Spider Bite Monoparesis

Carrell-Krusen Neuromuscular Symposium
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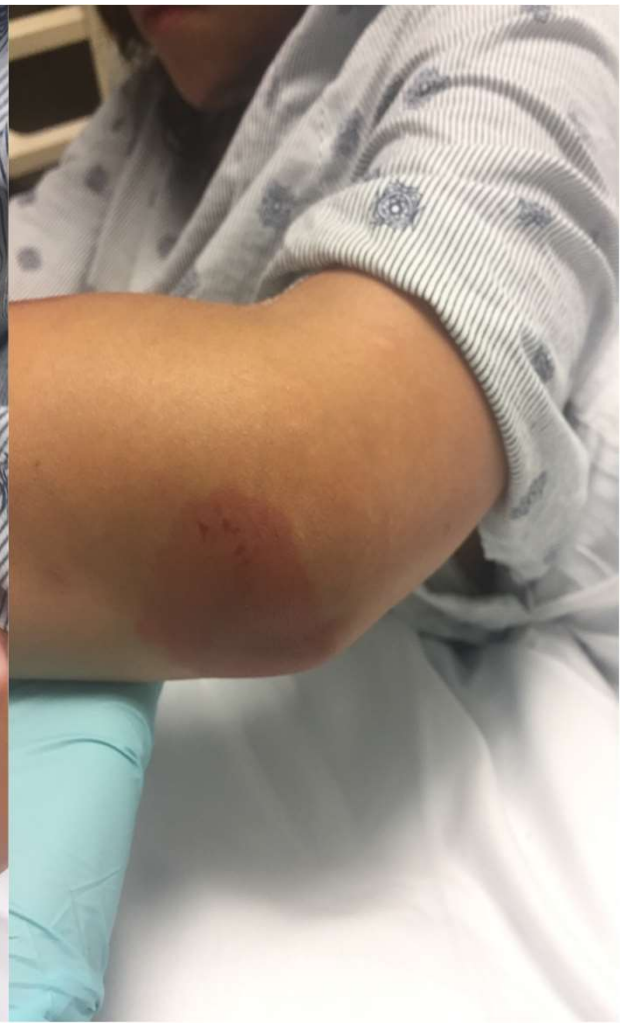
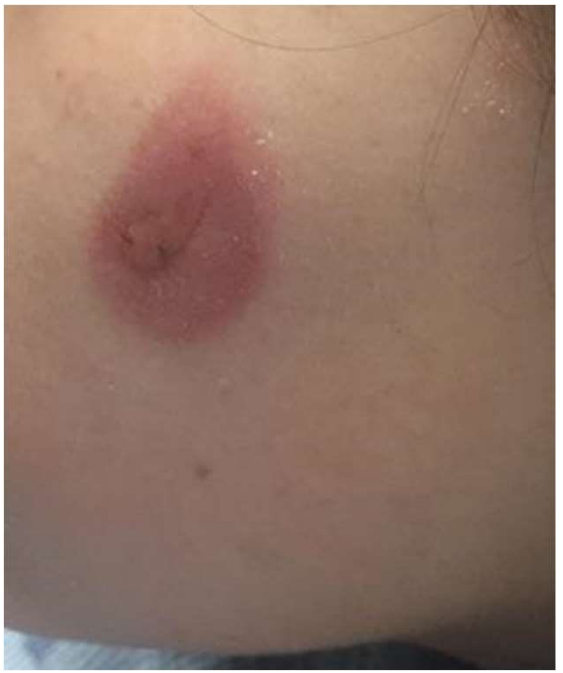
HPI

- 12 yr female with right arm cellulitis 1 month prior to presentation with acute onset of left arm monoparesis, swelling, and decreased sensation.
- Awoke with circumscribed erythematous patches on left arm and left cheek that were draining purulent material





Patient noted multiple small, black fuzzy spiders in her room the night prior to presentation.

Review of systems significant for 3 episodes of non-bloody vomiting on the morning of admission and **left arm pain of 2 days duration.**

Other Relevant History

PSH: None

Medications: None **Allergies:** NKDA **Immunizations:** Up-to-date

Birth History: Good prenatal care with no pregnancy complications. Born at term by SVD.

Social History: Lives with mother, step-father, brother and sister. No recent travel, no sick contacts. Has straight A grades in 7th grade.

Family History: Mother had prior ectopic pregnancy. No neurologic disorders.

Developmental History: No developmental delays

Neurological Exam on Admission

General: Active, alert, well-developed, well-nourished female

Cranial Nerves: CN II-XII intact. PERRLA, EOMI.

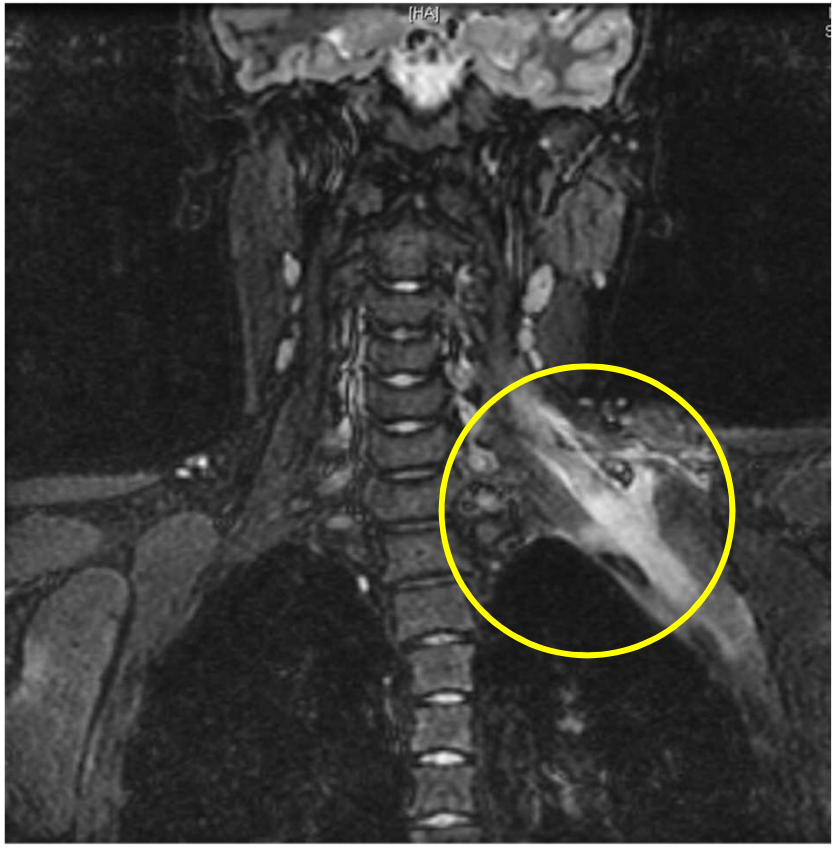
Motor: RUE 5/5 strength, **LUE 0/5 strength**

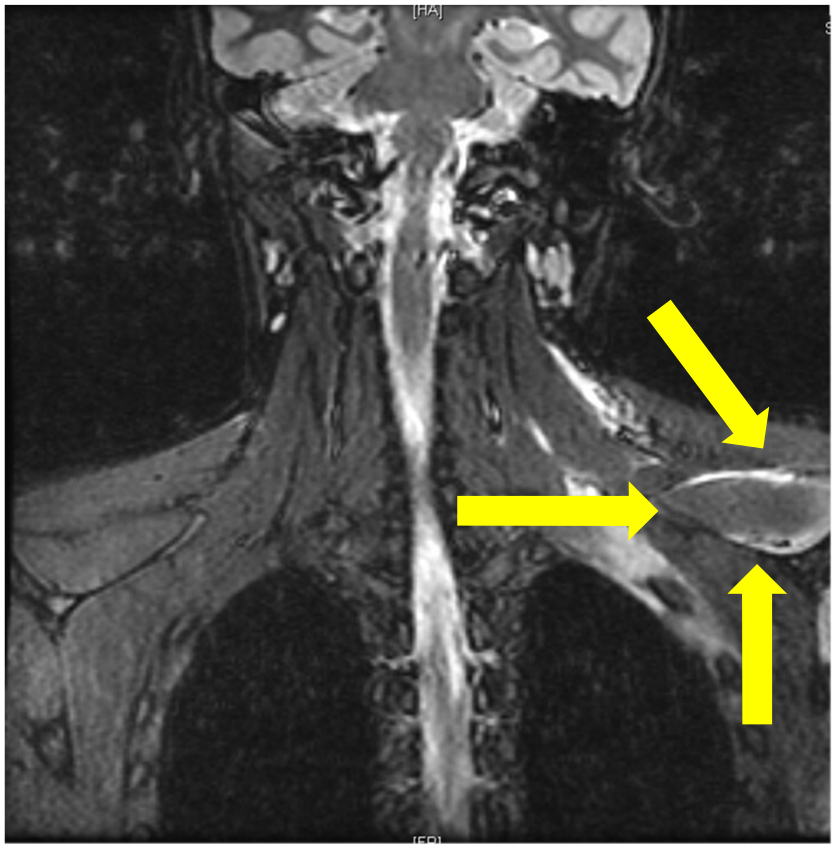
Sensory: hypoesthesia of left arm compared to right arm

- Right arm: light-touch/pain/temperature modalities intact.
- **Left arm: pinprick and temperature absent over arm, light-touch intact on left shoulder.**

DTRs: **2/4 except areflexia of left arm**

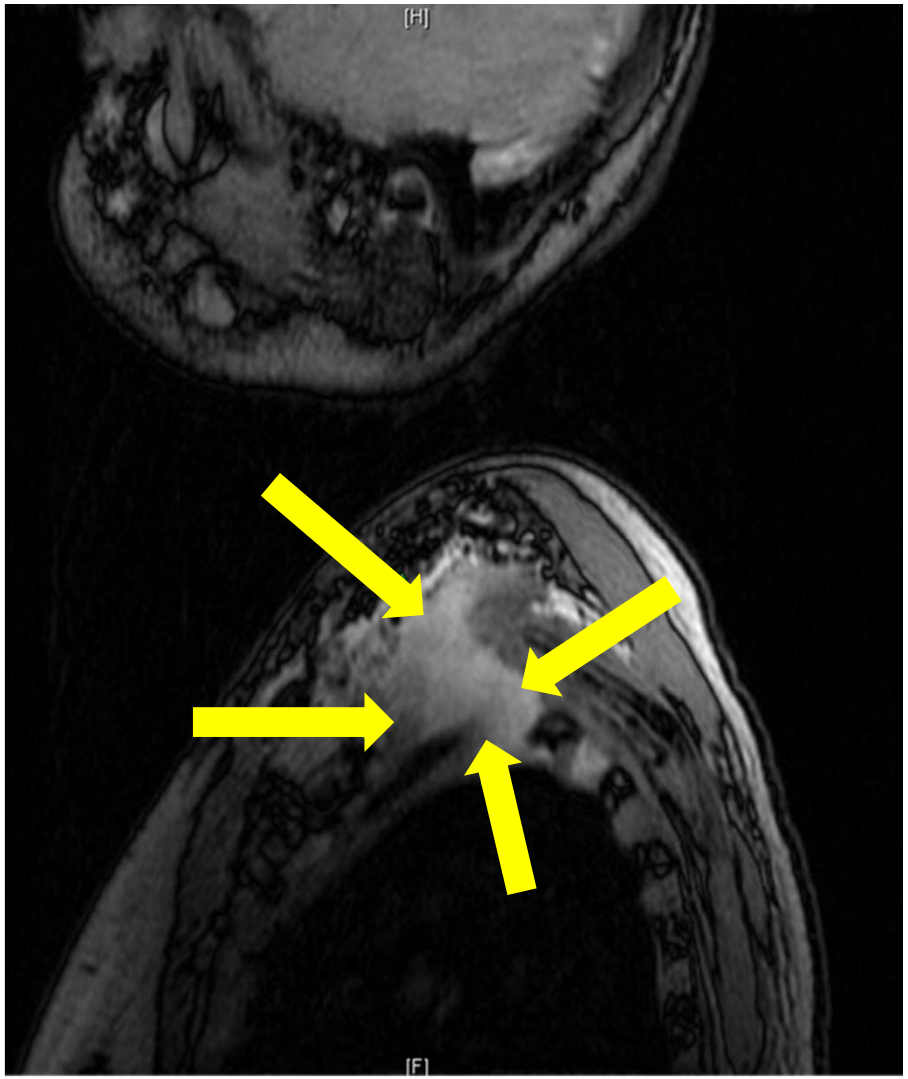
Coordination and Gait: No ataxia, normal gait.



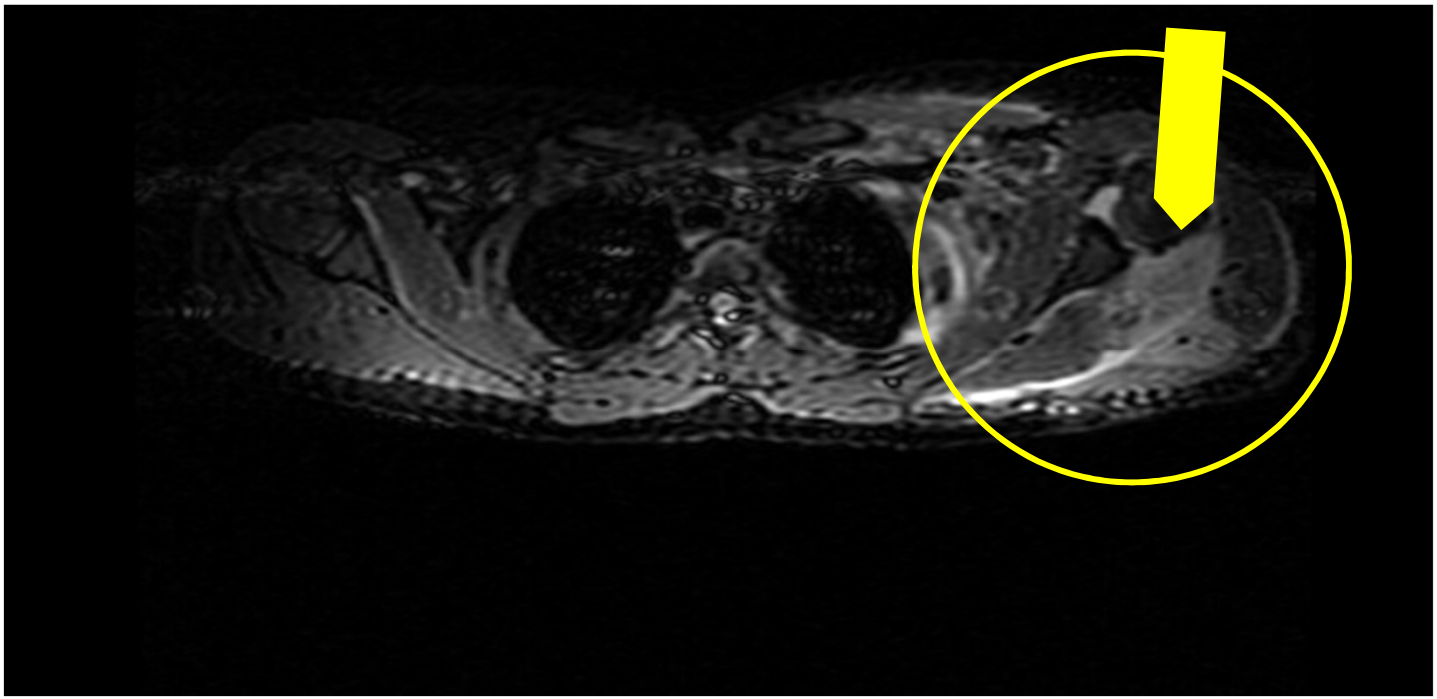


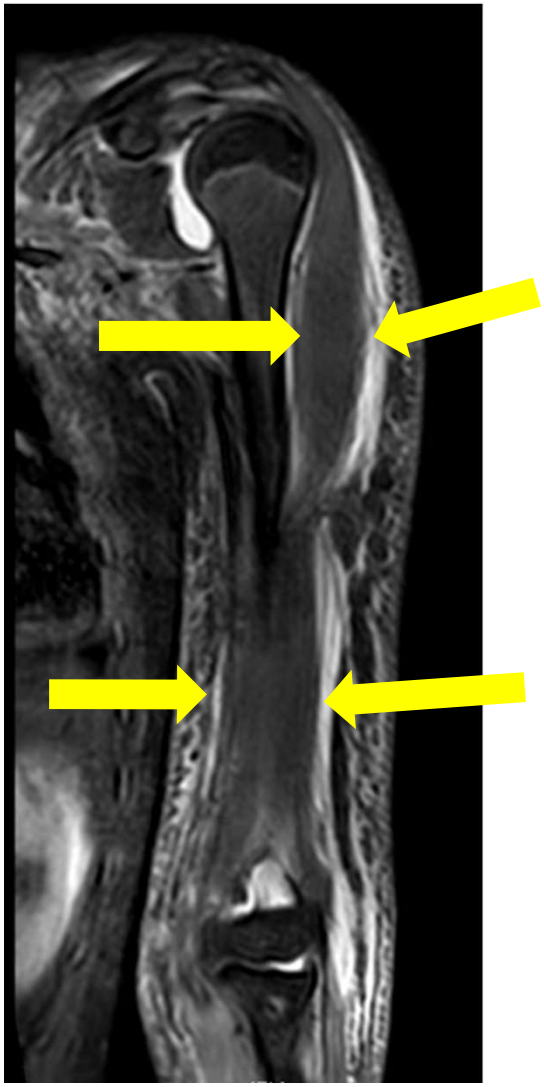
MRI (STIR Protocol) - Increased T2 signal in left lower neck and left superior thoracic cavity

Enhancing soft tissue mass within left inferior neck between scalene muscles, extending to lateral chest wall



Brachial Plexus MRI





Humerus MRI w/wo contrast showing edema and inflammation of the perimysium

Preliminary DDX

- Thoracic outlet syndrome
- Inflammatory disorder
- Infection i.e Lyme disease
- Malignancy
- Brachial neuritis
- Spider bite
- Epidural hematoma

Workup

	7/24	7/25	7/26	7/28
CK	**1606**		**651**	**195**
CRP		< 2.9		
ESR	7			
LDH			**411**	
AST	**86**	**72**		
Aldolase			**13.9**	
WBC	6.9			

Treated with clindamycin, vancomycin, ceftriaxone, lovenox, triamcinolone cream, and amitriptyline.

Consulted infectious disease, cardiology, rheumatology, dermatology, interventional radiology, and surgery.

EMG

Muscles showing increased insertional activity, fibrillations, and positive waves:

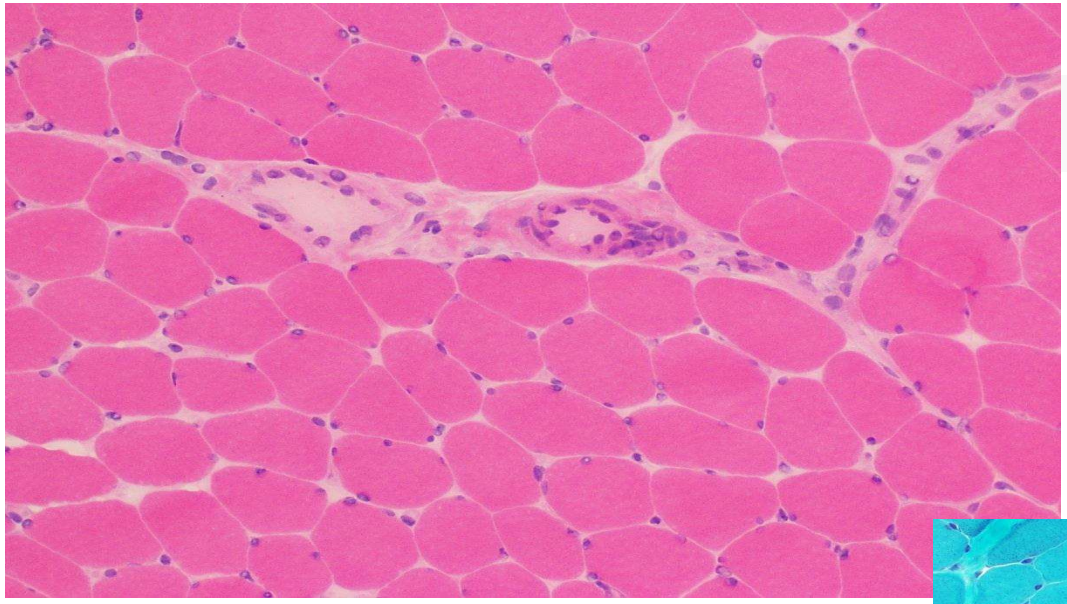
Flexor carpi ulnaris, abductor pollicis brevis, pronator teres, brachioradialis, extensor indicis, biceps, deltoids.

↓

Median, radial, ulnar, and axillary nerves + lateral/medial/posterior cords.
(No C5 nerve root involvement as rhomboid muscle was normal)

↓

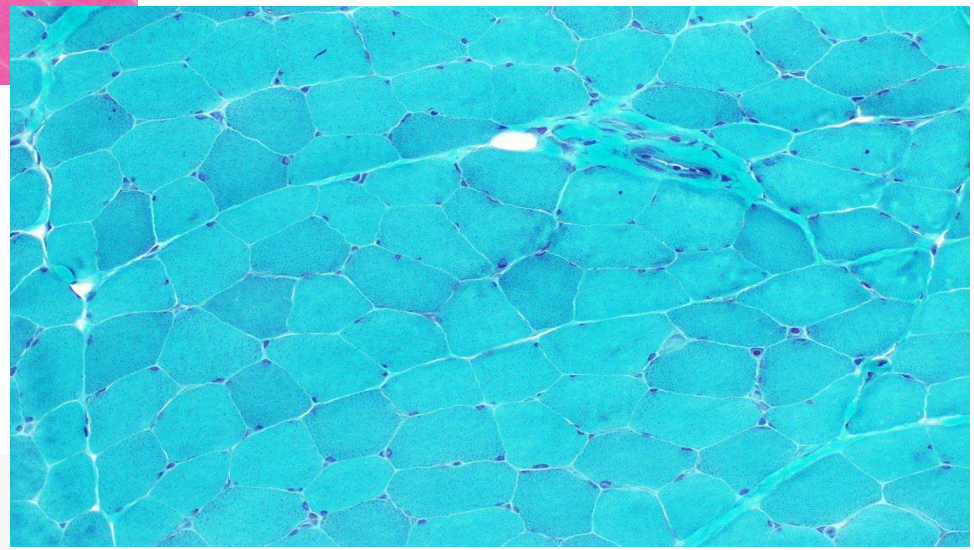
EMG suggestive of brachial plexus injury



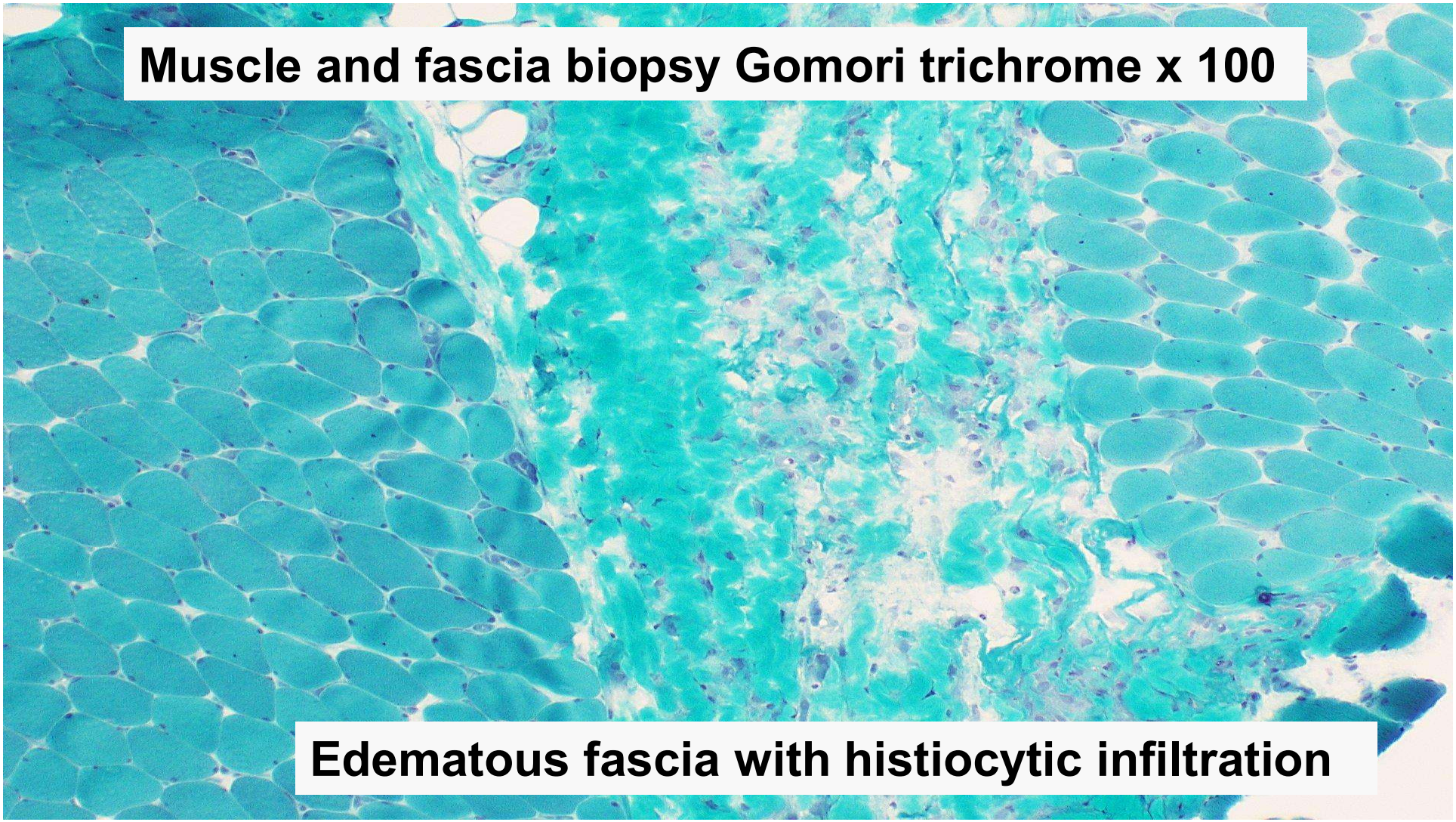
H&E x 100

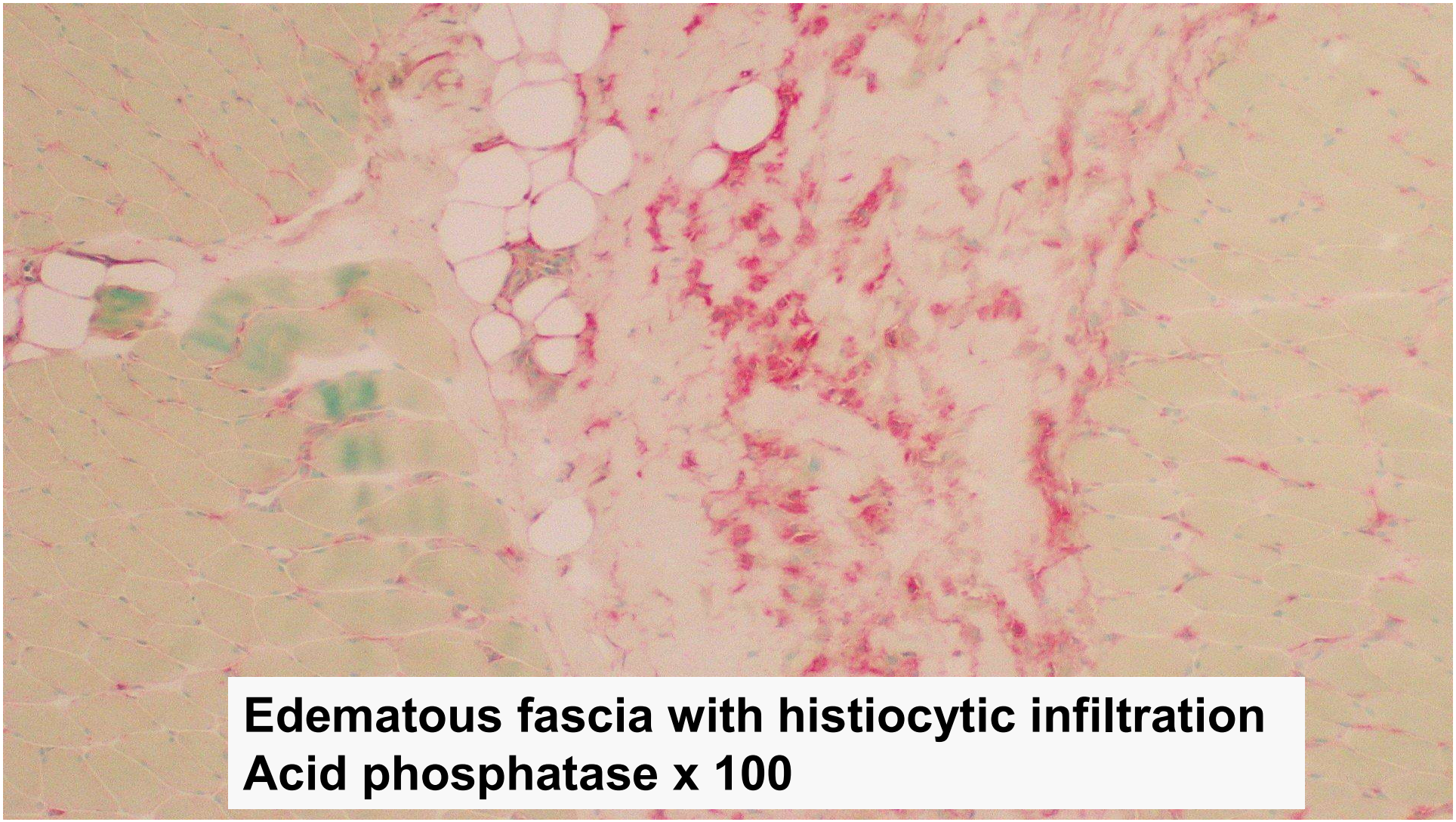
Histopathology

Left deltoid muscle

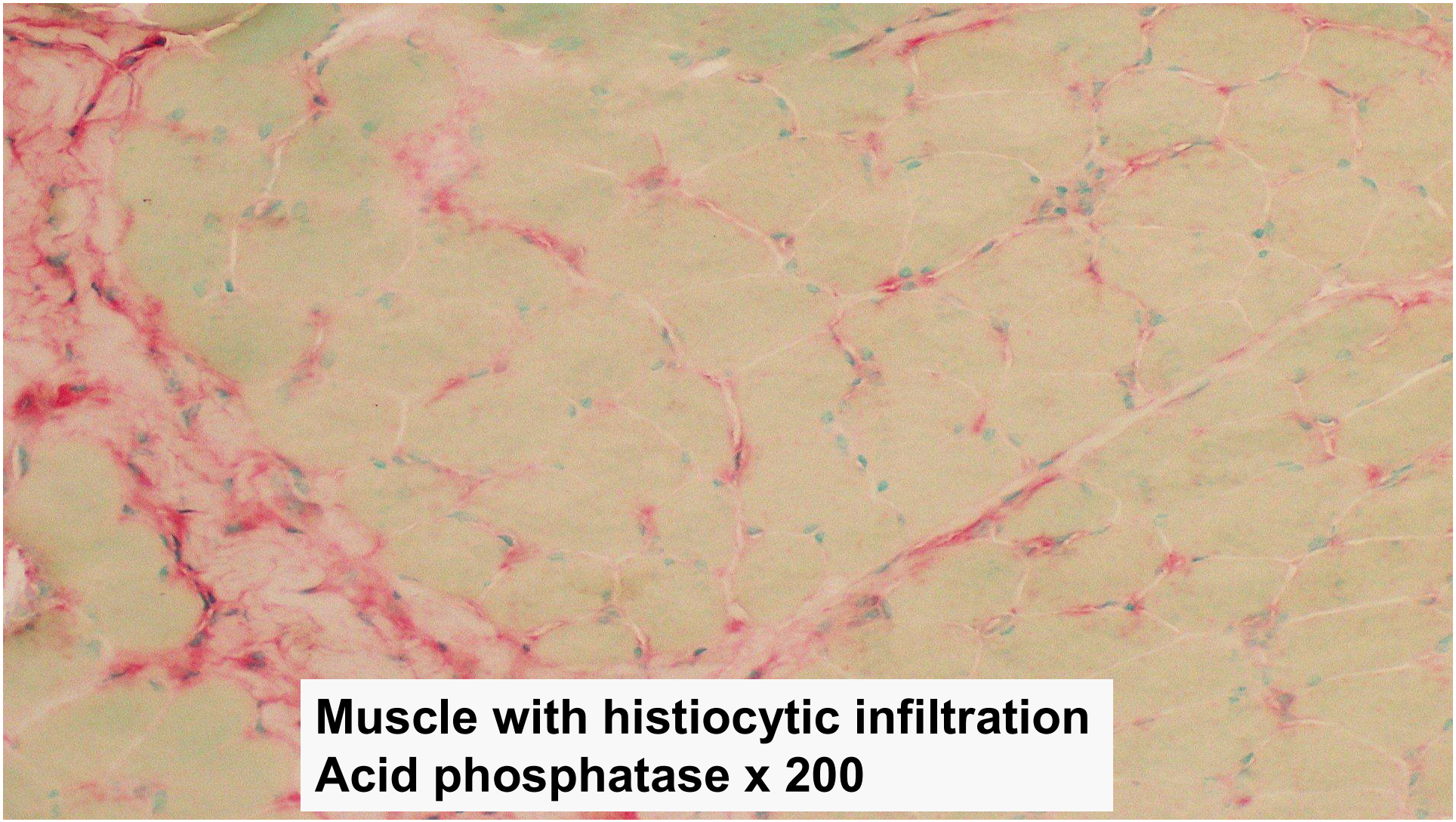


Gomori trichrome x 100

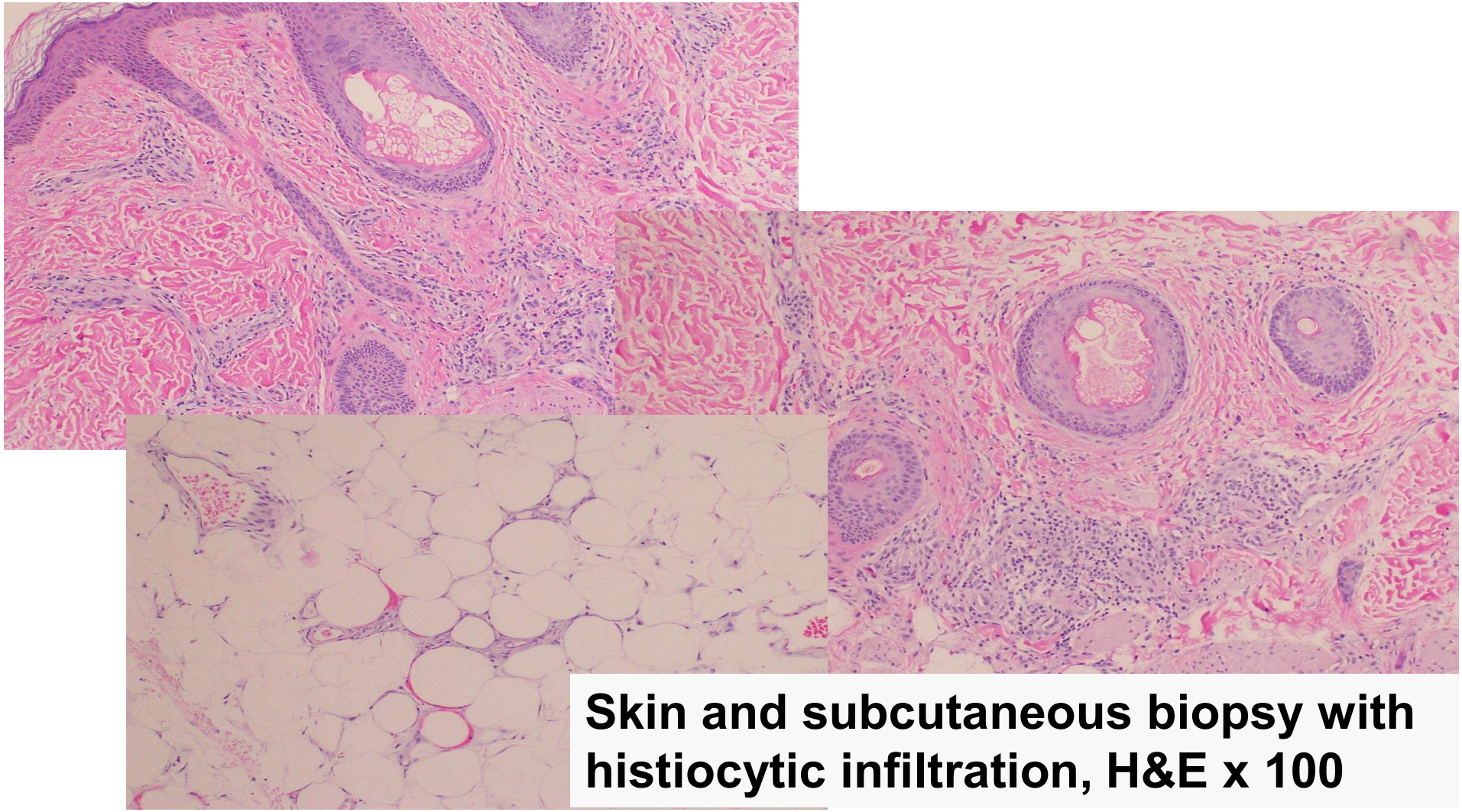

Muscle and fascia biopsy Gomori trichrome x 100
Edematous fascia with histiocytic infiltration



Edematous fascia with histiocytic infiltration
Acid phosphatase x 100



Muscle with histiocytic infiltration
Acid phosphatase x 200



Skin and subcutaneous biopsy with histiocytic infiltration, H&E x 100

Proposed Mechanism

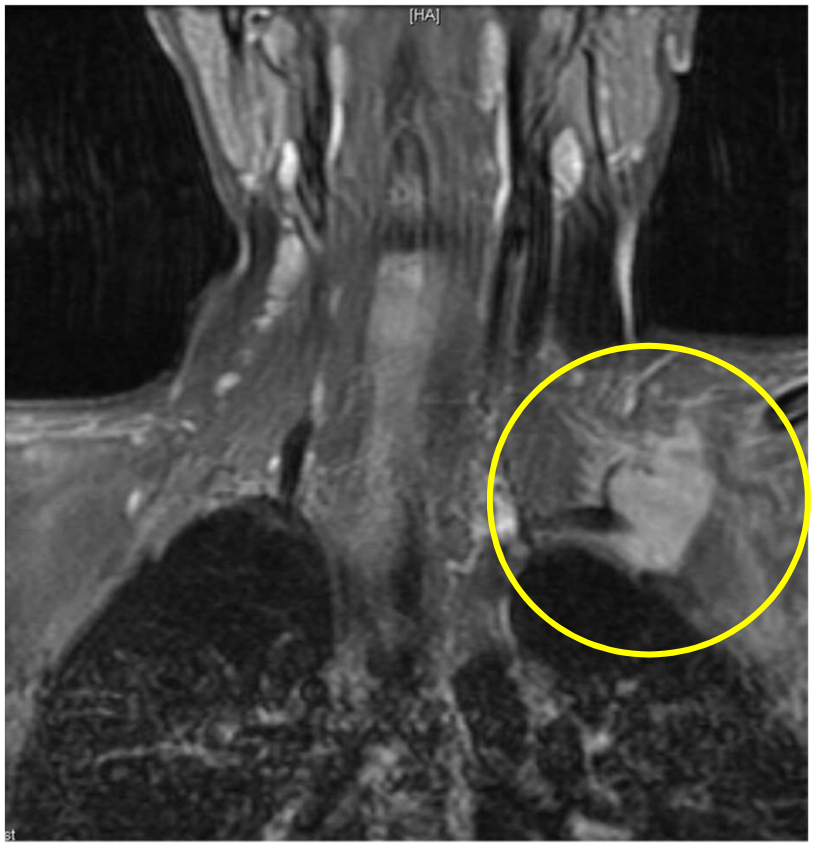
- Immune mediated allergic response
- Spider venom contains mucopolysaccharides, hyaluronidase, phospholipase, serotonin, histamine, and neurotoxins. Neurotoxins are the most important components of the venom. These low-molecular weight polypeptides cause severe adrenergic and cholinergic activities and affect sodium, potassium, and chloride channels of various cells.
- 1st - IL-1 and TNF-α mediated inflammation
- 2nd- TH1 cells secrete mainly INF, IL-2, and TNF-α. These cells are responsible for macrophage activation, TH2 induce and activate mast cells and eosinophils in the tissue

1: Ribeiro MF, Oliveira FL, Monteiro-Machado M, Cardoso PF, Guilarducci-Ferraz VV, Melo PA, Souza CM, Calil-Elias S. Pattern of inflammatory response to Loxosceles intermedia venom in distinct mouse strains: a key element to understand skin lesions and dermonecrosis by poisoning. Toxicon. 2015 Mar;96:10-23. doi: 10.1016/j.toxicon.2015.01.008. Epub 2015 Jan 16. PubMed PMID: 25600642.

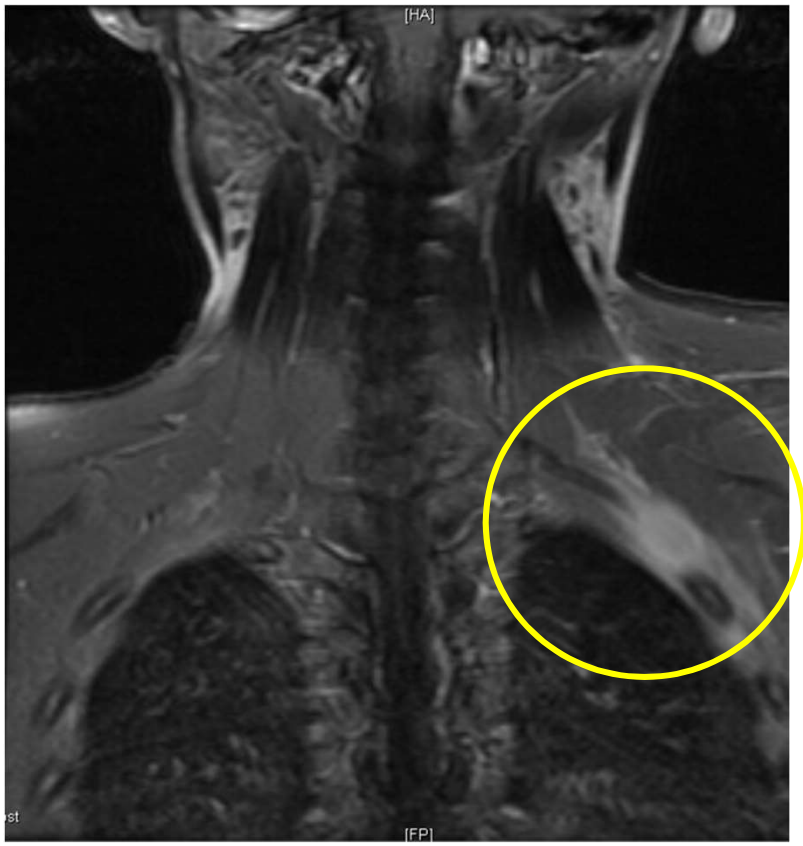
Discussion and Conclusion

- Spider venom → systemic inflammatory response →immune mediated reaction leading to brachial plexitis and focal myositis
- Follow up at 7 months shows 75% clinical improvement

MRI on admission



1 week later



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- Dr. Meenakshi Bhattacharjee (neuropathology)
- Dr. Rajan Patel (neuroradiology)

The End

Any Questions?

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Extra Slides

Motor Nerve Conduction:

Nerve and Site	Latency	Amplitude	Segment	Latency Difference	Distance	Conduction Velocity
Median.L						
Wrist	3.2 ms	2.1 mV	APB-Wrist	3.2 ms	mm	m/s
Elbow	6.8 ms	2.0 mV	Wrist-Elbow	3.6 ms	200 mm	55 m/s
Ulnar.L						
Wrist	2.3 ms	2.6 mV	ADM-Wrist	2.3 ms	mm	m/s
Below elbow	5.6 ms	1.9 mV	Wrist-Below elbow	3.3 ms	190 mm	58 m/s
Above elbow	8.2 ms	0.9 mV	Below elbow-Above elbow	2.6 ms	110 mm	43 m/s
Radial.L						
Forearm	2.1 ms	0.1 mV	Extensor indicis proprius-Forearm	2.1 ms	mm	m/s
Lateral brachium	NR ms	NR mV	Forearm-Lateral brachium	ms	mm	m/s

Sensory Nerve Conduction:

Nerve and Site	Onset Latency	Peak Latency	Amplitude	Segment	Latency Difference	Distance	Conduction Velocity
Median.L							
Wrist	2.2 ms	2.7 ms	40 µV	Digit II-Wrist	2.2 ms	130 mm	59 m/s
Elbow	2.2 ms	2.7 ms	39 µV	Wrist-Elbow	0.0 ms	mm	m/s
Ulnar.L							
	ms	ms	µV	Digit V-Elbow	1.8 ms	110 mm	62 m/s
Elbow	1.8 ms	2.7 ms	16 µV	Elbow-A3	0.2 ms	mm	m/s
	2.0 ms	2.7 ms	10 µV		ms	mm	m/s
Radial.L							
Forearm	1.6 ms	2.1 ms	15 µV	Snuff box-Forearm	1.6 ms	100 mm	48 m/s
	1.7 ms	2.1 ms	15 µV	Forearm-A2	0.1 ms	mm	m/s
Medial antebrachial cutaneous.L							
Elbow	1.3 ms	2.2 ms	6 µV	Forearm-Elbow	1.3 ms	120 mm	91 m/s
	1.4 ms	2.4 ms	6 µV	Elbow-A2	0.1 ms	mm	m/s
Lateral antebrachial cutaneous.L							
Elbow	2.0 ms	2.5 ms	9 µV	Forearm-Elbow	2.0 ms	120 mm	60 m/s
	1.9 ms	2.6 ms	10 µV	Elbow-A2	0.1 ms	mm	m/s

Needle EMG Examination:

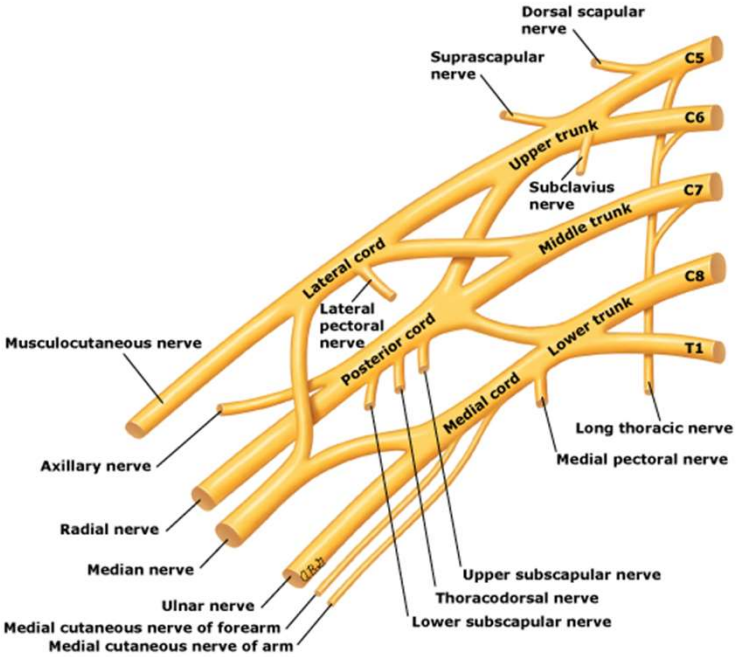
	Insertional	Spontaneous Activity			Volitional MUAPs					Max Volitional Activity		
Muscle	Insertional	Fibs	+Wave	Fasc	Duration	Amplitude	Poly	Config	Recruitment	Amplitude	Pattern	Effort
First dorsal interosseous.L	Normal	None	None	None	Normal	Normal	None	Normal	Reduced	800MV	Reduced	
Flexor carpi ulnaris.L	Increased	2+	2+	None								
Abductor pollicis brevis.L	Increased	1+	3+	None	Normal	Normal	None	Normal	Reduced	UP TO 600 MV	Reduced	
Pronator teres.L	Increased	2+	3+	None	Normal	Normal	None	Normal	Reduced	FF/600MV	Single unit	
Brachioradialis.L	Increased	None	None	None								
Triceps brachii.L	Normal	None	None	None								
Extensor indicis proprius.L	Increased	None	None	None								
Pronator quadratus.L	Normal	None	None	None								
Biceps brachii.L	CRD	None	3+	None								
Deltoid.L	Increased	None	3+	None								
Rhomboid major.L	Normal	None	None	None	Normal	Normal	None	Normal	Normal			
Supraspinatus.L	Normal	None	None	None	Normal	Normal	None	Normal	Reduced	400MV	Reduced	
C7 paraspinal.L	Normal	None	None	None								

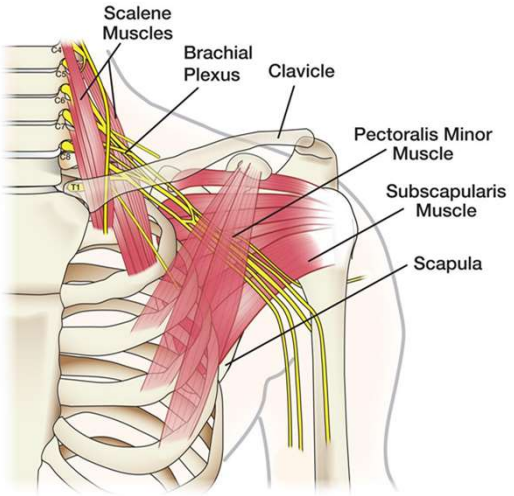
F-Wave Studies

Nerve	M-Latency	F-Latency
Median.L	6.8	23.1
Ulnar.L	2.3	28.1

Brachial Neuritis

Brachial Plexus Anatomy





https://www.uptodate.com/contents/images/NEURO/50576/Anatomy_of_brachial_plexus.jpg?title=Trunks+and+cords+of+the+brachial+plexus
http://www.assh.org/handcare/hand-arm-injuries/Brachial-Plexus-Injury

Brachial neuritis: Background

- Inflammatory change involving the brachial plexus
- Affects mainly the lower motor neurons of the brachial plexus, individual nerves, or nerve branches
- Characterized by acute onset of excruciating unilateral shoulder pain, followed by **flaccid paralysis and weakness of the shoulder and arm muscles**

Brachial neuritis: Etiology

- Traumatic
- Neoplastic and radiation induced plexopathy
- Viral
- Vaccinations
- Systemic
- Immunologic
- Hereditary
- **Idiopathic**

Brachial neuritis: Pathophysiology

- Brachial neuritis exists in an inherited and an idiopathic form
- In the idiopathic form, the pathophysiology is unknown, but is thought to be an immune system mediated inflammatory reaction against nerve fibers of the brachial plexus
  - Herpes simplex, varicella zoster virus[9]
  - Altered lymphocyte subsets[10]
- The inherited form is autosomal dominant and has been linked to mutations in the SEPT9 gene on chromosome 17q25[11]
  - Septins involved in the formation of the cytoskeleton and cell division

Brachial neuritis: Epidemiology

- Brachial plexitis has been reported in individuals from age 3 months to 74 years, most commonly presenting in the 3rd-7th decades of life
- Prevalence is highest in men (M:F 2:1 to 11.5:1)
- Incidence of brachial plexitis 1.6 cases per 100,000 person-years in the United States[12]

Brachial neuritis: Presentation

- Abrupt onset of pain at shoulder and upper arm
- Pain may be bilateral in 10-30% of patients
- A minority of patients do not experience the initial painful stage
- Pain intensity is usually very high, lasting from a few hours to several weeks
- As the pain subsides, weakness becomes apparent.
- Affects a wide variety of muscles, usually those innervated by upper trunk (supraspinatus, infraspinatus, deltoid, biceps, serratus anterior)
- Atrophy and reduced muscle strength of affected muscles become prominent after approximately 2 weeks
- Can also present with numbness, reduced reflexes, and sensory loss
- Course of the neuritis is usually gradual improvement and recovery of muscle strength in 3-4 months, although some patients may have several years of muscle weakness or slight permanent weakness

DDx of brachial neuritis

- Cervical spondylosis
- Cervical radiculopathy
- Suprascapular notch masses
- Rotator cuff deficiency
- Shoulder impingement
- Calcific tendonitis
- Adhesive capsulitis
- Disuse atrophy of shoulder girdle muscles
- Quadrilateral space syndrome
- Trauma
- Acute poliomyelitis
- Cervical disc disease
- Mononeuritis multiplex
- Neoplastic brachial plexopathy
- Pediatric HIV infection
- Polymyalgia rheumatica
- Thoracic outlet syndrome

Brachial neuritis: Workup

- Laboratory studies usually **within the reference range**
- Electrodiagnostic testing (EMG, NCS)
  - Show abnormalities c/w brachial plexus lesions (**axonal loss**)
  - **Fibrillation potentials** and **positive waves** suggestive of **muscle denervation**
- MRI
  - **Denervation changes in muscles**
  - Muscle initially appears normal
  - Over the next few weeks, **high T2 signal develops**
  - Gradually, atrophy and fatty infiltration will develop with the increase in T1 signal and decreased muscle bulk
- Ultrasound

BN: Treatment and Management

- No specific treatment
- Typically **self-limiting**, requiring only supportive therapy and conservative management
- Majority of patients have a slow but steady recovery of motor function over 6-18 months
- 90% of patients have excellent recovery by 3 years
- **Physical therapy** to maintain strength and mobility
- **Occupational therapy** for functional conditioning of the upper extremity
- Medical pain management: corticosteroids?[13]
- Surgical intervention: nerve grafting or tendon transfers

Evaluation of prednisolone treatment in the acute phase of neuralgic amyotrophy: an observational study. *van Eijk JJ, van Alfen N, Berrevoets M, van der Wilt GJ, Pillen S, van Engelen BG. J Neurol Neurosurg Psychiatry. 2009 Oct; 80(10):1120-4.*

Focal Myositis

- Rare and benign dysimmune disease that presents as isolated intramuscular mass, or pseudotumor.
- First described in 1977 by Heffner et al.[7]

"... myopathy affecting a single skeletal muscle without systemic manifestation with a histologically proven inflammatory myositis process" - Flaisler

Etiology Unknown

Proposed mechanisms:

1. Neurologic: denervation in setting of genetic susceptibility, radiculopathy, or **nerve lesion**.
2. Immune: triggered by dysimmune/autoimmune diseases i.e. Behcet.[1]

Other causes:

1. Trauma/mechanical lesions
2. Infection: associated with Lyme, coxsackie, TB, influenza, etc. Suspect if fever, diarrhea, or **skin lesion present**.
3. Iatrogenic: associated with statin use.[2]

FM: Epidemiology

- Affects any age, preferably middle-aged adults.
- Affects males and females equally.
- Only 200 cases reported as of 11/2016.[3]

FM: Clinical Presentation

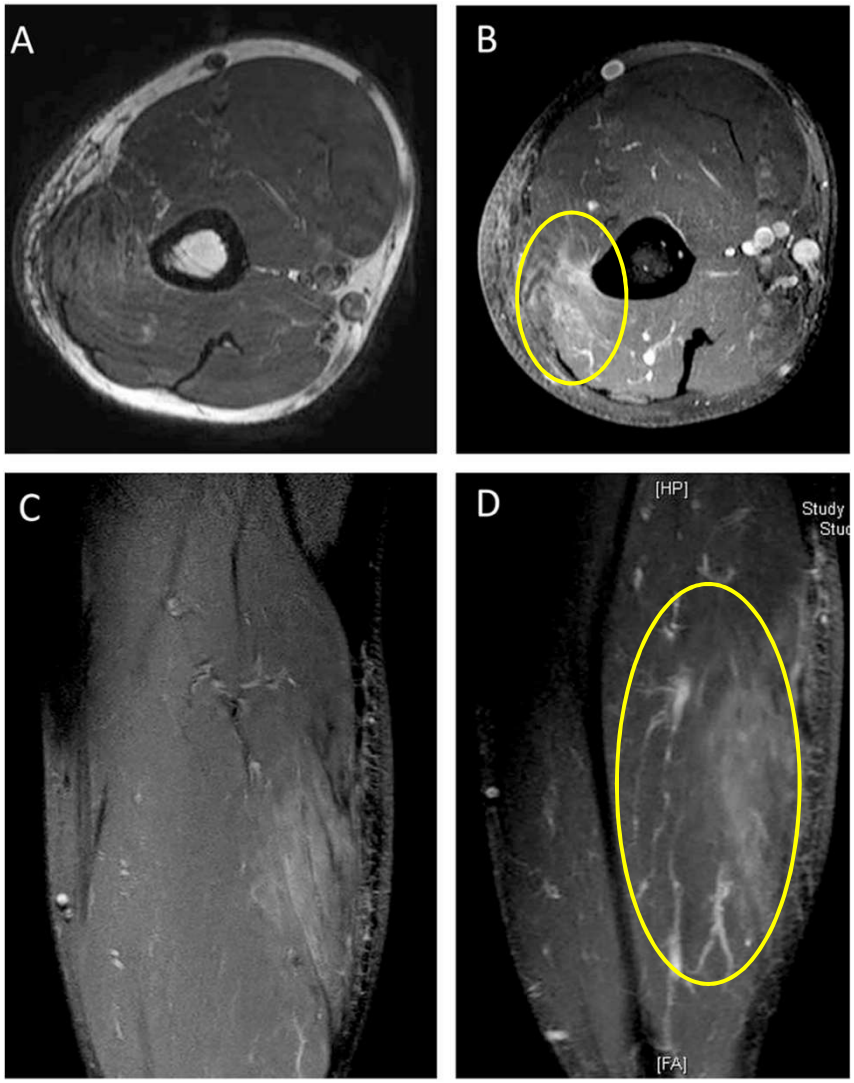
- **Intramuscular mass or swelling** within one muscle. Grows slowly. Can be **painful**.
- Usually in lower extremities
- **No systemic symptoms** usually.
- No weakness or sensory impairments if no nerve association.
- No recurrence in 2-6 vears.

FM: Differential Diagnoses

- Benign tumors: similar MRI as FM
- Malignant tumors: invade adjacent structures.
- Myositis ossificans: calcification of a muscle.
- Proliferative myositis: presence of basophilic fibroblasts.
- Polymyositis: symmetric involvement of proximal muscles.
- Muscular dystrophy: assess clinical history.

FM: Workup

- CK and ESR (usually moderately elevated), CRP normal
- Order MRI
- Order EMG
- Confirm with muscle biopsy



MRI Triceps

FM: EMG

Exclusively in affected muscle

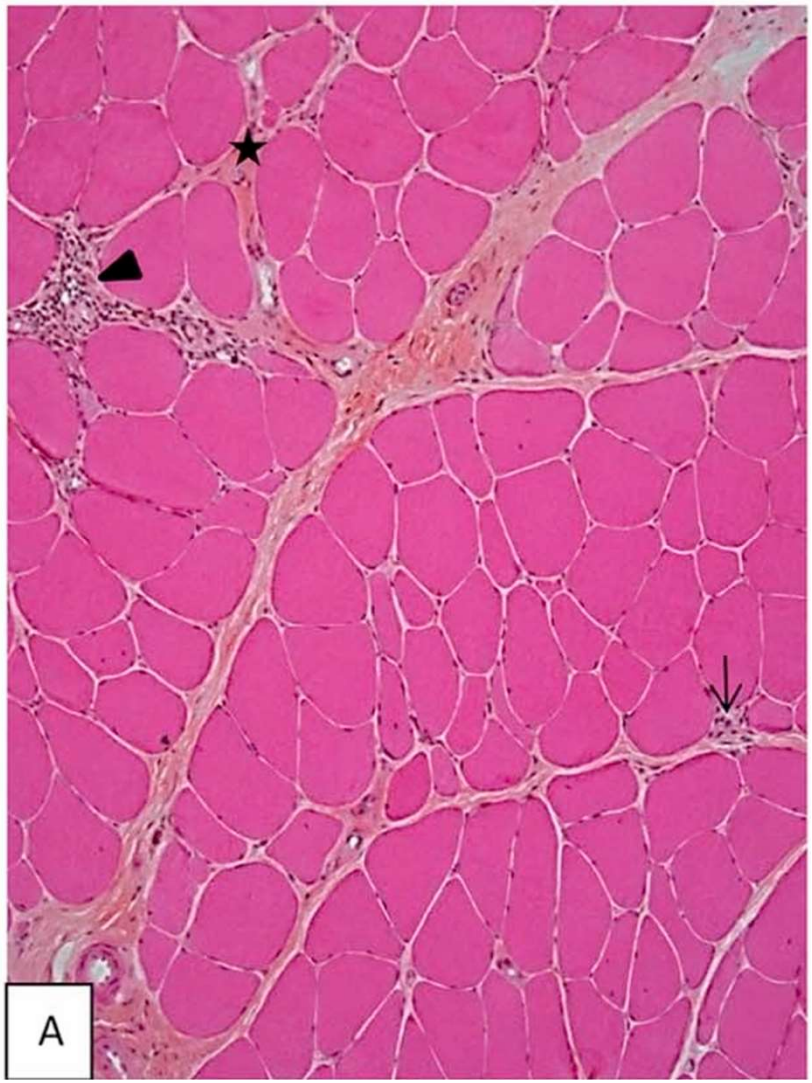
Spontaneous activity associated with positive sharp wave or fibrillation

Small amplitude polyphasic motor unit potentials

http://www.iomonitoring.org/imgemg/Fig-5.jpg

FM: Muscle Biopsy

- Macrophage (**arrow**) and inflammatory cell infiltration (**arrow head**).[3]
- Hypertrophic muscle fiber regeneration.
- Interstitial fibrosis (**star**).



FM: Treatment

Self-limited. Usually regresses within one-two weeks.

Relapse occurs in 1-18% of cases.[4]

Steroids recommended IF associated with elevated CK/ESR, nerve lesion, or autoimmune process.

- 0.75 mg/kg/day for 4–12 weeks