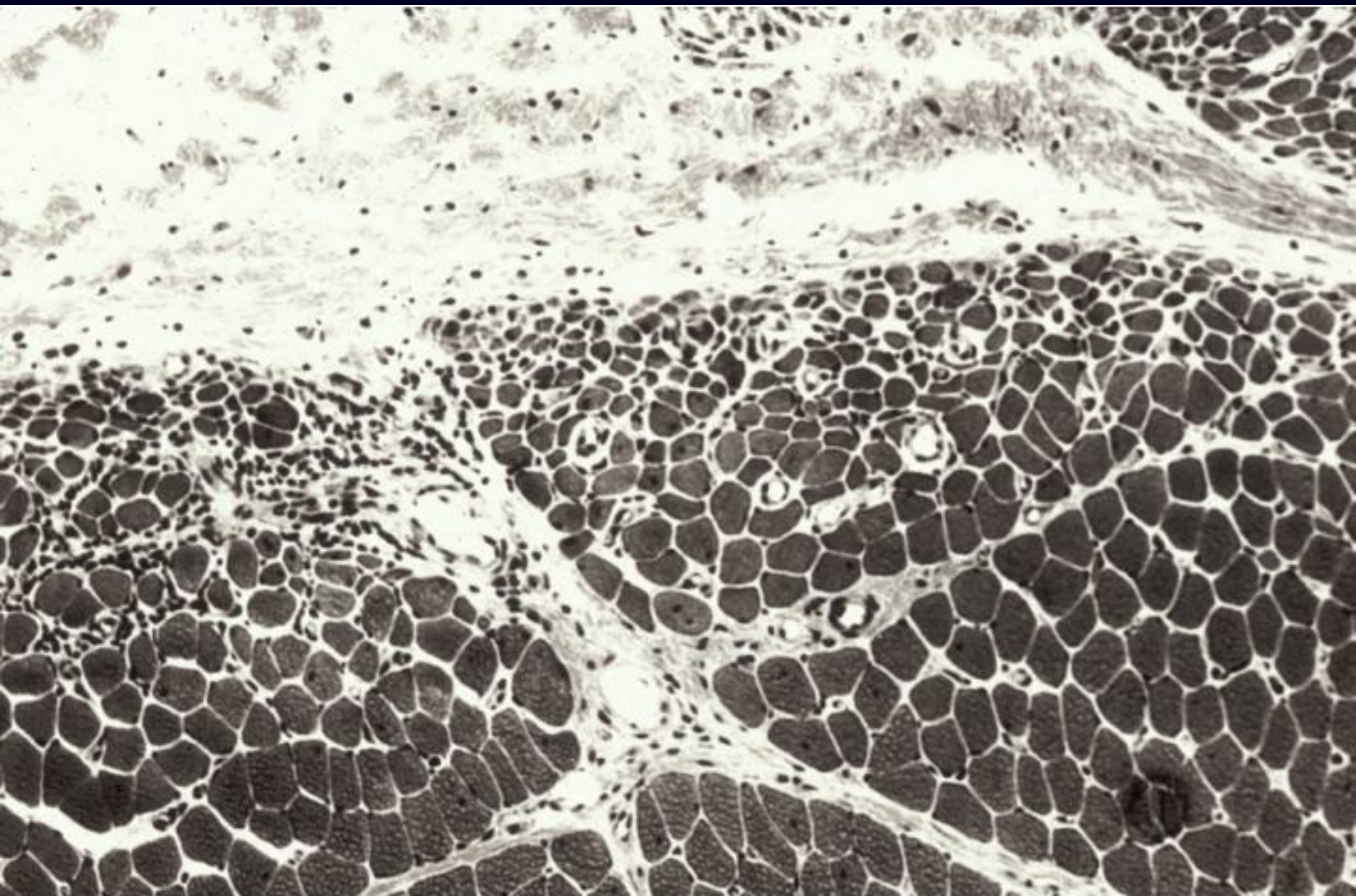












Dermatomyositis

■ Subsets

- ◆ Adult dermatomyositis
- ◆ Juvenile dermatomyositis
- ◆ Amyopathic dermatomyositis







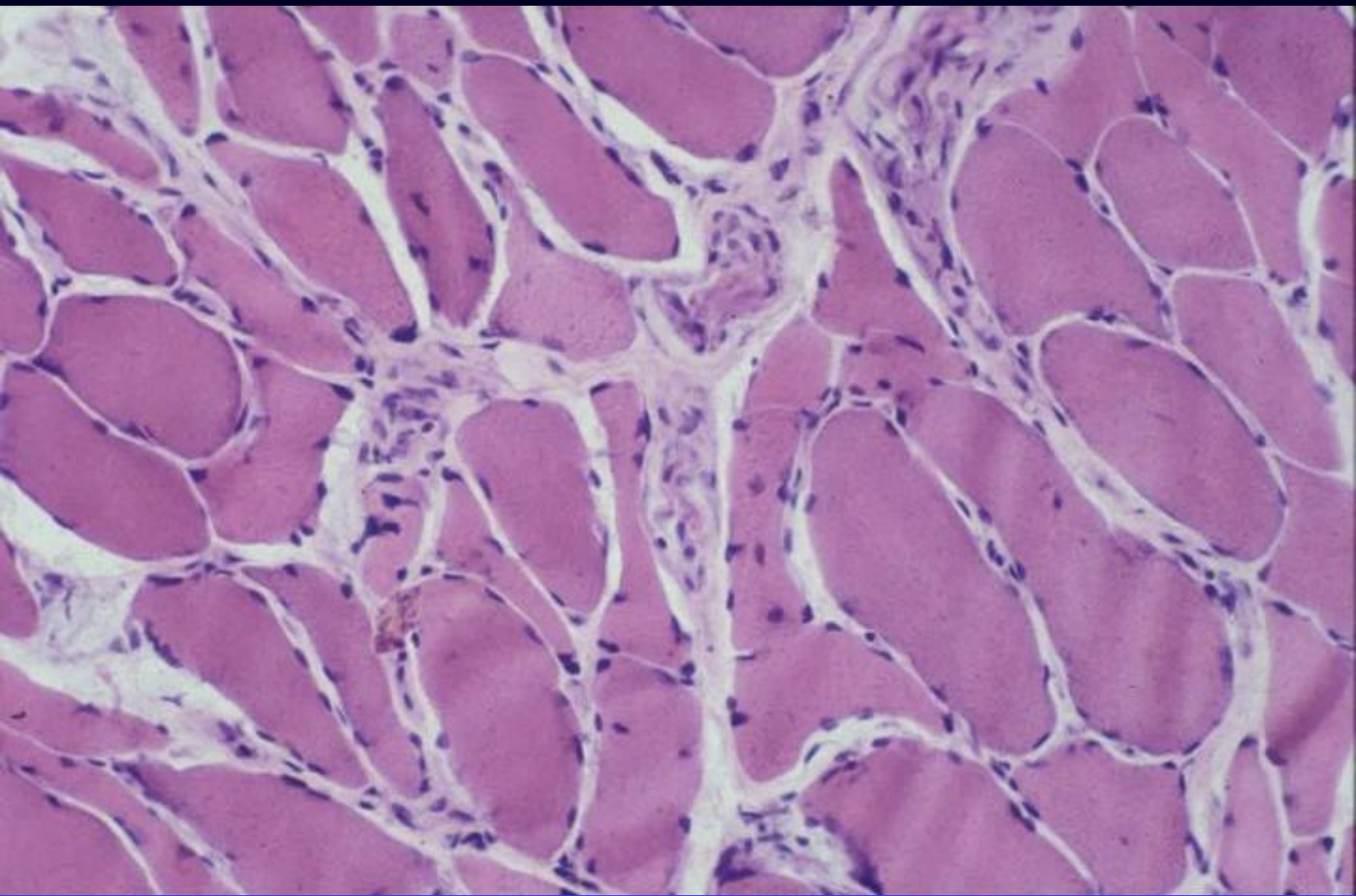
Inclusion Body Myositis – Clinical

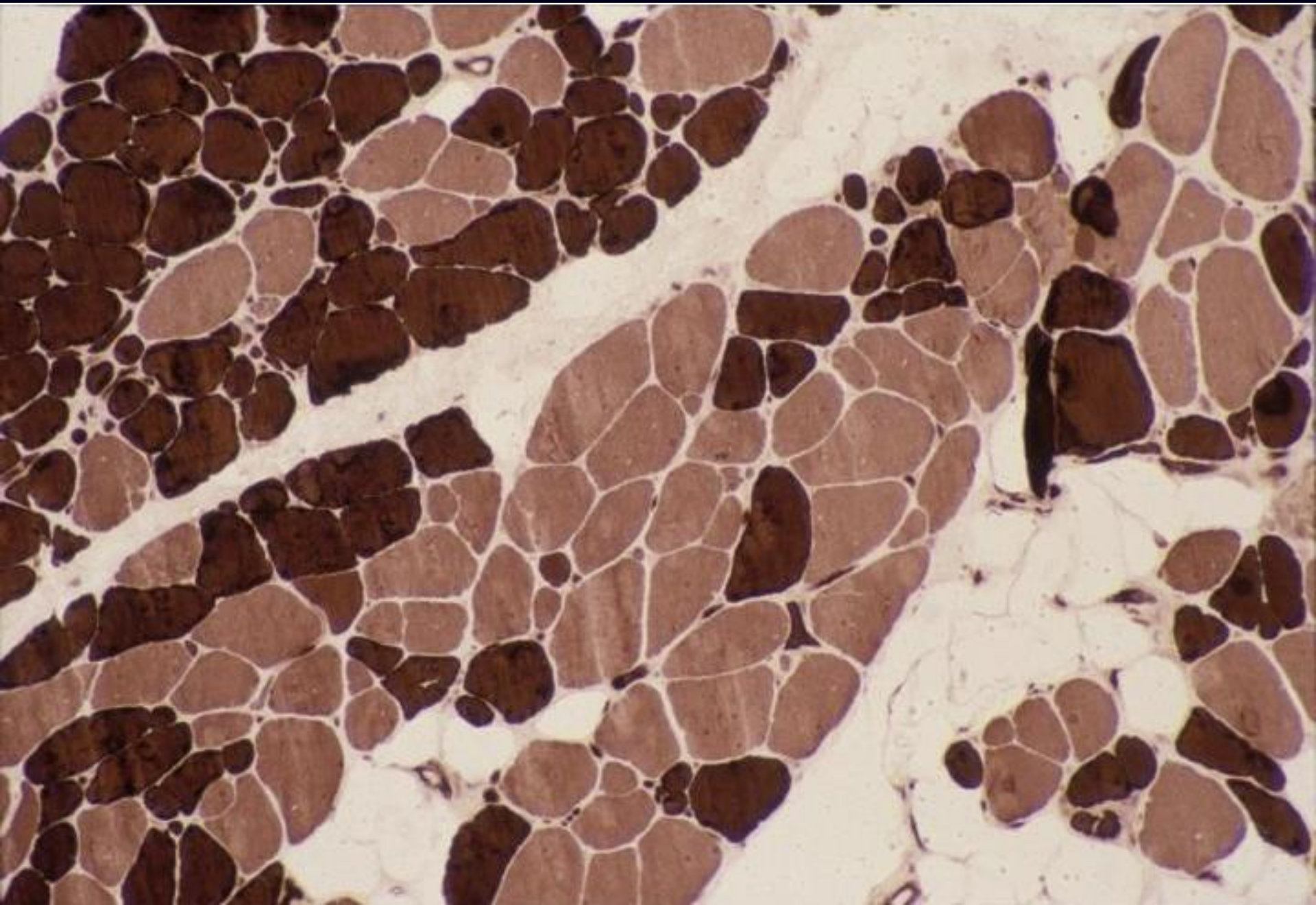
- Weakness
 - ◆ Proximal and symmetric
 - ◆ Distal
 - ◆ Asymmetric
- Response to Therapy
 - ◆ Poor if any

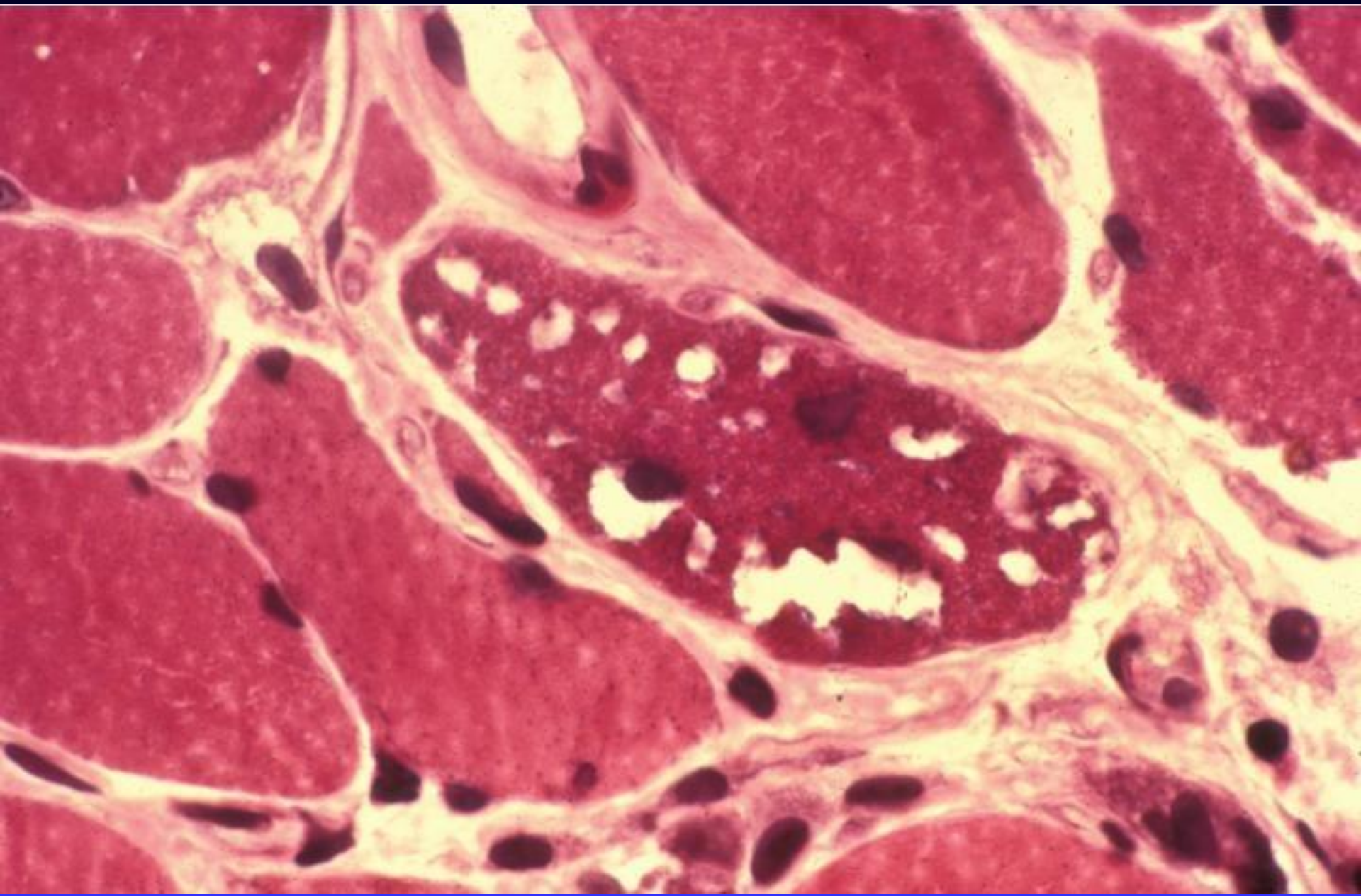


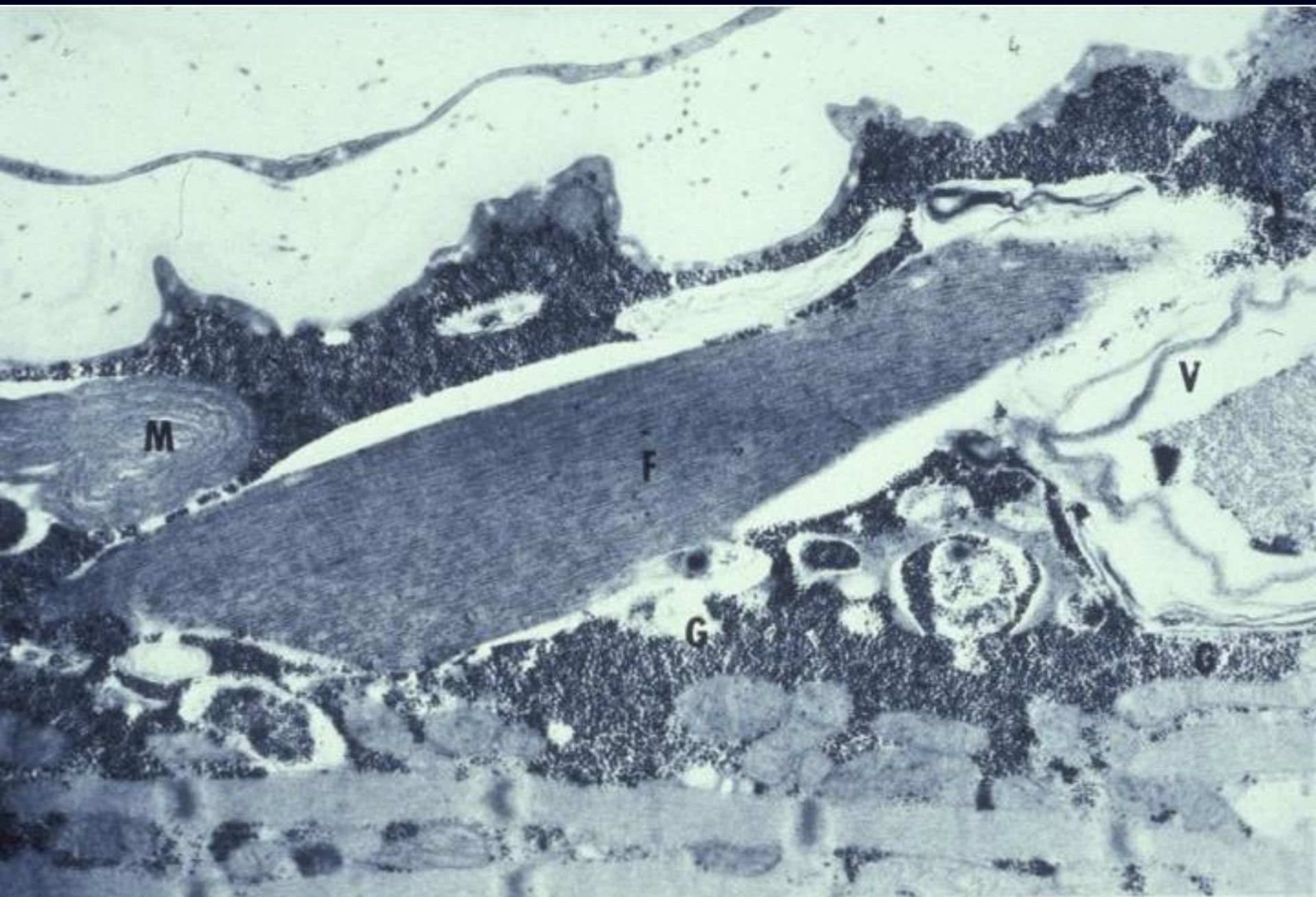
Inclusion Body Myositis

- Cellular infiltrate-like polymyositis but disappears
- Lined vacuoles
- Inclusions
- Amyloid deposits
- Mitochondrial abnormalities









Myositis and Connective Tissue Diseases

- Systemic lupus erythematosus
- Scleroderma
- Mixed connective tissue disease

Myositis and Cancer

- Increase risk with dermatomyositis
- Cancers are those most common for age and gender except for ovarian cancer
- Risk is greatest within one year of diagnosis
- Treatment of cancer often treats the myositis

Prognosis

■ PM and DM

- ◆ 55% do great
- ◆ 35% have variable results
- ◆ 10% do poorly

■ IBM

- ◆ Does not respond to drug therapy
- ◆ Typically progresses slowly

Myositis Specific Autoantibodies

- May help predict outcomes

Anti-Synthetase Syndrome

- Polymyositis > dermatomyositis
- Interstitial lung disease
- Fever
- Arthritis
- Raynaud's
- Mechanic's hands
- Difficult to treat

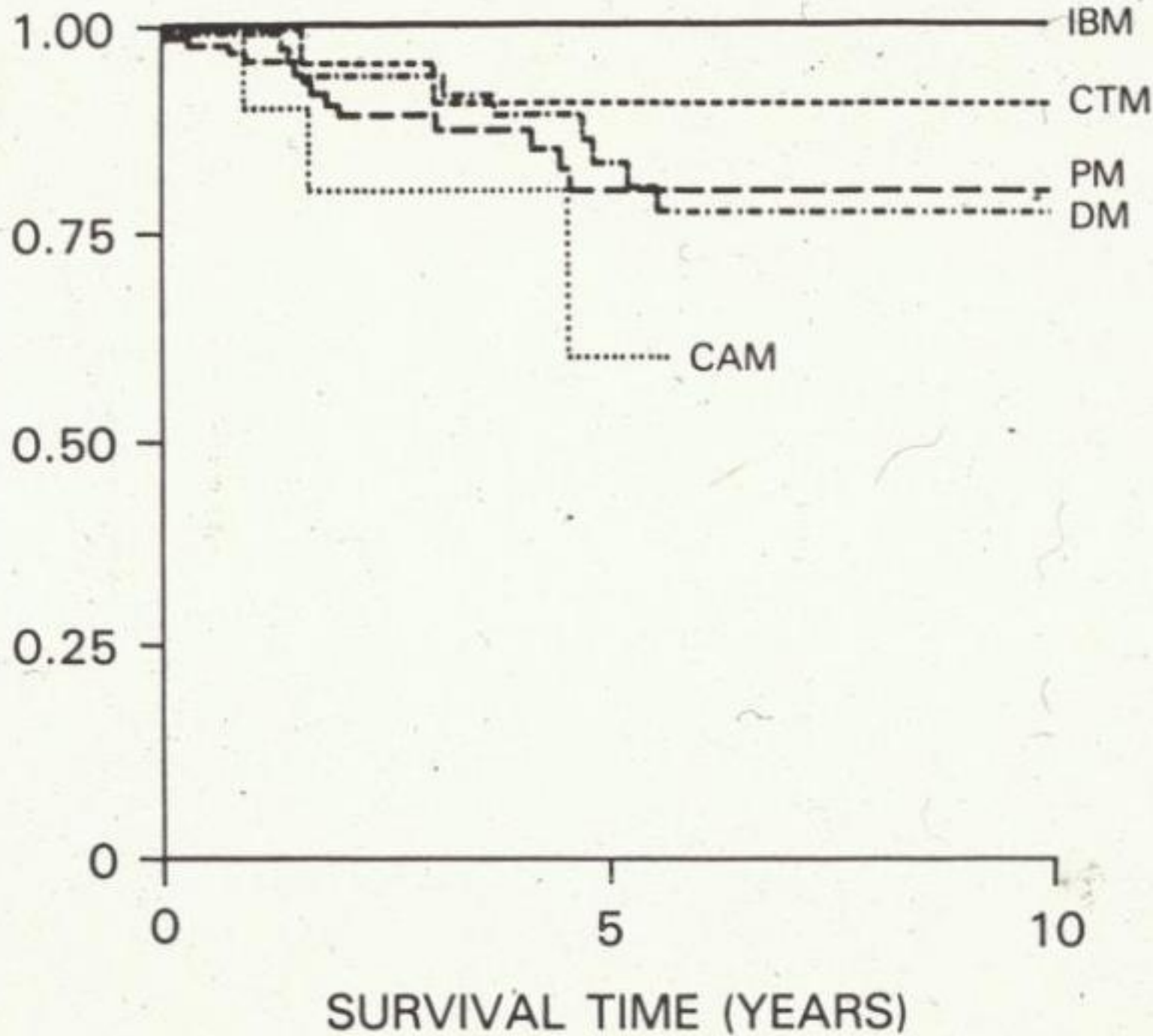
Anti-SRP

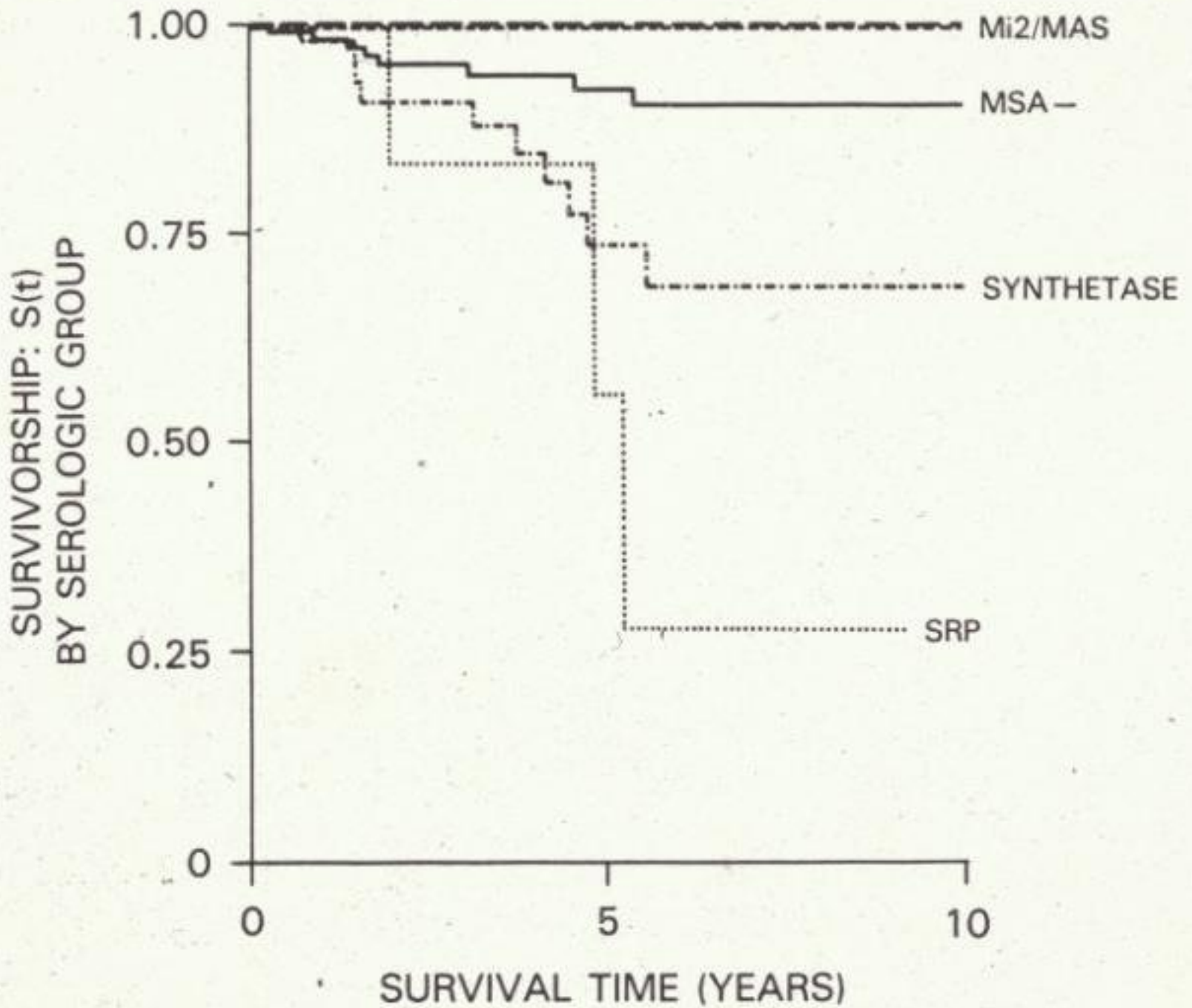
- Polymyositis >>> dermatomyositis
- Cardiomyopathy
- Distal weakness
- Very poor prognosis*

Anti-Mi 2

- Dermatomyositis
- Excellent prognosis*

SURVIVORSHIP: S(t)
BY CLINICAL GROUP





Although the criteria are nonspecific, when occurring together, and without other explanations, they allow the diagnosis of an idiopathic inflammatory myopathy,

*Management of Inflammatory
Myopathy*

Impact of Cortisone on Polymyositis

- Changed the mortality from over 50% to less than 10%
- Steroid therapy may prove curative to 50%

Exercise is good!

- Aerobic
- Anaerobic

Both have been shown to improve strength
and have anti-inflammatory effects

Controlled Trials

- Azathioprine + Prednisone
 - ◆ Bunch, 1981, 20 patients
- Plasma and leukophoresis
 - ◆ Miller, 1992, 39 (26) patients
- IV immune globulin
 - ◆ Dalakas, 1993, 15 patients
- Methotrexate/azathioprine – IV methotrexate
 - ◆ Villalba, 1997, 30 patients

RIM Trial

- 202 subjects
 - ◆ 75 PM
 - ◆ 55DM
 - ◆ 52 JDM
- Negative results
- Regardless, over 80% of patients met the definition of improvement and average dose of prednisone dropped significantly

Others Used

- Cyclosporine
- Cyclophosphamide
- Chlorambucil
- Etanercept
- Infliximab
- Intravenous immune globulin
- Mycophenolate
- Plasmapheresis
- Rituximab
- Tacrolimus

Lack of Response

- Treatment insufficiency
 - ◆ (not enough drug prescribed or taken)
- Refractory disease
 - ◆ (IBM, interstitial lung disease, cancer, anti-SRP)
- Steroid toxicity
- Incorrect diagnosis