34 yo M with HIV, DM 2, hyperlipidemia and acute myopathy

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Disclosures

Relevant financial relationships

None

Off-label/investigational uses

None

HPI

- 34 yo M, subacute severe shoulders pain, moved to neck
- Unable to raise arms all the way due to pain/weakness.
- Cannot pull on pants because of difficulty lifting leg
- Without Norco unable to walk.
- No breathing difficulties, No dysphagia.

Medications:

- **ARVs: Genvoya** (elvitegravir, cobicistat, emtricitabine, tenofovir)
  Biktarvy (bictegravir, emtricitabine & tenofovir)
- **WAS taking Lipitor, stopped**
- **Norco 1 pill every 6 hours.**
- **Naproxen twice a day**
- **Steroids 60 mg/day**

FHx:

- **Sister: diagnosed with Lupus.**
- **Diabetes runs in family.**

Exam

- **Motor exam limited by pain**
- **Could raise arms above shoulders with difficulty (at least 4/5 throughout proximally and distally)**
- **Could squat with difficulty (holding to objects)**
- **Calf is tender with some swelling**
- **Diffuse swelling of various muscles that is visible on lower back paraspinal, upper arms, forearm, upper thigh (lateral and medial side)**

Labs

	1/5/19	1/6/19	1/22/19	1/25/19	
CK	**4687**	**4290**	**8764**	**9162**	**8193**
Aldolase					**33**

Labs-Cont

	10/2017	11/2017	12/2017	4/2018	1/2019
HbA1C	13.3	16.2	12.3	6.8	10.1
LDL			124		
TRIG		1275	217		

Labs-Cont

MyoMarker Panel 3: Negative

(SRP, J0-1 and other AyS Abs)
HMGCR Ab: Negative

Labs-Cont

	3 1/22/2019 1858	2 1/25/2019 1542	1 1/30/2019 1302
SEROLOGY (NON-MIC...			
ANA AB, IF			*<1:80*
ANA INTERP			*See Note **
ANA, SER QL		*POSITIVE **	
JO-1 AB ID	*<0.2*		
SCL-70 AB	*<0.2*		
DNA DS AB	*1 **		
SS-A AB	*<0.2*		
SS-B AB	*<0.2*		
RNP AB	*0.2*		
SMITH AB	*<0.2*		
CENTROMERE AB, SER QN	*1.4*		
ENA SM+ RNP AB, SER	*<0.2*		
CHROMATIN (NUCLEOS...	*<0.2*		
RPP	*<0.2*		

Labs-Cont

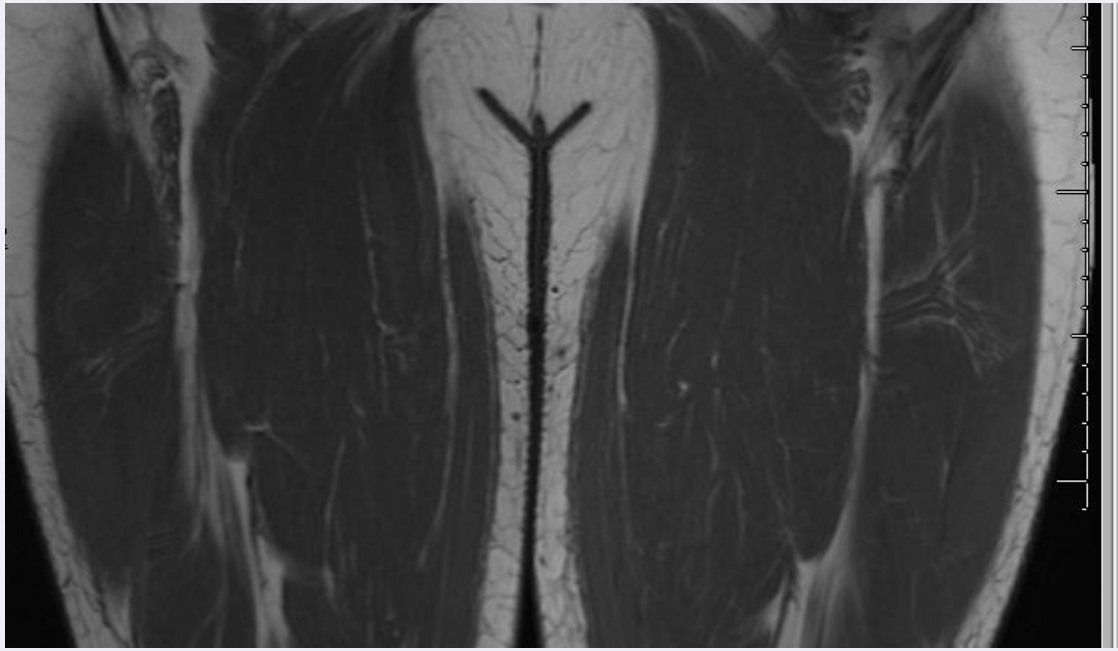
	... 10 9/5/2013 1122	9 4/23/2014 1128	8 1/7/2015 1810	7 11/14/2015 1020	6 2/17/2017 1305	5 10/1/2017 1542	4 12/2/2017 1010	3 4/24/2018 1855	2 1/5/2019 1215	1 1/25/2019 1542
OTHER -										
CD3 CELLS, CT QN	2,315 *	1,908 *	2,110	1,712	1,457 *	1,747	1,245	2,165	840	
CD4 CELLS % QN	20 *	23 *	23	19	21 *	22	24	22	25	
CD4 CT	593 *	600 *	701	436	410 *	553	383	679	301	
CD4/CD8 CELLS, NUM QN	0.38 *	0.50 *	0.54 *	0.35 *	0.42 *	0.50 *	0.47 *	0.49 *	0.63 *	
CD8 CELLS % QN	54 *	47 *	43	54	49 *	45	51	46	40 *	
CD8 CELLS, CT QN	1,579 *	1,204 *	1,287	1,238	981 *	1,103	819	1,396	478 *	
ESR									27 *	49

LE Doppler:

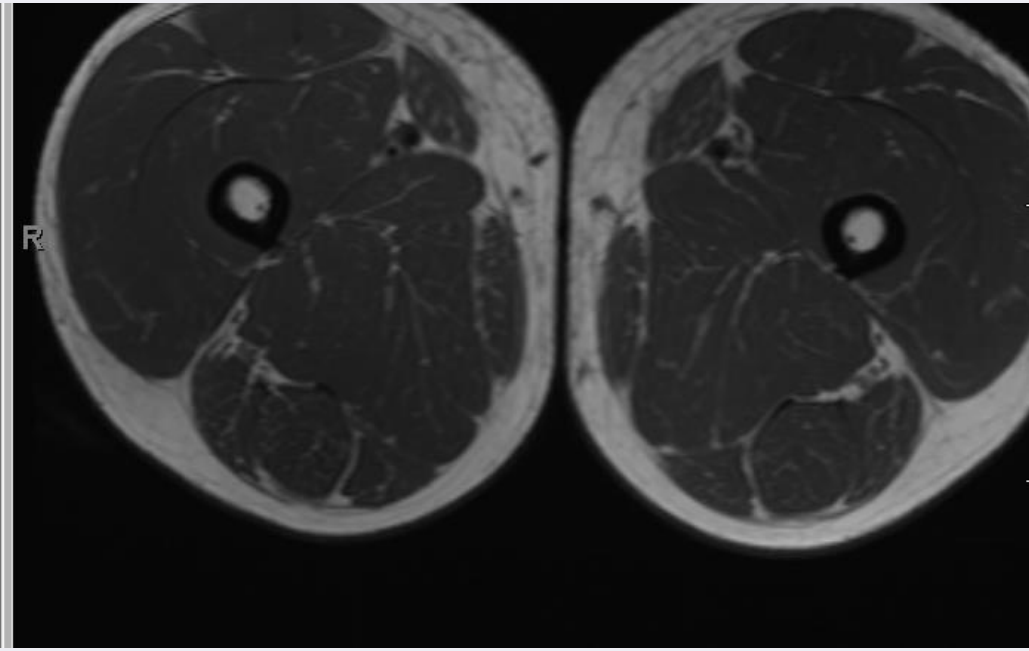
No deep vein thrombus in the left upper extremity.

Muscle MRI

T1

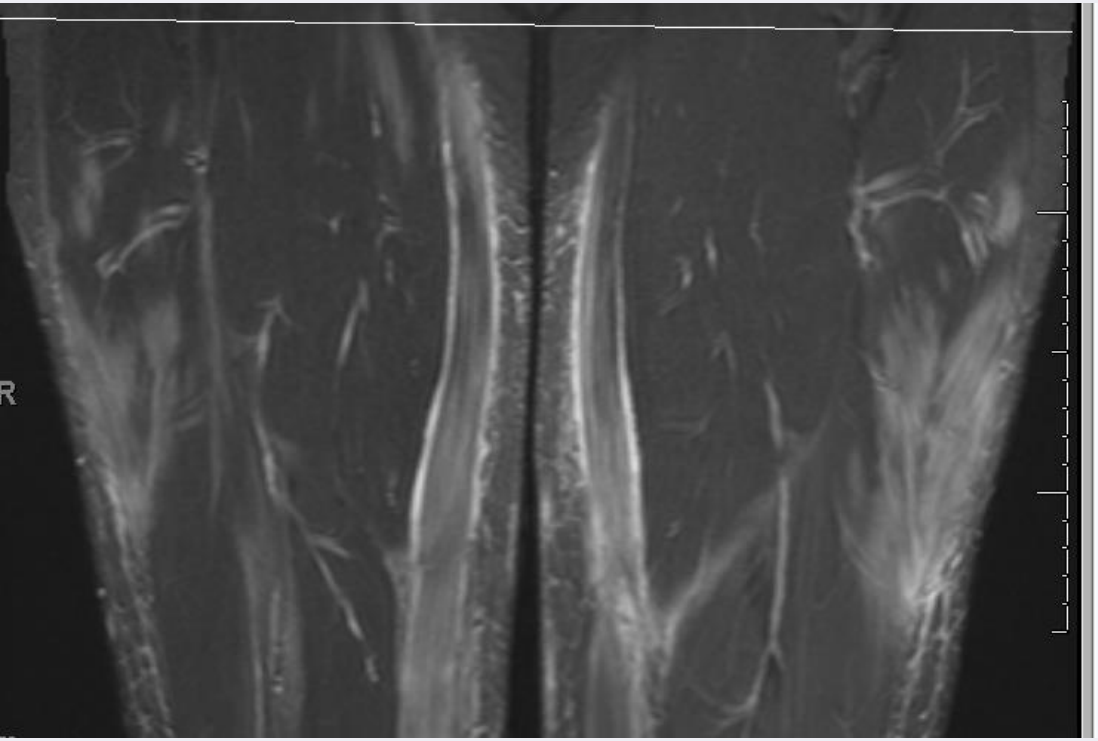


T1

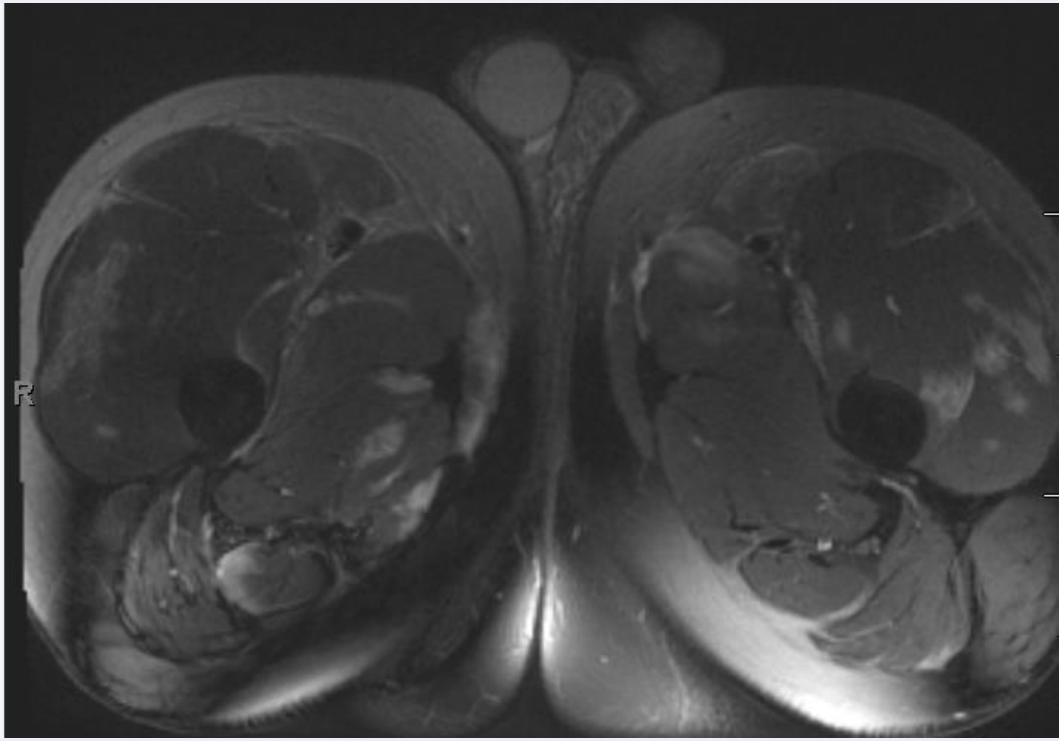


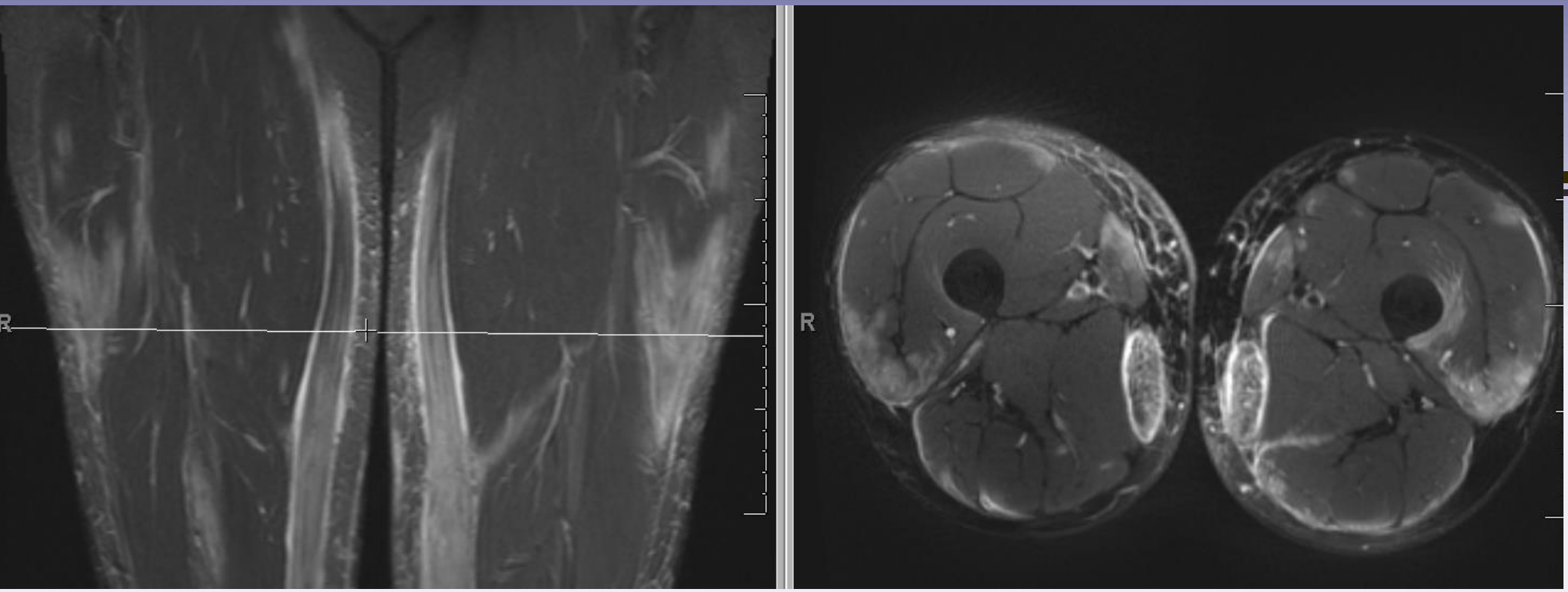
Muscle MRI

STIR

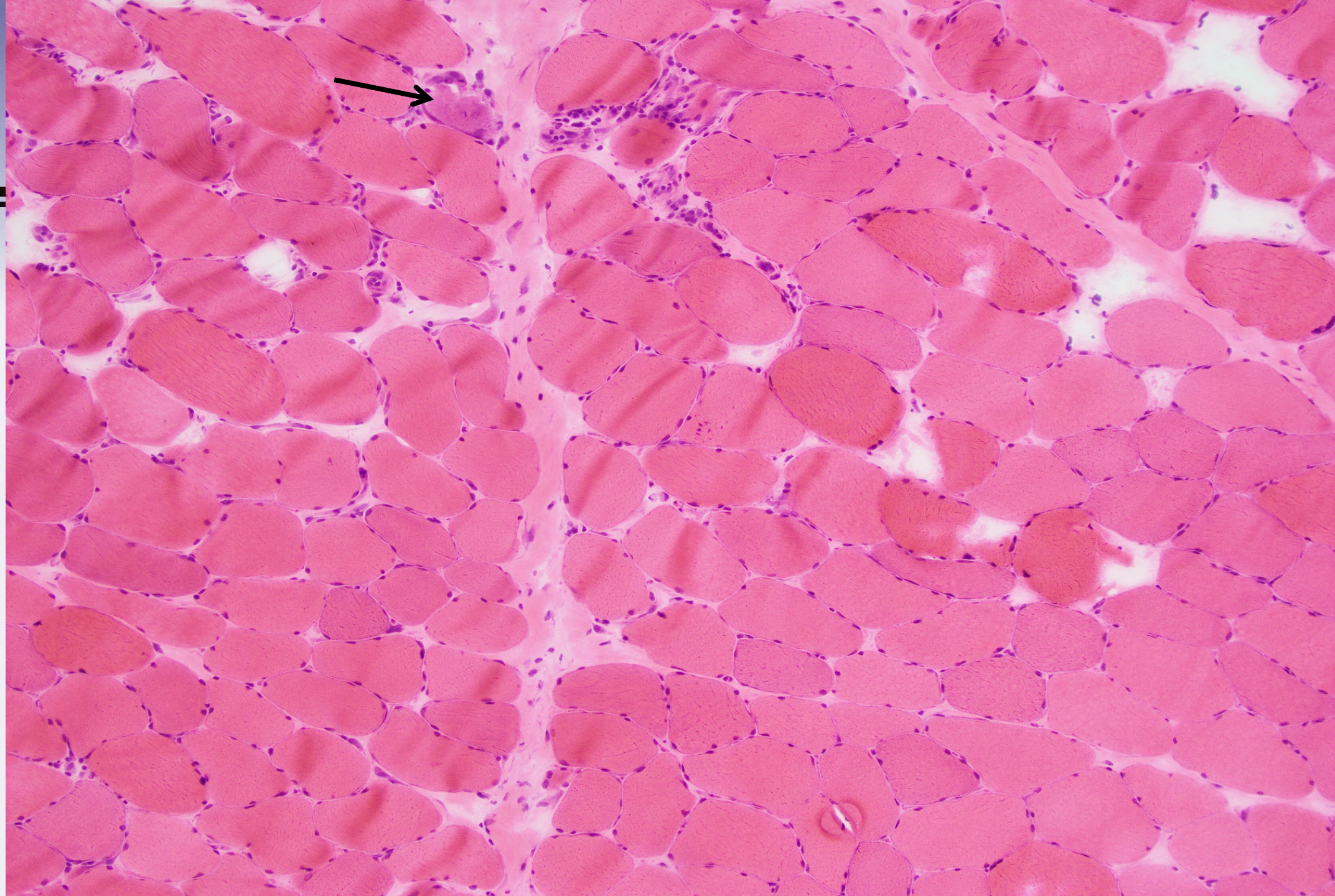


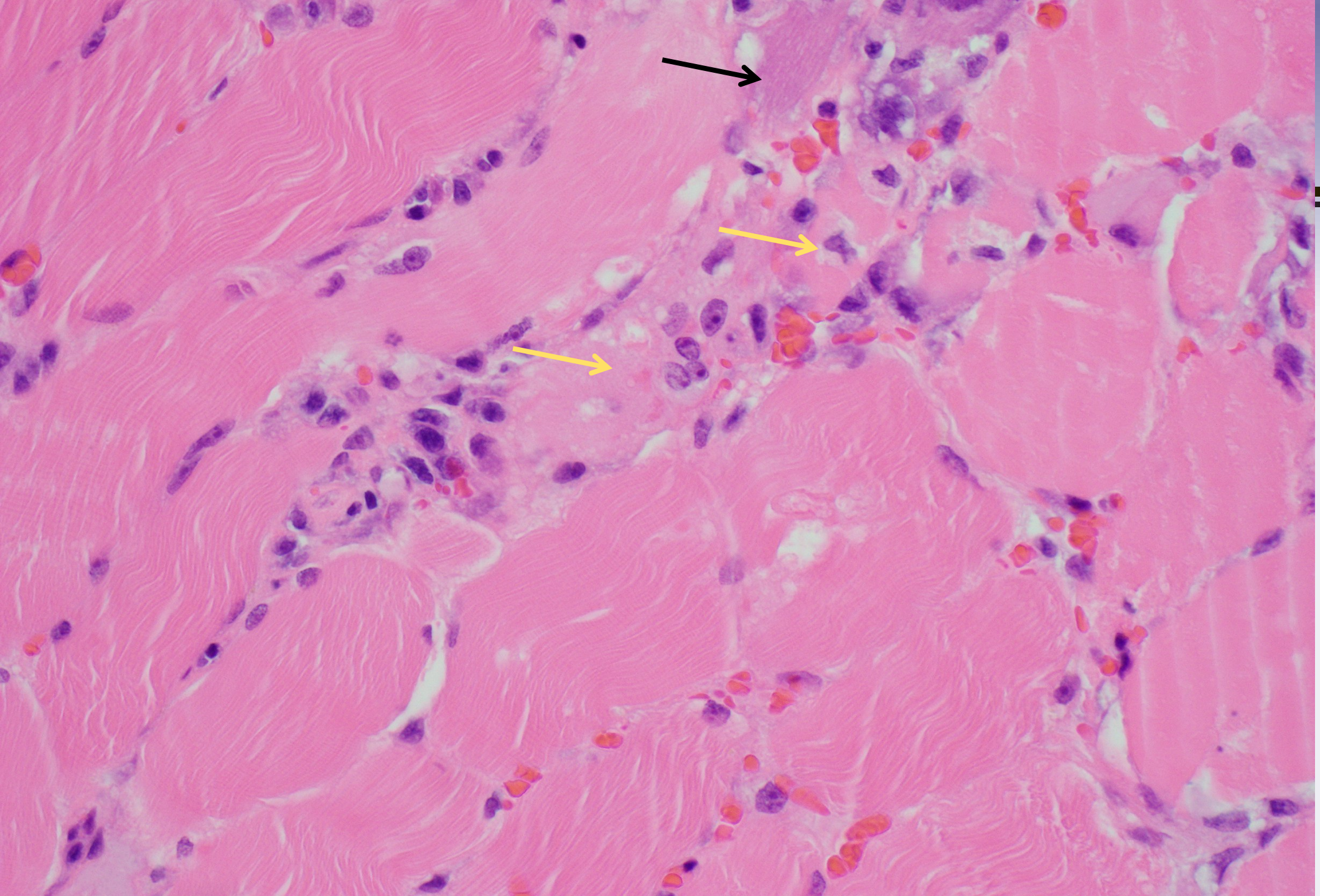
T2 Fat Sat

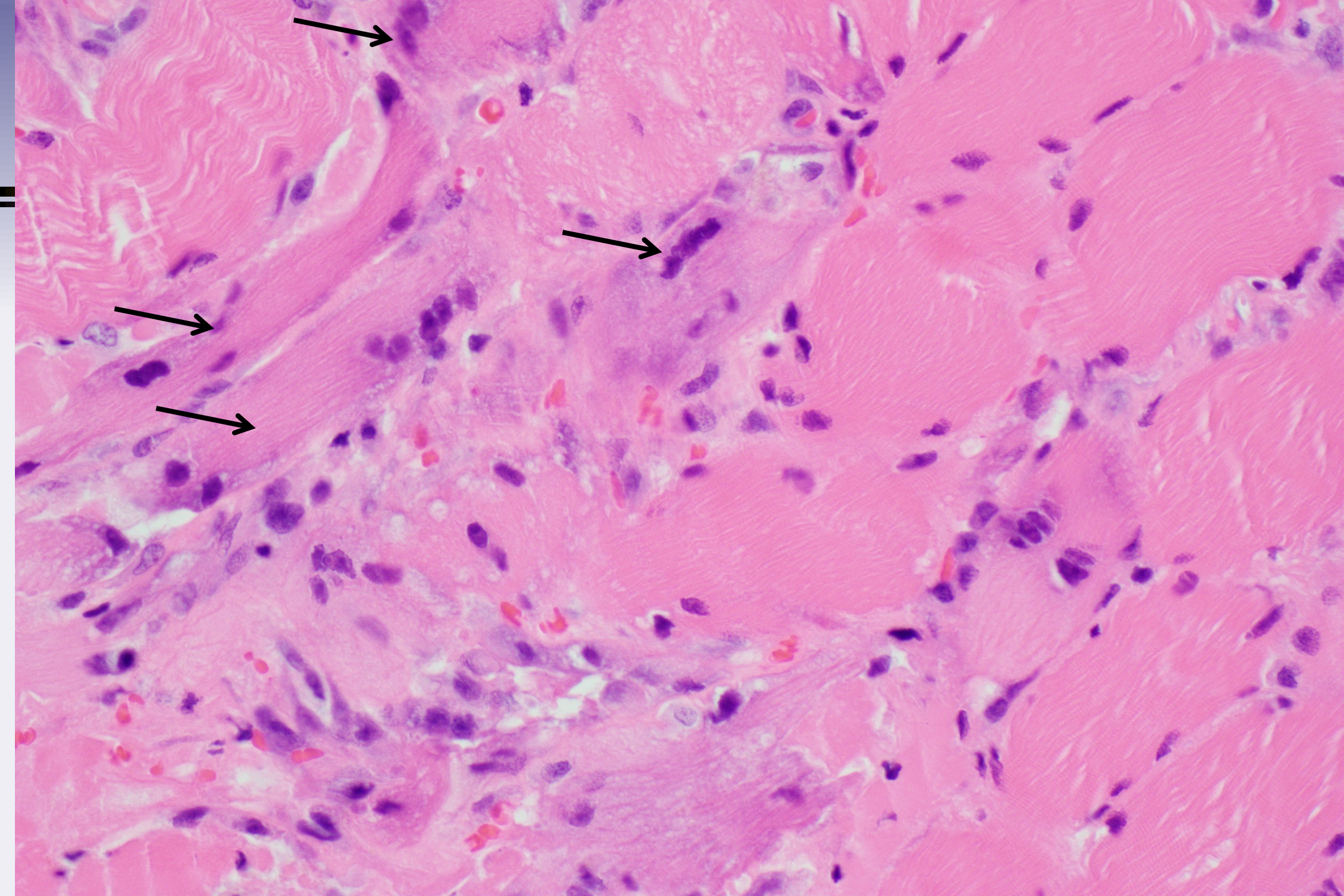


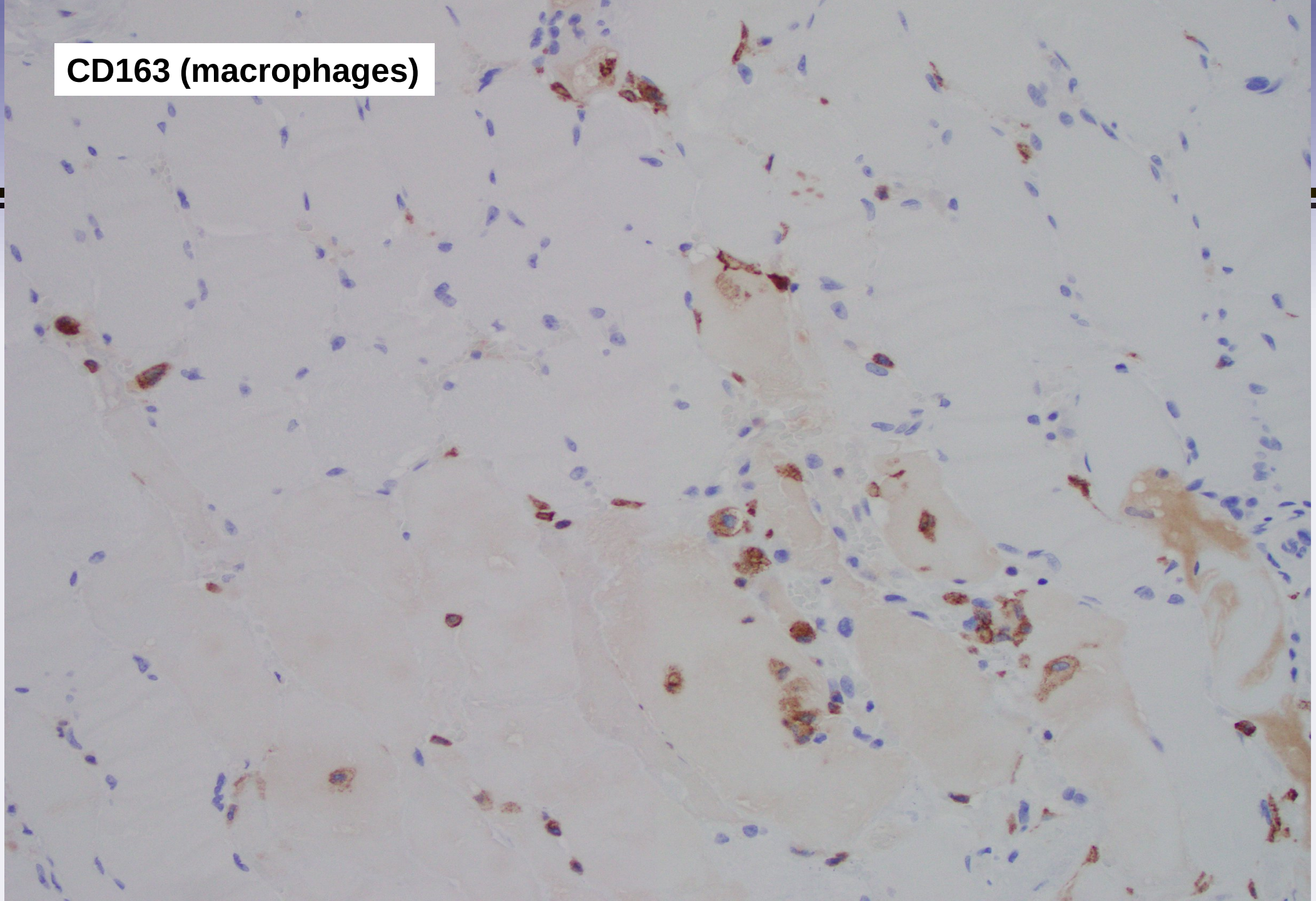


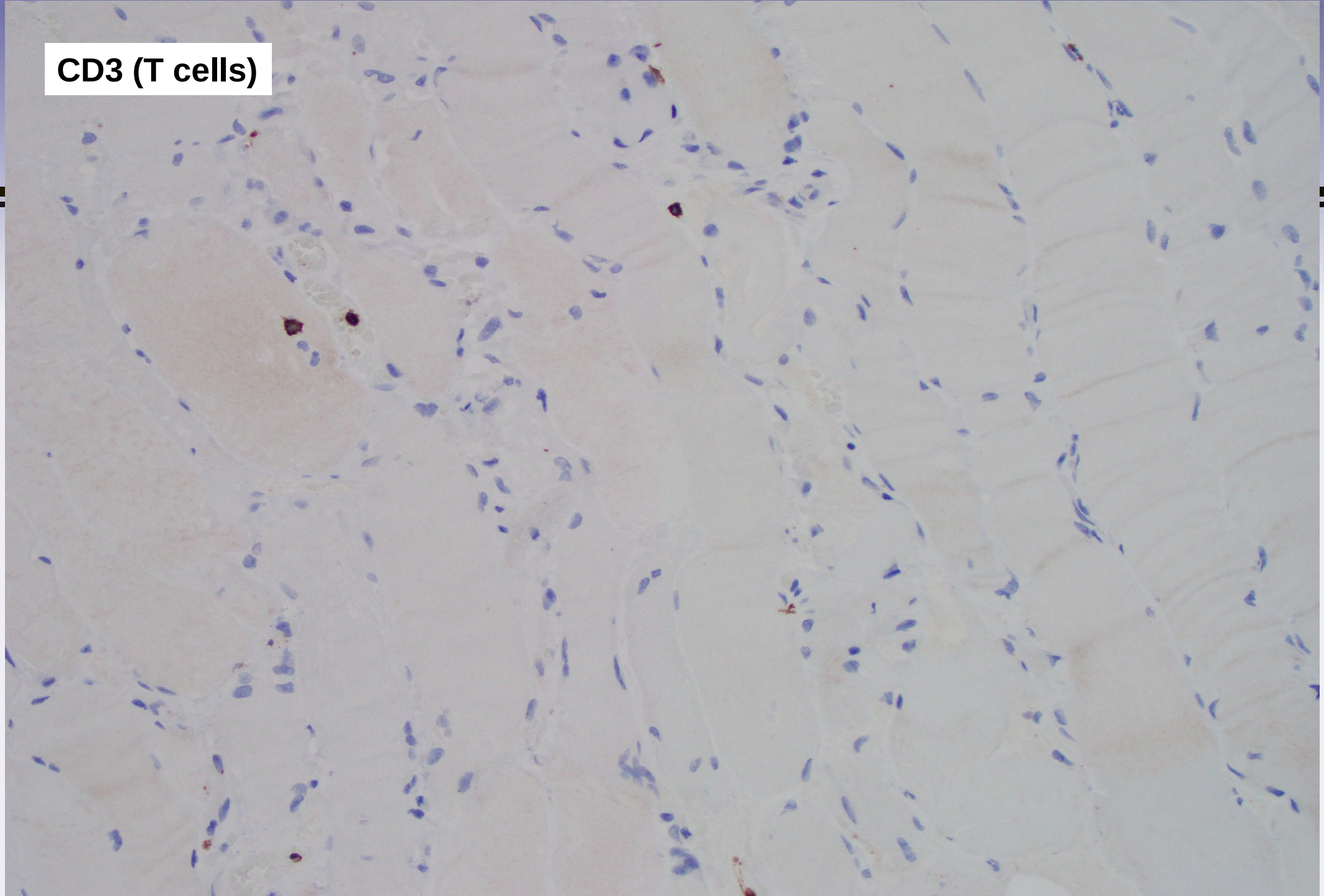
"Multifocal intramuscular edema in the pelvic and bilateral thigh musculature, most notably involving the bilateral adductor and gracilis muscles as described **in keeping with myositis**"

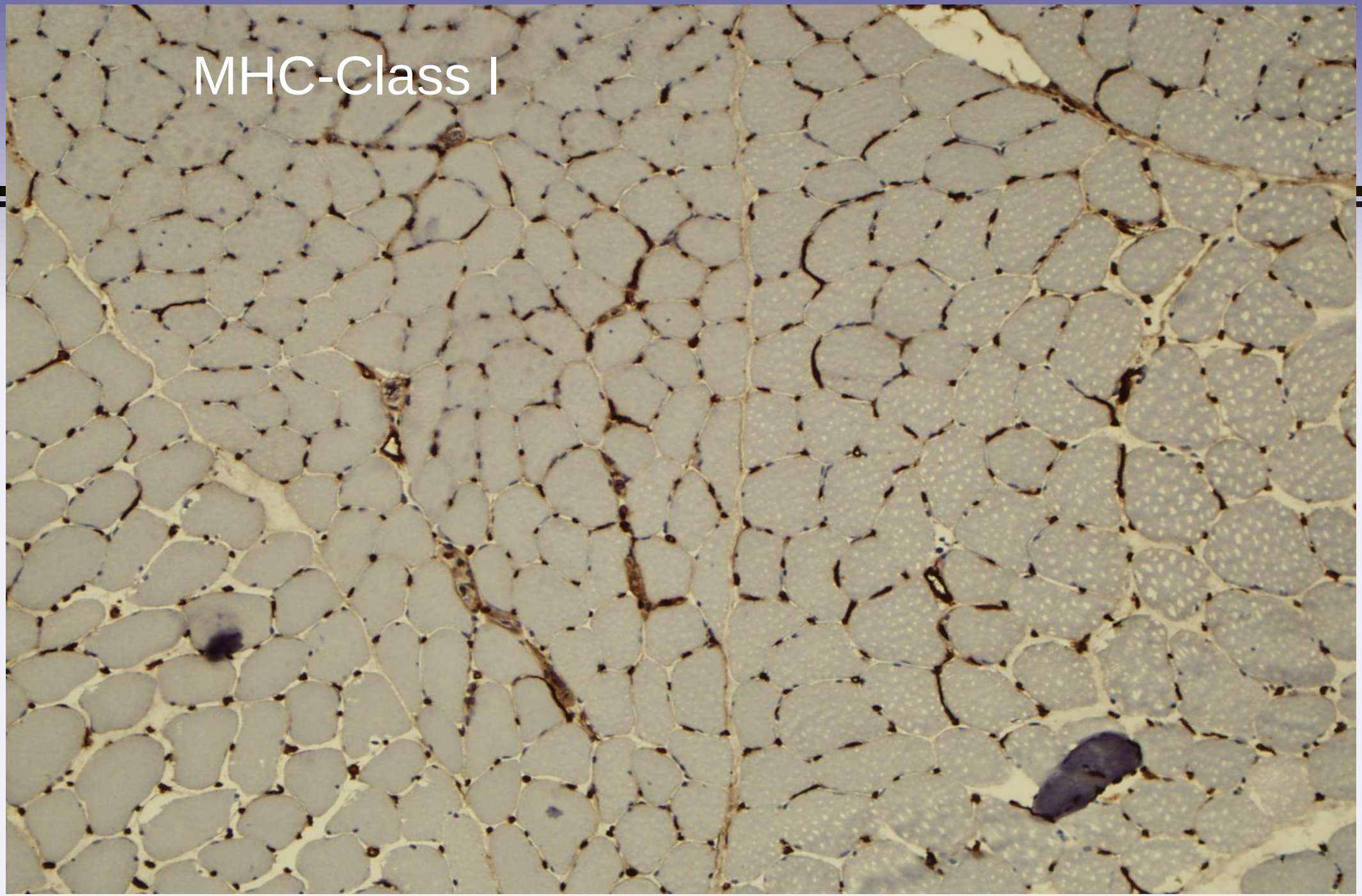


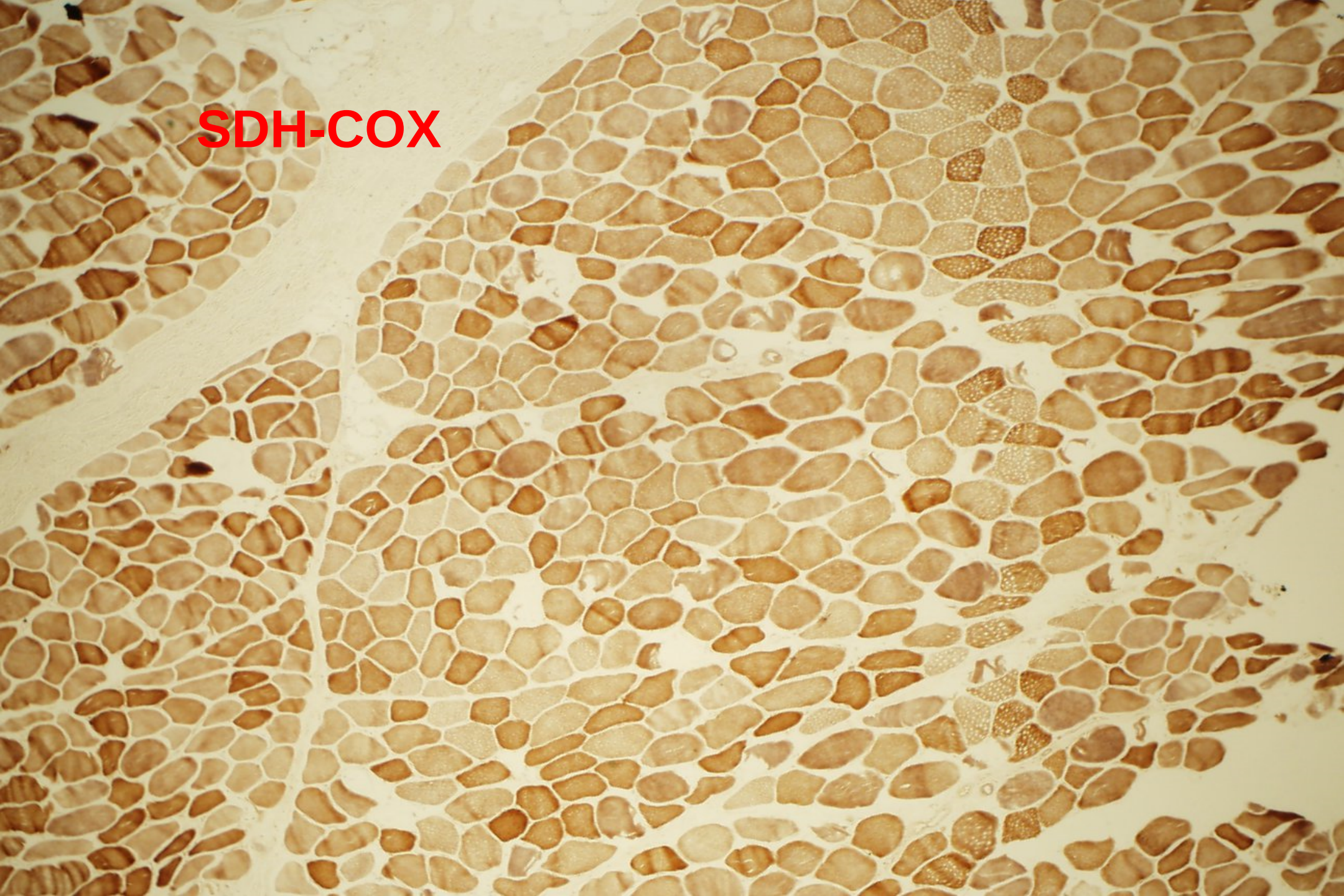


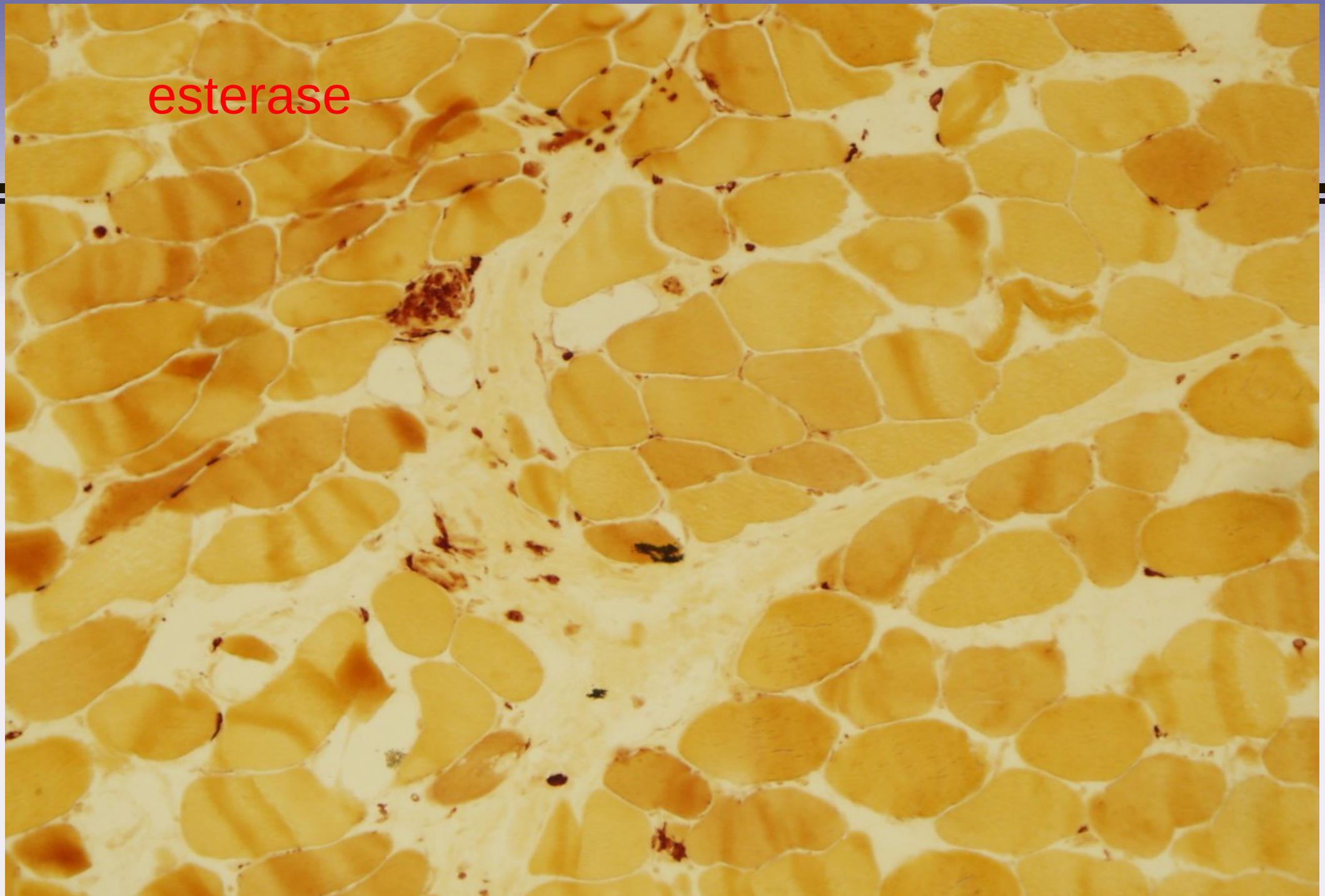


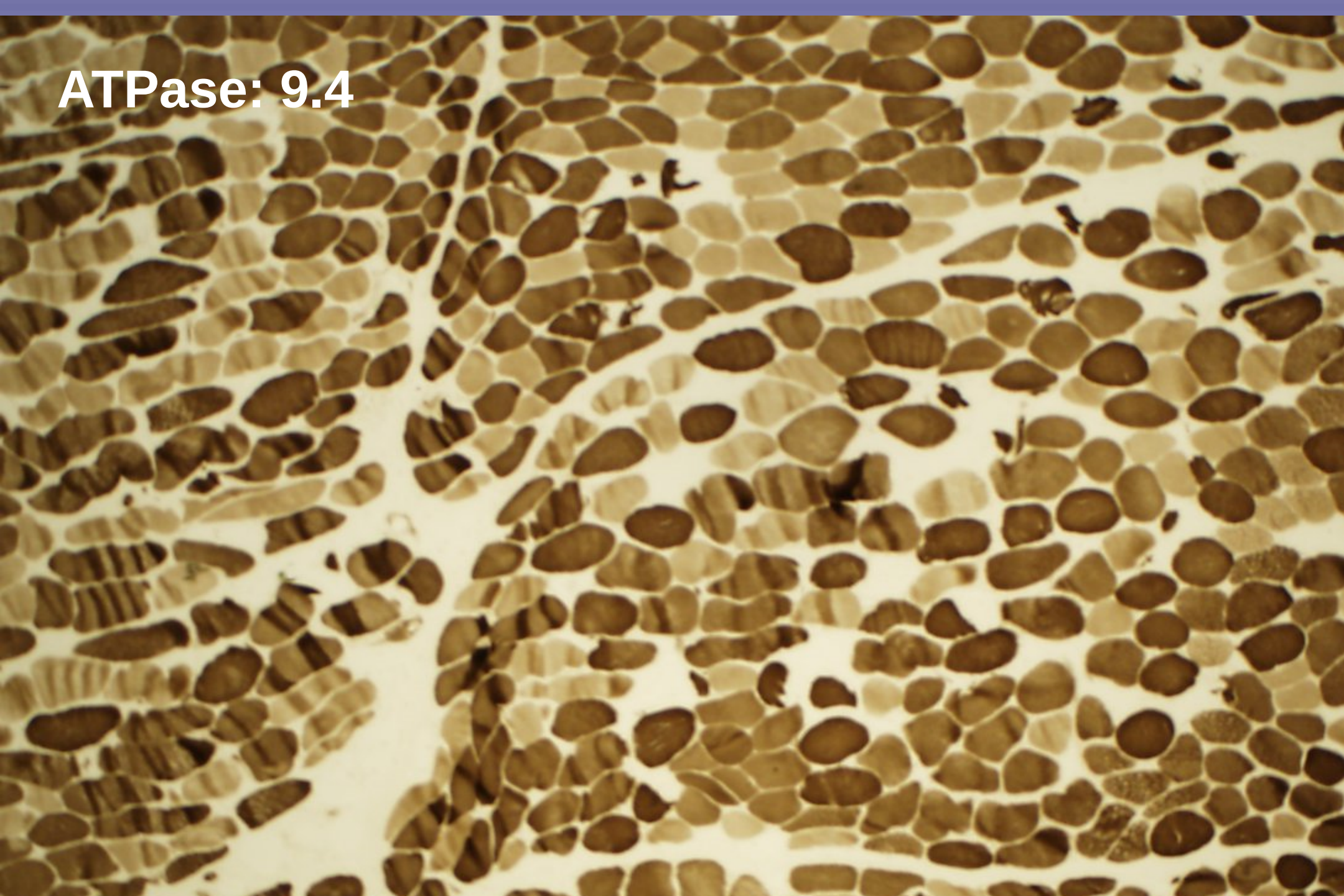

CD163 (macrophages)


CD3 (T cells)


MHC-Class I


SDH-COX


esterase


ATPase: 9.4

Pathologic diagnosis

“Necrotizing Myopathy”

However, this could be:

- **Inflammation**
- **Infection**
- **Toxic**
- **Metabolic**
- **Vascular**
- **…**

Clinical correlation!

The 10 P's:

A Mnemonic to Increase Accuracy of Diagnosis in Acquired Myopathies

1) **Pattern: Proximal UE and LE (Exam limited by pain), No asymmetry**
2) **Period: 1 mo PTA, fairly subacute/acute**
3) **Phenomena: Swelling, severe pain, poorly controlled DM, and non-compliance with HIV Meds**
4) **Pills (/toxins): Statins; discontinued, not been on "D-Drugs" (AZT, etc)**
5) **Plasma: CK: 4K-9K, ? normal levels, negative myositis Ab panel (HMGCR, SRP)**
6) **Pathology: Necrotizing myopathy**
7) **Picture (MRI): asymmetric patchy edema in muscles, no atrophy**
8) **Physiology : NA**
9) **Pedigree: NA**
10) **Pharmacology s/p Steroids 60 mg/day for short time.**

Comments/questions?

Final Diagnosis

Diabetic Myonecrosis

Alternative considerations:

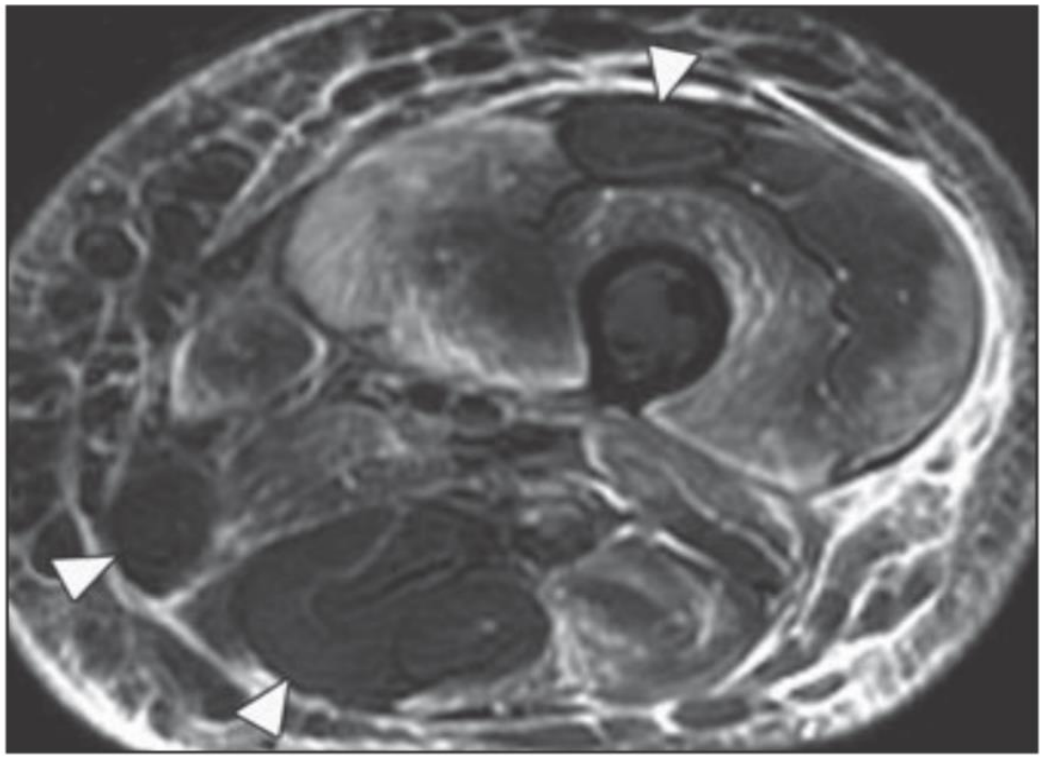
- **Seronegative necrotizing myopathy (MRI, Edema, Pain, Increasing CK with Steroids)**
- **"Myositis"**
- **DVT**
- **Rhabdomyolysis**

AJR Am J Roentgenol. 2010 Jul;195(1):198-204. doi: 10.2214/AJR.09.2494.

Diabetic myopathy: MRI patterns and current trends.

Huang BK[1], Monu JU, Doumanian J.

CONCLUSION: Diabetic myopathy may occur more frequently in patients with type 2 diabetes than previously reported. In this population, T2-weighted and contrast-enhanced images have similar findings, and the increased coexistence of nephropathy makes administration of gadolinium-based contrast agents ill-advised. With a typical clinical presentation and MRI findings, a confident diagnosis can be made, and potentially harmful biopsy is avoided. Diabetic myopathy encompasses a spectrum of diseases, including muscle inflammation, ischemia, hemorrhage, infarction, necrosis, fibrosis, and fatty atrophy. It is usually seen with long-standing, poorly controlled diabetes.



Ann Med Surg (Lond). 2018 Sep 13;35:141-145. doi: 10.1016/j.amsu.2018.09.003. eCollection 2018 Nov.

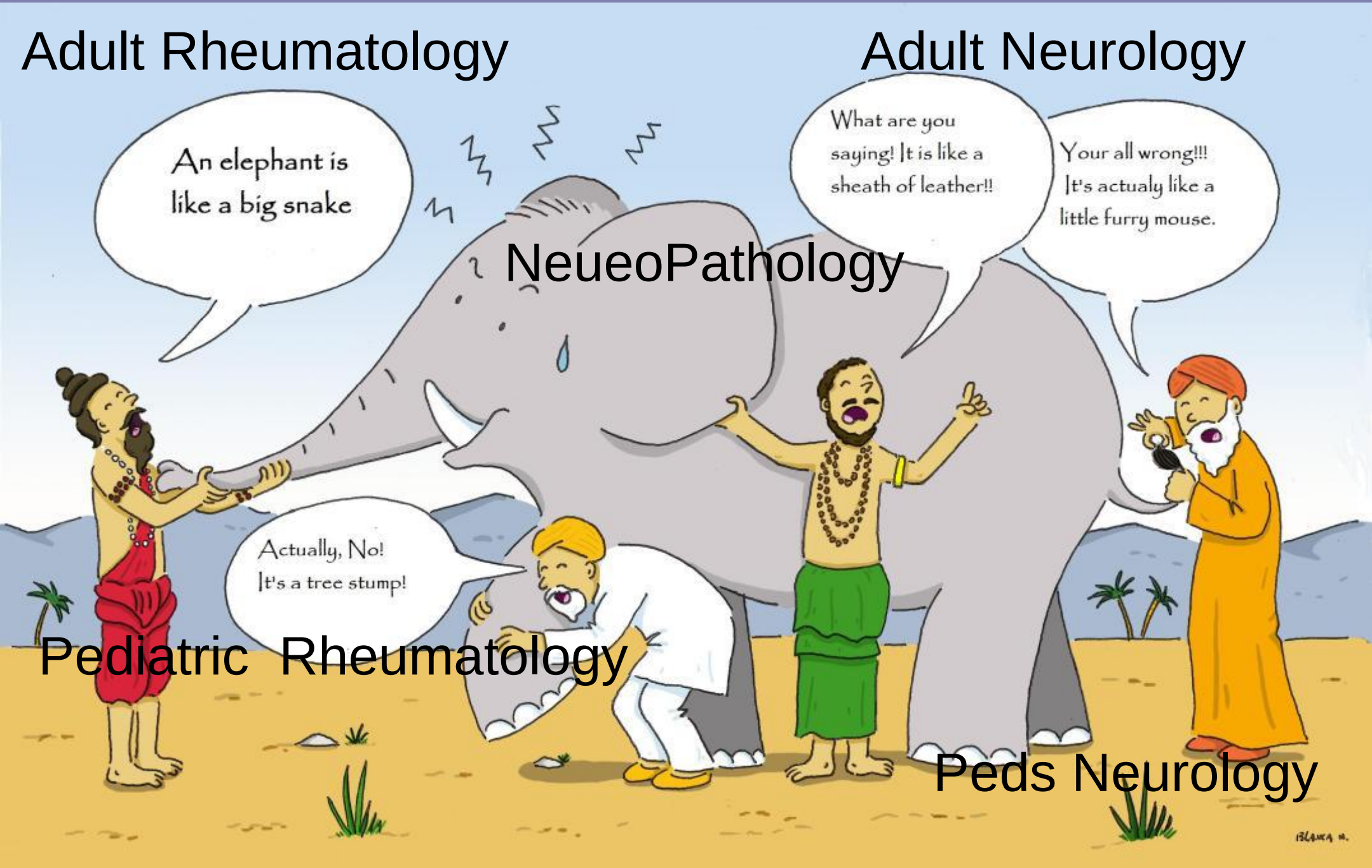
Recurrent diabetic myonecrosis -an under-diagnosed cause of acute painful swollen limb in long standing diabetics.

Gupta S[1,2], Goyal P[2,3], Sharma P[4], Soin P[5], Kochar PS[4].

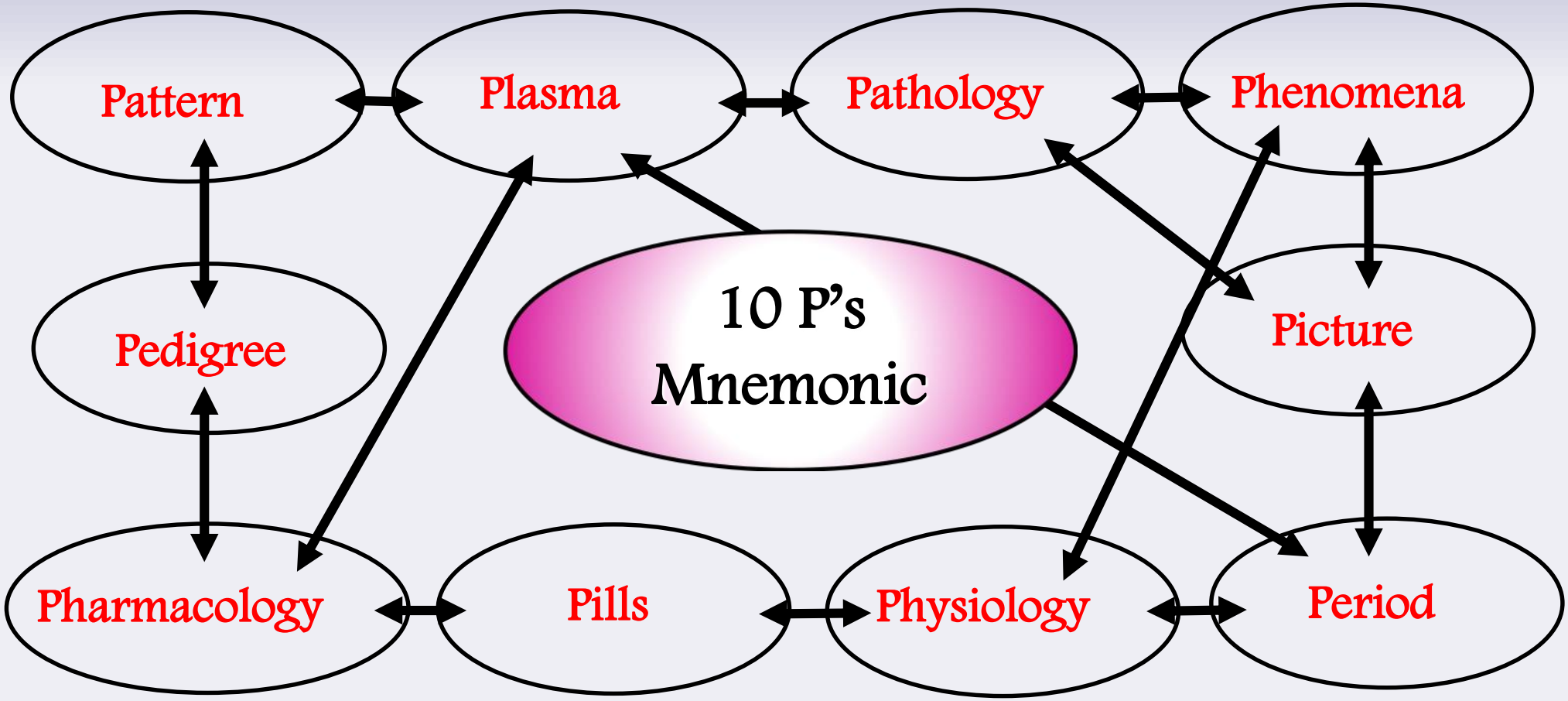
Author information

Abstract

Diabetic myonecrosis (DMN) is an under-diagnosed complication of long-standing poorly controlled diabetes mellitus. It presents as abrupt pain and swelling of the extremity, mostly lower limbs. Diagnosis is often delayed as it mimics a number of clinical entities such as deep vein thrombosis (DVT), cellulitis, necrotizing fasciitis and malignancy. Failure to properly identify this condition can result in increased morbidity through exposure to unnecessary tests and biopsy. A 56-year-old male with a history of complicated type 2 diabetes mellitus, hypertension presented to emergency with gradually worsening left calf pain for last 2 weeks. A lower-extremity venous Doppler was negative for DVT. Magnetic resonance imaging (MRI) was suggestive of muscle edema likely of inflammatory etiology. Muscle biopsy revealed myonecrosis with ischemic myopathy and was negative for vasculitis or inflammatory myopathy. He was managed conservatively and his symptoms resolved in 4 weeks. After 6 months he had recurrence in right thigh which was managed conservatively too. Given these findings, a diagnosis of recurrent diabetic myonecrosis was made. Myonecrosis is a less known microvascular complications of diabetes and should always be keep in mind when evaluating a diabetic patient with muscle pain. Diagnosis can be made on MRI in appropriate clinical settings. The clinical course is usually self-limiting and patients respond well to supportive medical therapy that involves bed rest, strict glycemic control along with analgesic.


Adult Rheumatology
Adult Neurology
An elephant is like a big snake
What are you saying! It is like a sheath of leather!!
Your all wrong!!! It's actualy like a little furry mouse.
NeueoPathology
Actually, No! It's a tree stump!
Pediatric Rheumatology
Peds Neurology

The 10 Ps Mnemonic for acquired myopathies



Monthly online interdisciplinary neuromuscular pathology Case conferences
Kaiser Northern California

4th Wednesday of the month 8:30-10:00

Trainees and colleagues are welcome to join.

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Thank you!

Questions?!