

# **54 YEAR OLD MAN WITH BILATERAL FOREARM ATROPHY**

Samah K Aburahma, MD  
University of Colorado Denver

# 54 YEAR OLD MIXED RACE GENTLEMAN...

10 year history of progressive bilateral hand weakness

Onset of fluctuating bilateral arm weakness, stepwise progression over time

Radiating pain down the arms to the hands

Setting of chronic neck pain

# HISTORY

PMhx of arthritis, hypertension, hypercholesterolemia, peptic ulcer disease, neck pain, and low back pain with lumbar spine surgery in the past

Family history noncontributory

Medications: Atorvastatin, Amitriptyline, cyclobenzaprine, omeprazole, lisinopril, Zolpidem, oxycodone

Prior cervical spine CT myelogram that showed foraminal narrowing without spinal stenosis

Prior labs showed CPK 300-400 range, strongly positive ANA titers, negative GM1/MAG serologies, and negative paraneoplastic/ myositis panels

# EXAMINATION

Normal mental state examination and cranial nerve examination

Mild left sided scapular winging

Atrophy of bilateral forearm muscles

Bilateral distal upper limb weakness involving wrist flexors, wrist extensors, and intrinsic hand muscles MRC grade 3/5

Lower limb power generally preserved

Deep tendon reflexes normal

Sensation diminished over the medial arms, hands, and back



**INVESTIGATIONS?**

# NEUROPHYSIOLOGY

## Nerve conduction studies:

- Normal bilateral upper and lower limb sensory nerve conduction studies, including lateral and medial antebrachial cutaneous sensory studies.
- Normal motor nerve conduction studies and F wave latencies

## EMG of left upper and lower limb muscles

- Abnormal spontaneous activity in the deltoid, biceps, flexor carpi radialis, and extensor carpi radialis muscles
- Motor unit potentials of short duration, decreased amplitude, and polyphasic morphology with early recruitment were seen in above muscles
- Motor unit potentials in the tibialis anterior and medial gastrocnemius muscles were of normal to increased amplitude and duration.

***Consistent with a myopathy with muscle membrane instability involving the left upper extremity.***

# LAB INVESTIGATIONS

ANA strongly positive 1:320, RF negative, ESR normal

CK 200-400 range

GM1 antibodies negative

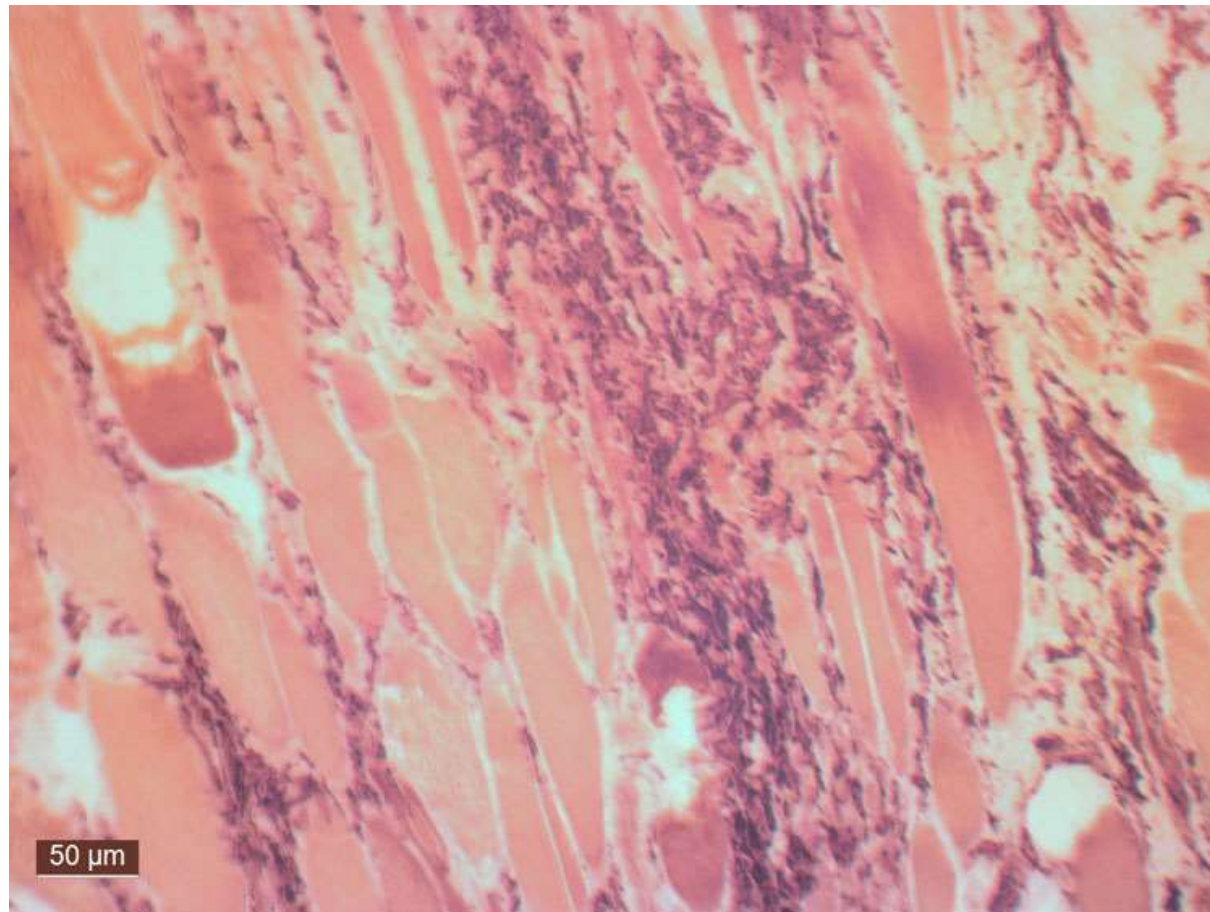
Anti MAG negative

SPEP normal

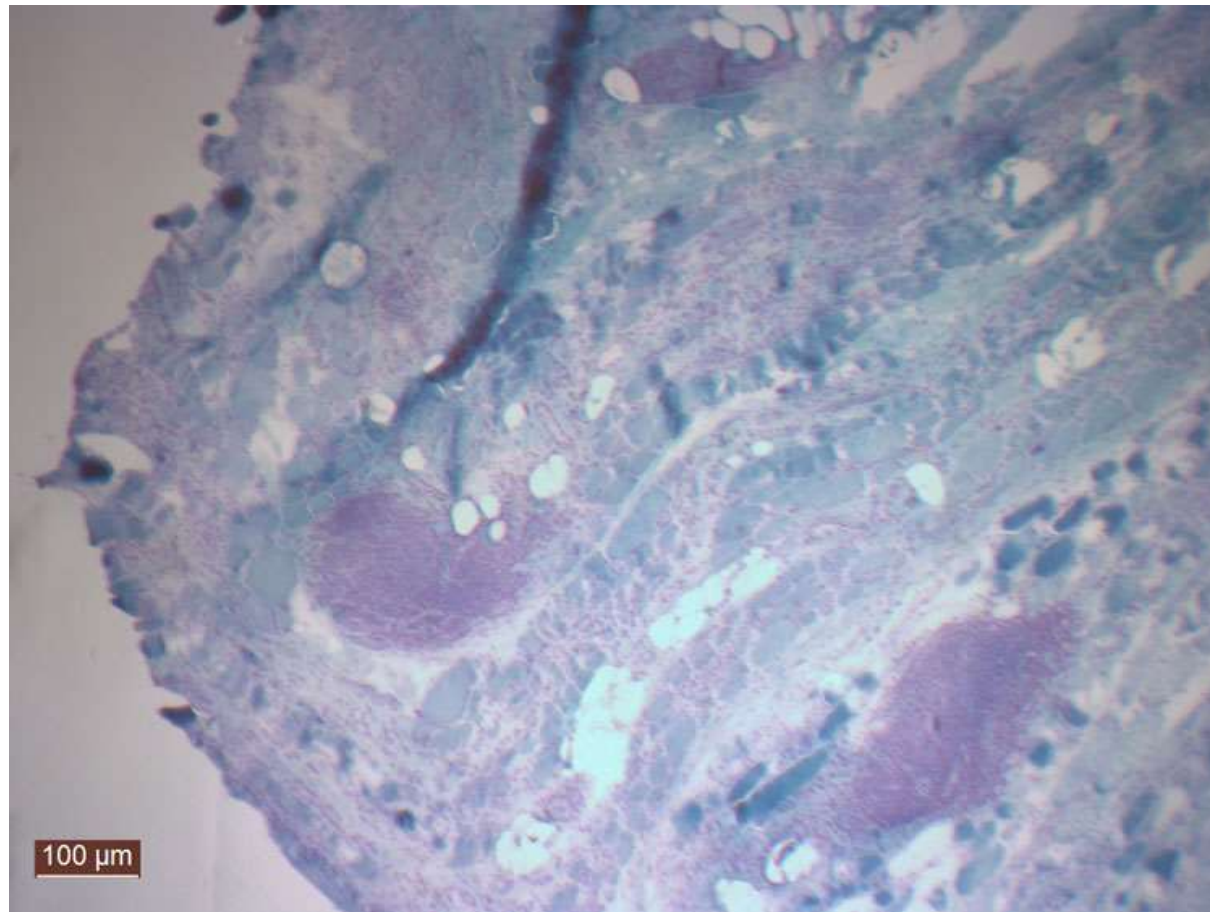
Paraneoplastic panel negative

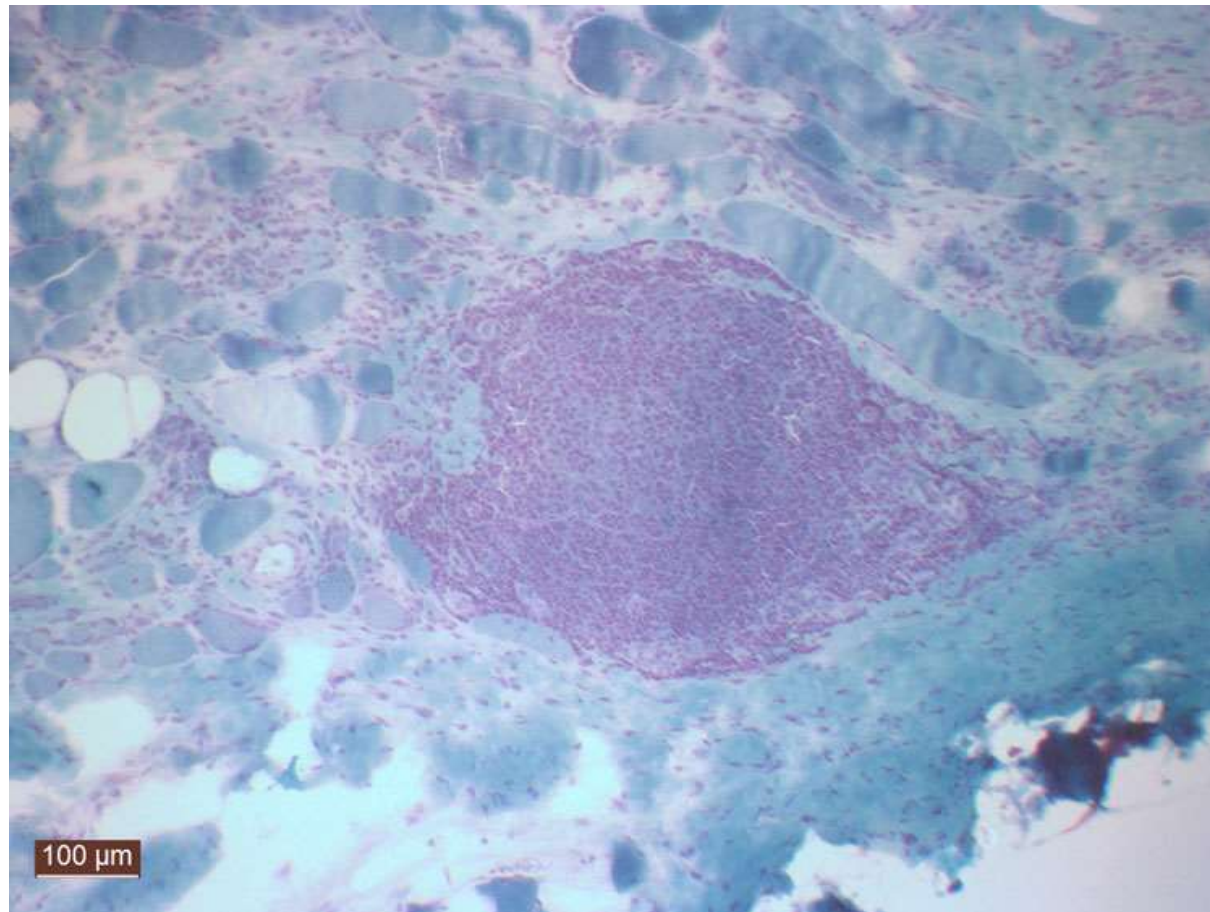
Anti GAD negative

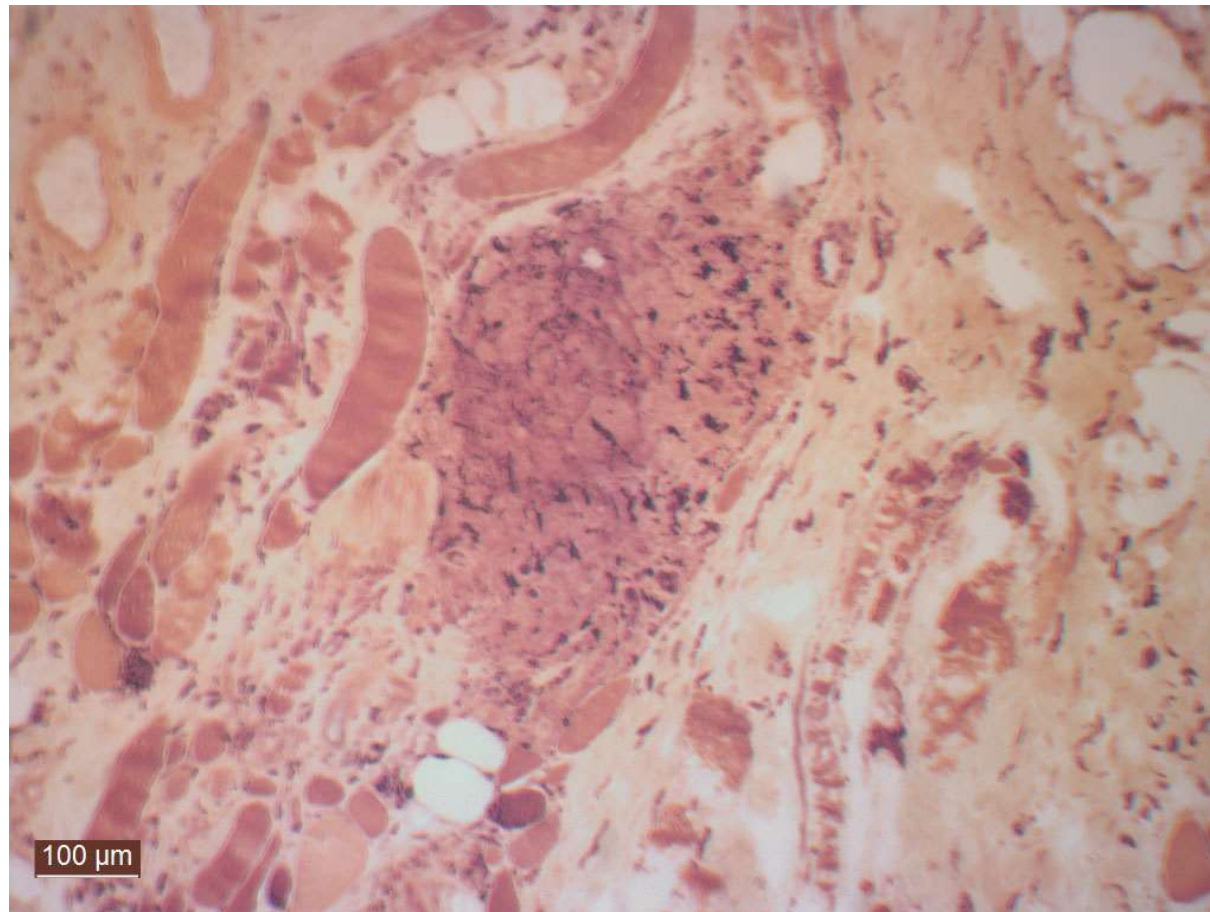
HMGCR and myositis panels negative



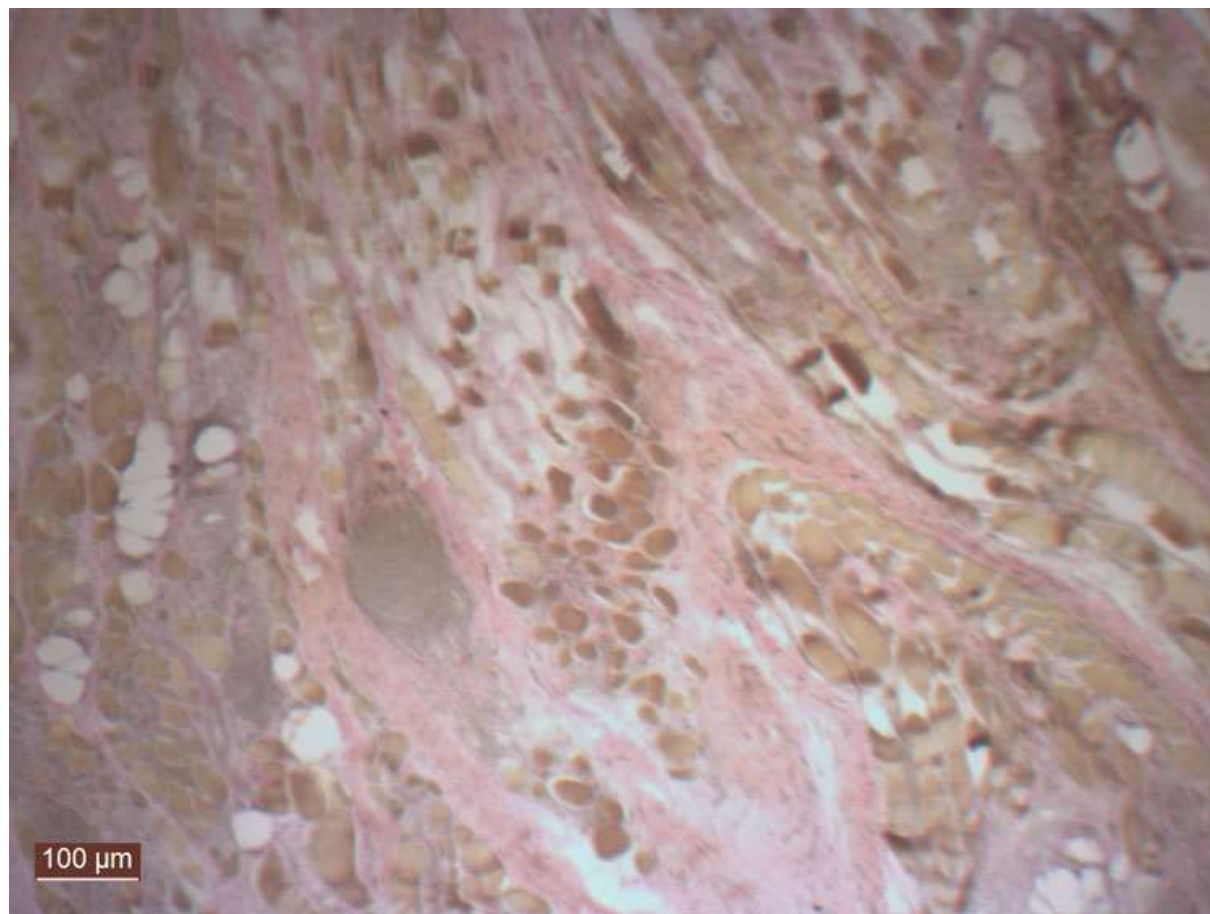


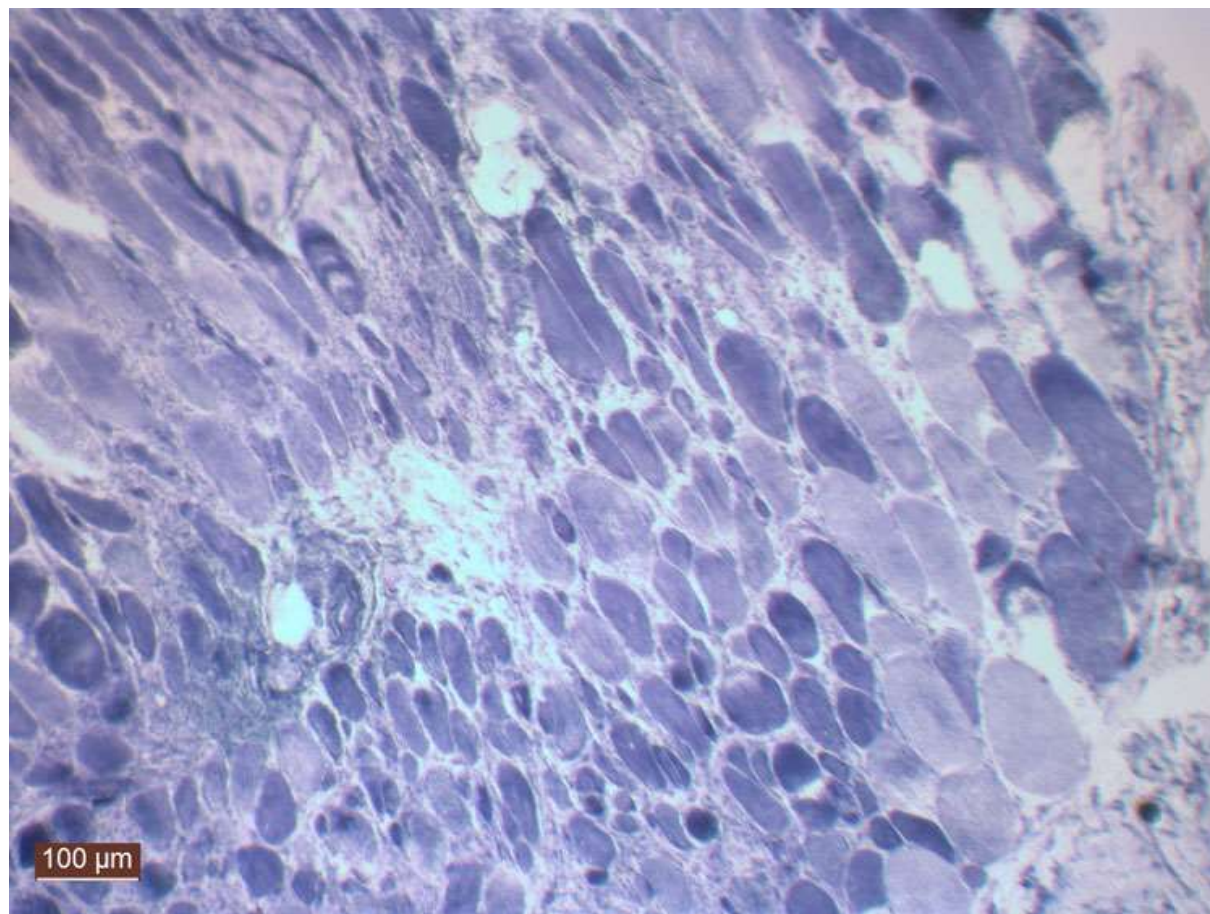


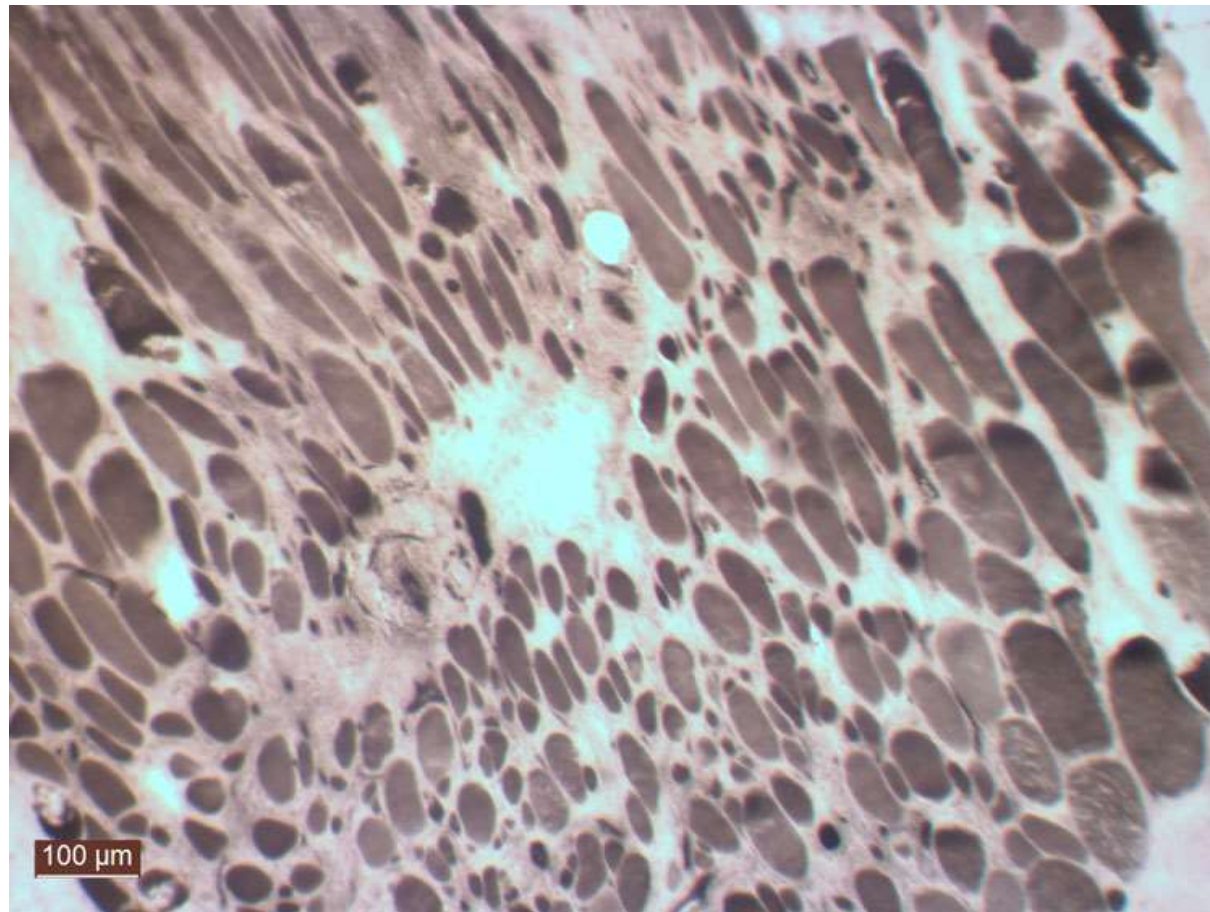




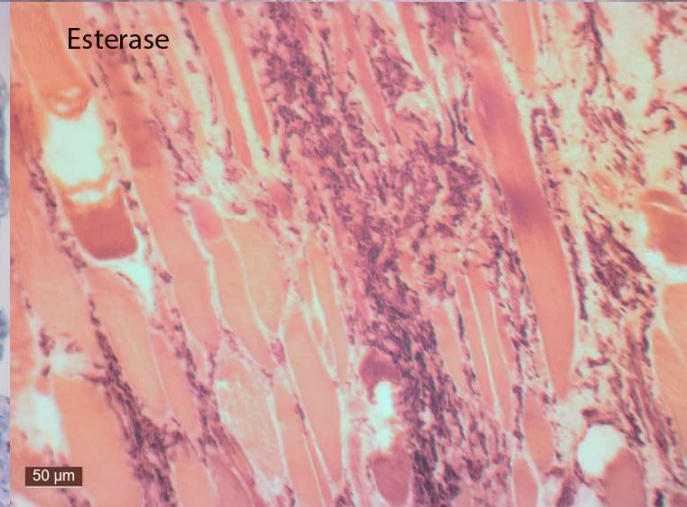
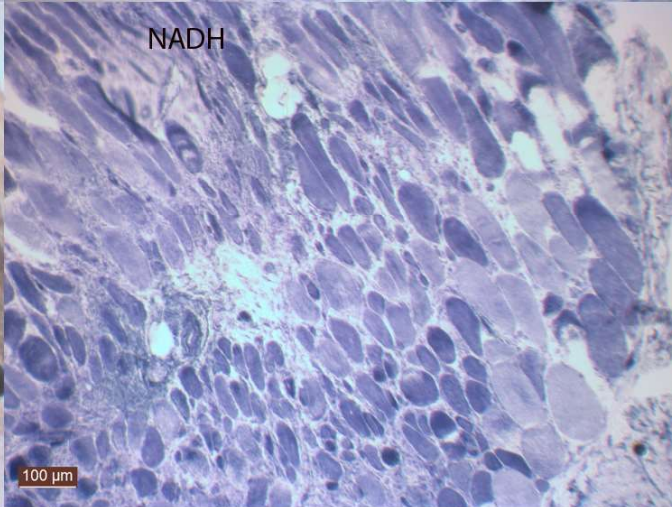
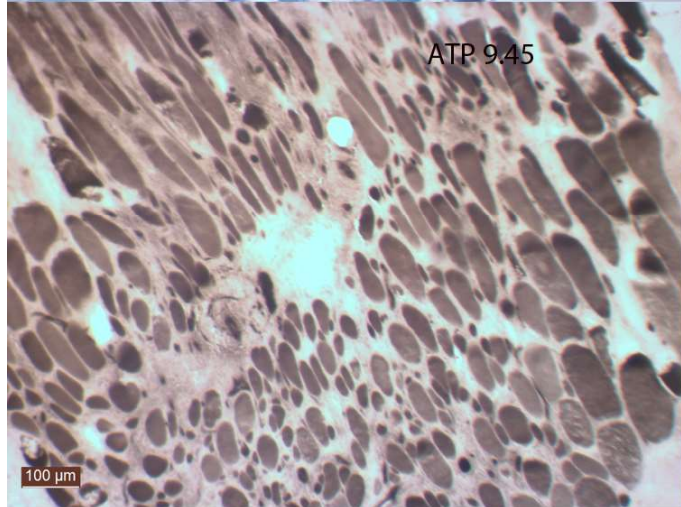
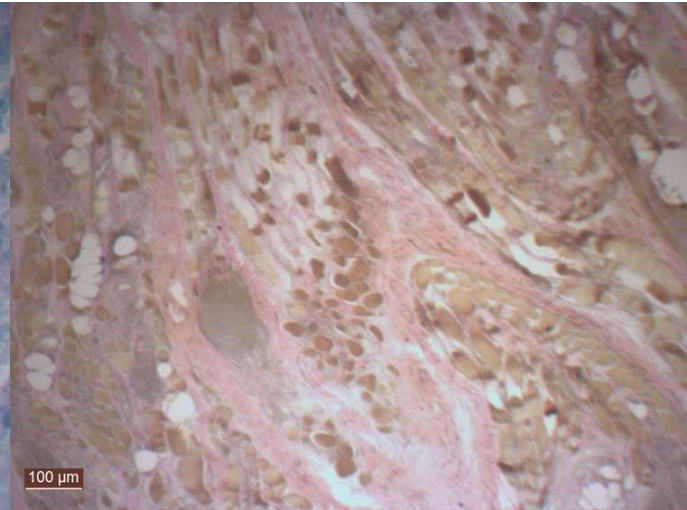
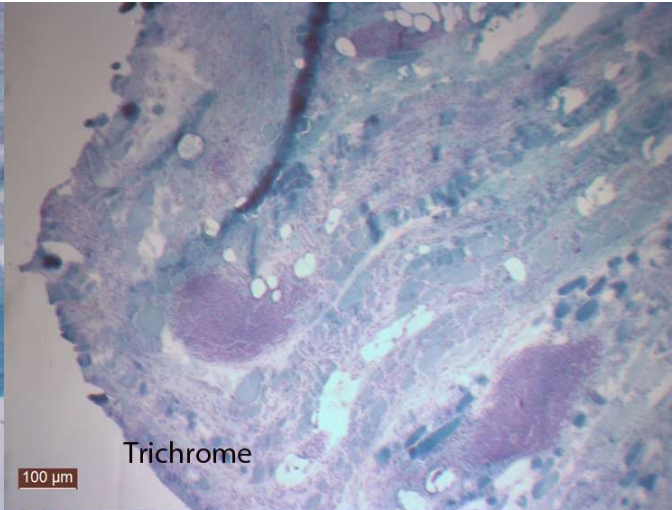
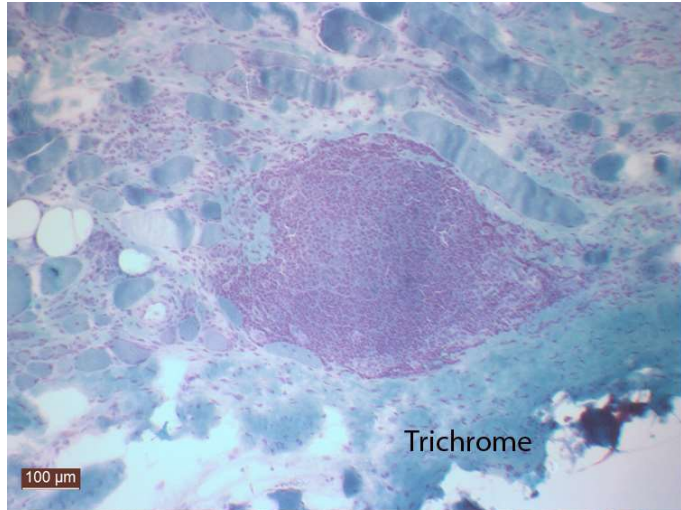












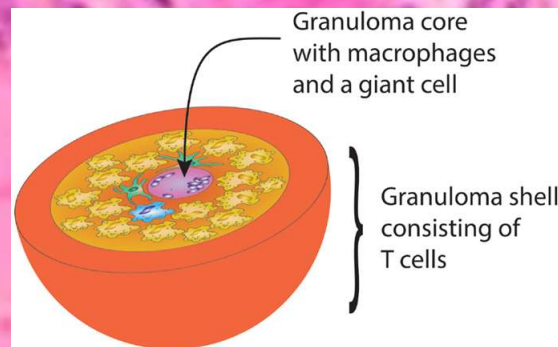


# GRANULOMA...WHAT'S IN A NAME

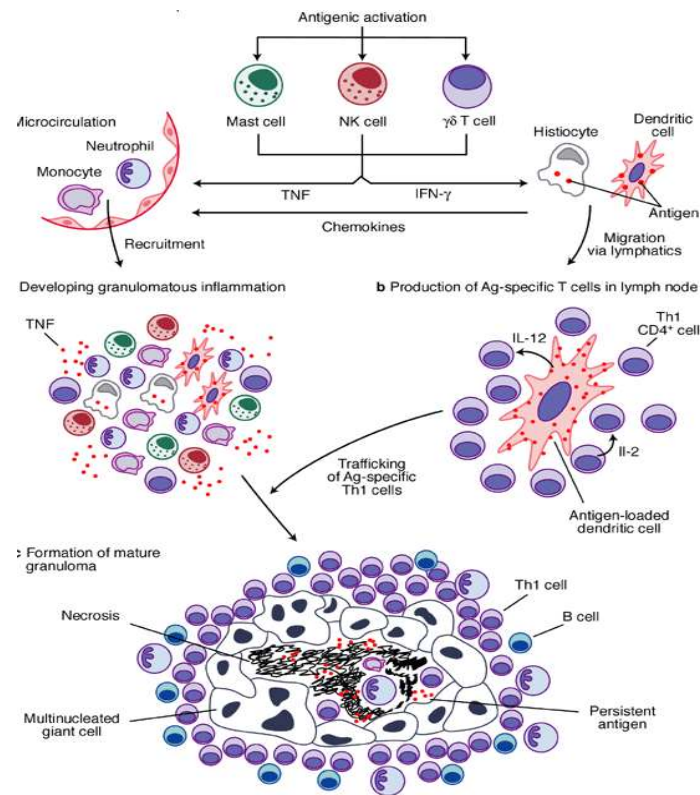
**Granuloma: Focal aggregates of macrophages, epithelioid histiocytic cells, inflammatory lymphocytes, and occasional giant cells of Langerhans “in muscle More than 60% macrophages as evidenced by esterase staining or CD 68**

## **Key players in pathogenesis**

- Antigenic triggers: Mycobacteria, P acnes
- Macrophages: primarily the M2 polarity
- CD4+ T helper cells
- B cells







# GRANULOMATOUS MYOSITIS (GM)

Granuloma formation in skeletal muscle

Variable response to immunosuppressive therapies

No evidence of systemic sarcoidosis



# GRANULOMATOUS MYOSITIS

Sarcoidosis

Idiopathic

Infections

Myasthenia gravis

Autoimmune disease

Malignancy

*SILVERSTEIN A, SILTZBACH LE. MUSCLE INVOLVEMENT IN SARCOIDOSIS. ASYMPTOMATIC, MYOSITIS, AND MYOPATHY. ARCH NEUROL 1969;21:235-41.*

Asymptomatic granulomatous muscle involvement in up to 50-80%

Symptomatic involvement

Chronic

- Most frequent
- Bilateral, “proximal with sarcoid, distal with idiopathic”, with muscle atrophy
- Insidious, late onset
- Acute
  - Younger patients
  - Myalgias and weakness
  - Early stages of the disease
- Nodular
  - Least common, painful solitary nodules, early disease stage

# SARCOIDOSIS AND GRANULOMATOUS MYOSITIS

## Several case series of GM

- Mozaffar et al: 10 patients with GM, 4 of whom had systemic sarcoidosis
  - Sarcoid GM was proximal and steroid responsive, while idiopathic GM presented distally and was steroid resistant...*granulomatous myopathy*
- Le Roux et al: 13 cases with GM; 8 sarcoid myopathy, 5 isolated GM
  - Sarcoid GM was proximal, with variable response to steroids
- Fayad et al: 5 cases of sarcoidosis with symptomatic muscle involvement
  - 3 with nodular muscle involvement, 2 with acute myositis
  - Variable response to steroids and frequent relapse

# GRANULOMATOUS MYOSITIS

## Laboratory evaluation

- CPK, inflammatory markers, TB

## Imaging:

- MRI muscle
- Gallium scintigraphy for muscle and extra muscular involvement

## Therapy

- Unpredictable response to steroids

# GRANULOMATOUS MYOSITIS...NOT SO SIMPLE

## GM and inclusion body myositis (IBM)

- Larue et al, 2017: Distal muscle involvement in GM can mimic IBM
- Sanmaneechai et al, 2014: Occurrence of histological features of IBM concomitant with GM
  - ? IBM and GM associated diseases, both share common immune pathways
- Sakai et al, 2015: Identification of granulomas in retrospective study of patients with clinicopathological diagnosis of IBM
  - ? New form of IBM associated with granuloma formation
- Zakaria et al, 2017: ?sarcoidosis as a possible trigger of IBM

# GRANULOMATOUS MYOSITIS...NOT SO SIMPLE

## Sarcoid myopathy and polymyositis

- Awano et al, 2015: a case of sarcoid myopathy with anti-Ku antibodies, and criteria compatible with polymyositis
- Toujani et al, 2014: a case of systemic sarcoidosis and anti-Jo antibodies, leading to a diagnosis of antisynthetase syndrome