



117TH BOSTON MARATHON

Rheumatology Review



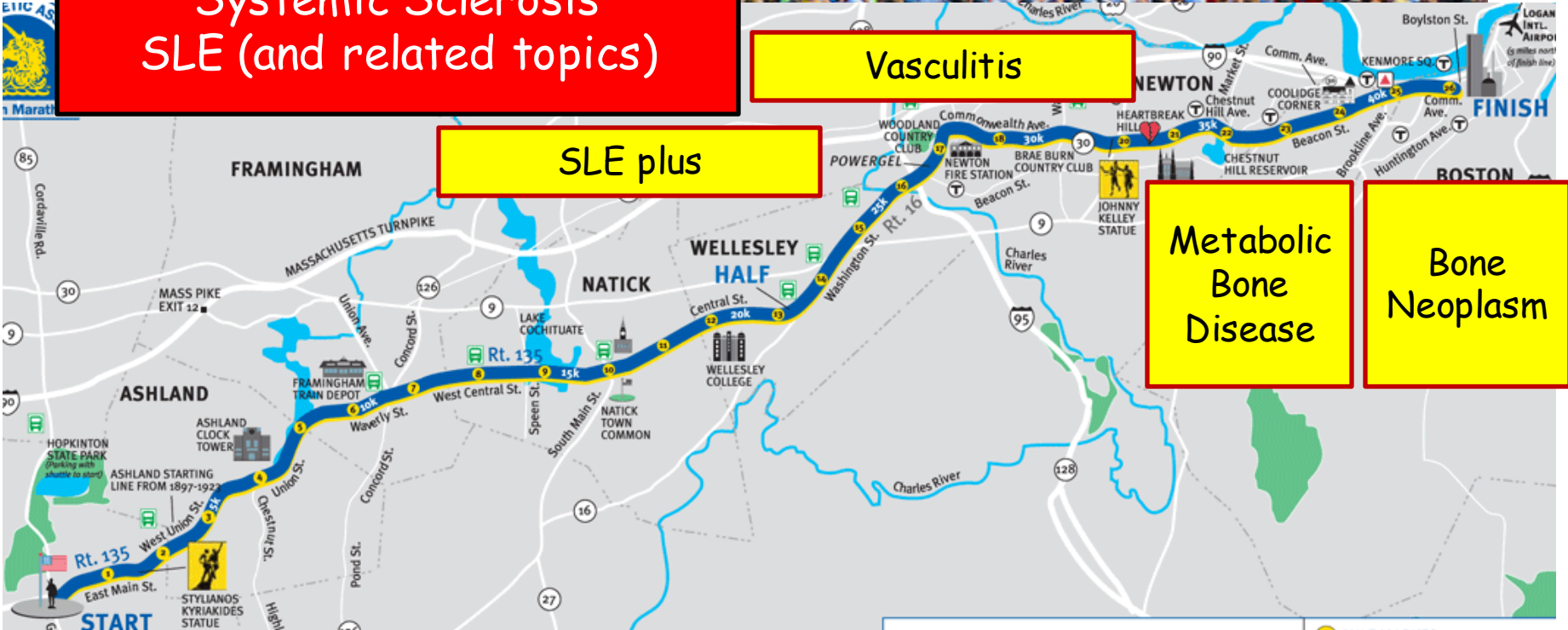
Myositis
Systemic Sclerosis
SLE (and related topics)

Vasculitis

SLE plus

Metabolic
Bone
Disease

Bone
Neoplasm



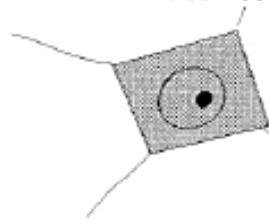
Classification of muscle disease

Diseases of motor neurons

spinal muscular atrophies
motor neuron disease [ALS]

local damage

- trauma, ischaemia,
tumours, infections



**Inflammatory
Myopathies**

**Primary muscle
disease -
myopathies**

Destructive myopathies

Dystrophies
Polymyositis
Dermatomyositis
Local damage - infections
trauma, tumours

Disorders of intracellular organelles and enzymes. e.g. mitochondria, sarcoplasmic reticulum, glycogen metabolism, rods, cores etc

Diseases of peripheral nerves

Axonal degeneration
Segmental degeneration

Disorders of neuromuscular transmission

Myasthenia gravis
Eaton-Lambert syndrome

Myopathies associated with systemic disorders
endocrine and metabolic diseases, carcinoma

Pathology, Myositis (prototypical)

- Common Features: Inflammatory mm disorder

Key Diagnostic Features:

1. Symmetric & Proximal Weakness
2. Elevated CK
3. Characteristic EMG findings
4. MM bx abnormalities

Key Associations:

Skin findings
Systemic complications (i.e. malignancy/IF)

Myositis + EOS

- Heliotrope/Gottron rash

thy
emic

Pathology, Myositis (prototypical)

- **Common Features:** Inflammatory mm disorder
- **Presentation:** *Symmetric AND Proximal mm weakness*
- **Diagnosis:** *Elevated CK; Characteristic EMG findings; MM bx abnormalities*
- **Pathology:** All myopathies have evidence of mm fiber injury (necrosis/regeneration) and mononuclear inflammation (location?)

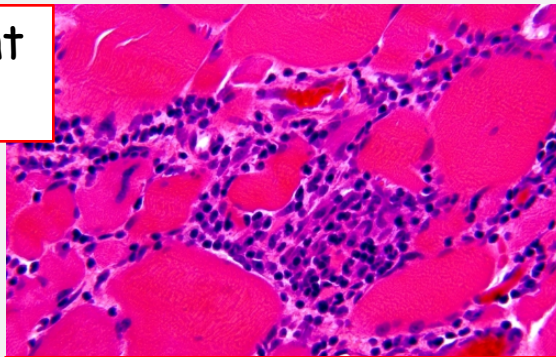
Pathology, Myositis

- **Common Features:** Inflammatory mm disorder
- **Presentation:** *Symmetric AND Proximal mm weakness*
- **Diagnosis:** *Elevated CK; Characteristic EMG findings; MM bx abnormalities*
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Dermatomyositis is **NOT** simply Polymyositis with skin involvement

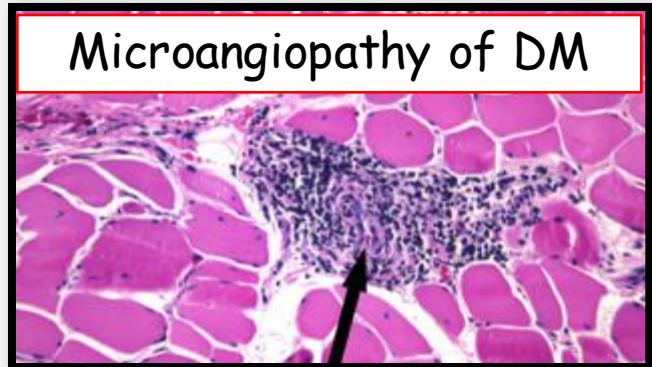
PM is mediated by **cytotoxic T cells**; DM is **humorally** mediated.

Patchy involvement
of endomysium



Polymyositis

Microangiopathy of DM



Pathology, Myositis

- **Common Features:** Inflammatory mm disorder
- **Presentation:** **Symmetric** AND Proximal mm **weakness**
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- **Pathology:** All myopathies have evidence of mm fiber injury (necrosis/regeneration) and mononuclear inflammation

Polymyositis Features:

- Pathology/Pathogenesis:
 - **Cytotoxic T cell mediated**
 - T-cell **infiltrates scattered in mm**
 - No vacuoles/inclusion bodies

Dermatomyositis Features:

- Pathology/Pathogenesis:
 - **Microangiopathy**, humoral mediated
 - **Perivascular injury** leads to ischemic appearance w/ mm atrophy

Pathology, Myositis

- **Common Features:** Inflammatory mm disorder
- **Presentation:** **Symmetric** AND Proximal mm **weakness**
- **Diagnosis:** Elevated **CK**; Characteristic EMG findings; MM bx abnormalities
- **Pathology:** All myopathies have evidence of mm fiber injury (necrosis/regeneration) and mononuclear inflammation

Polymyositis Features:

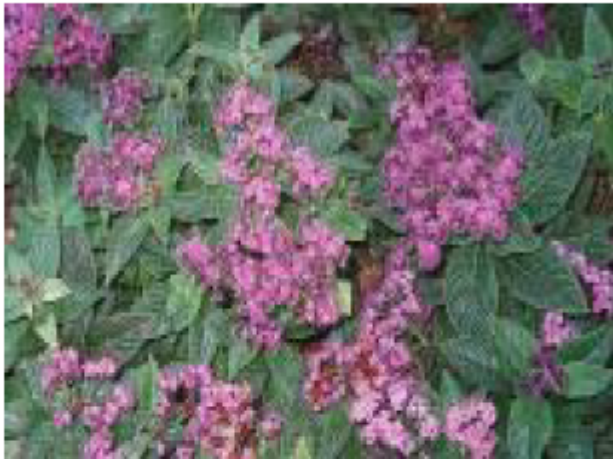
- **Pathology/Pathogenesis:**
 - Cytotoxic T cell mediated
 - T-cell infiltrates scattered in mm
 - No vacuoles/inclusion bodies
- **Complications:**
 - **ILD** more common, esp if **Anti-Jo-1 +**
 - Malignancy < DM

Dermatomyositis Features:

- **Pathology/Pathogenesis:**
 - Microangiopathy, humoral mediated
 - Perivascular injury leads to ischemic appearance w/ mm atrophy
- **Complications:**
 - **Malignancy** > PM
 - ILD
- **Myositis PLUS**
 - Heliotrope/Gottron rash

Dermatomyositis – specific rash

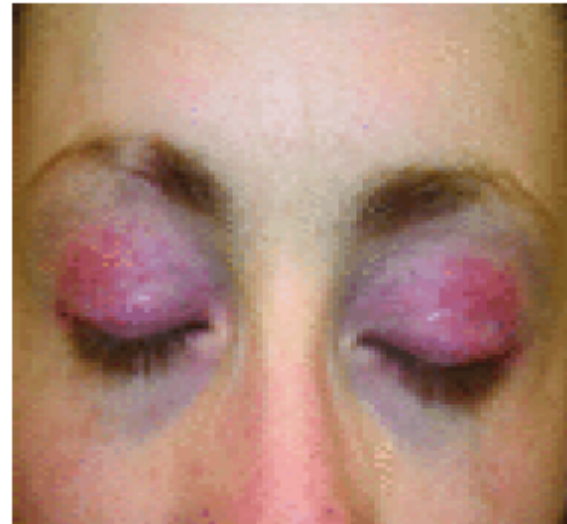
Heliotrope flower



Helio - Gk: sun
Trope - Gk: turn

Heliotrope

Violaceous to dusky periorbital rash



Violaceous

Dermatomyositis – specific rash

Gottron's papules – slightly elevated
violaceous papules

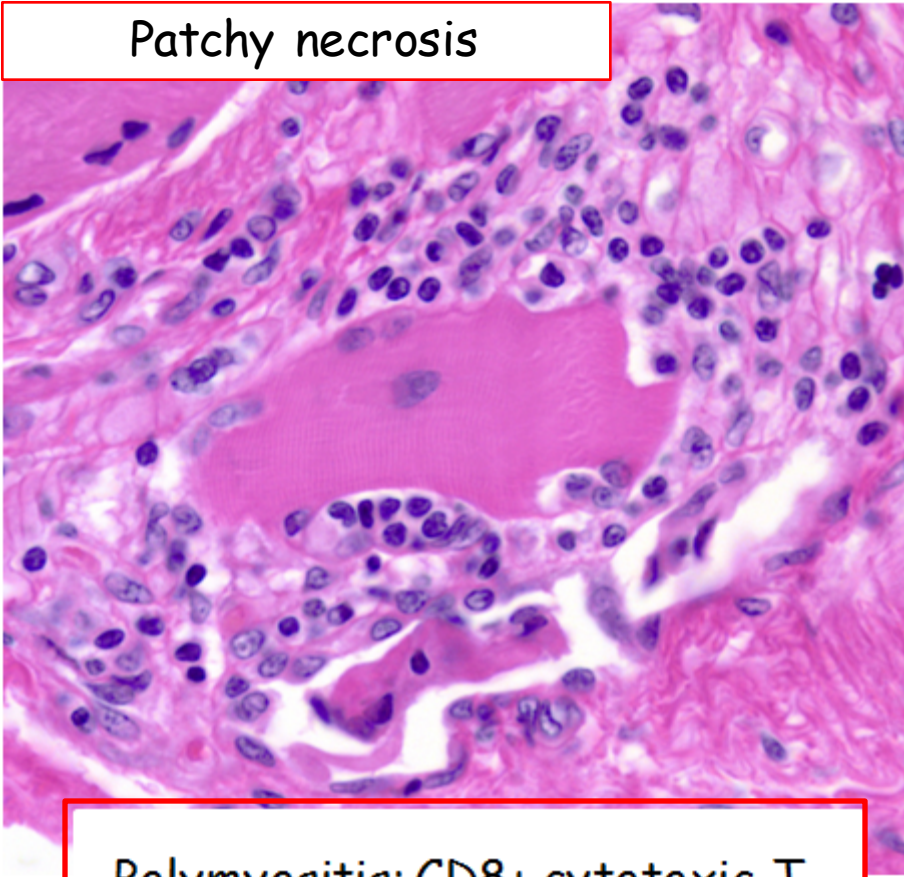


Photodistribution but photo-
sensitivity unusual



Gottron's: Language of DM

Patchy necrosis



Polymyositis: CD8+ cytotoxic T-cells b/w myofibers

Endomyosial capillaritis (vasculitis)

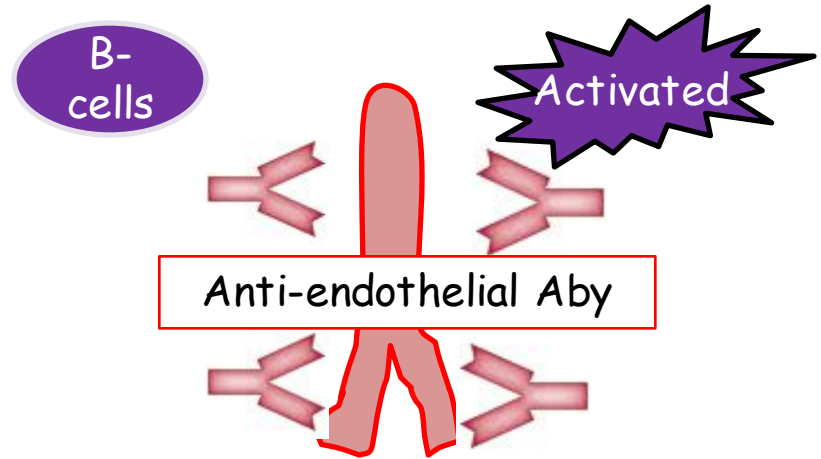
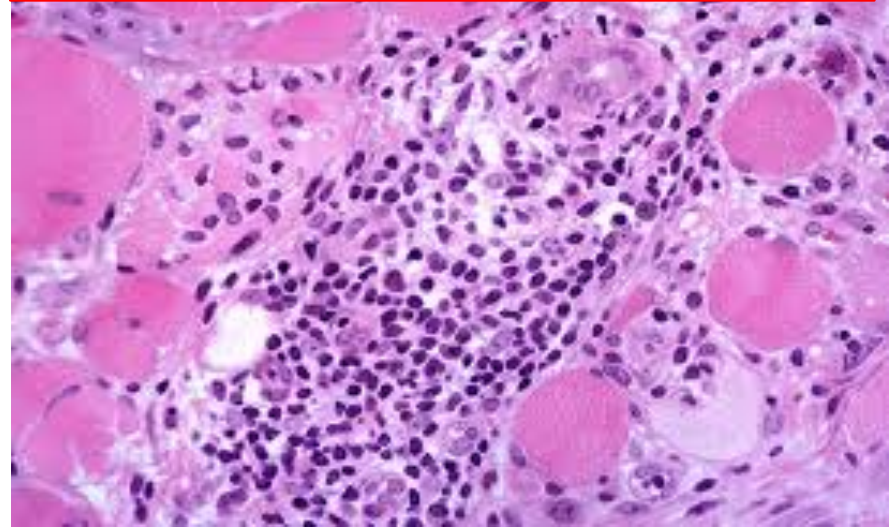


TABLE 6-9 -- Antinuclear Antibodies in Various Autoimmune Diseases

Nature of Antigen	Antibody System	Disease, % Positive					Inflammatory Myopathies
		SLE	Drug-Induced LE	Systemic Sclerosis—Diffuse	Limited Scleroderma—CREST	Sj?gren Syndrome	
Many nuclear antigens (DNA, RNA, proteins)	Generic ANA (indirect IF)	>95	>95	70–90	70–90	50–80	40–60
							<5
							<5
							<5
							<5
							10
							<5
							<5
							<5
Histidyl-tRNA synthetase	Jo-1	<5	<5	<5	<5	<5	25

Be familiar w/ nomenclature



Anti Jo-1 (histidyl-tRNA synthetase)

1. Correlates with disease activity
2. Useful marker for polymyositis, esp associated IF

Alphabet Soup ('Myositis Panel'):
Anti: MI-2, PL-7, PL-12, EJ, OJ, KU, SRP, U2 SN RNP

Myositis “mimickers”



Hypothyroidism
Cushing's syndrome



Steroids
Statins
EtoH, cocaine



Myasthenia gravis
ALS



Viral (flu, HIV, CMV)
Parasitic (trichinosis, toxo)



Muscular dystrophy
Metabolism disorders

cores etc

Myopathies associated with systemic disorders
endocrine and metabolic diseases, carcinoma

Myositis “mimickers”



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Muscular dystrophy
Metabolism disorders

Key Differential Dx: PMR

Elevated ESR
Shoulder/Thigh ache (mimics weakness)
Normal CK

Myositis

- Background
 - Proximal/symmetric weakness
 - Elevated CPK
 - Characteristic EMG findings
 - **Dx:** MM bx
- Polymyositis
 - T-cell infiltrate (**cytotoxic T-cells**) w/ patchy necrosis in endomysial location (between mm fibers).
 - Anti-Jo 1; histidyl-tRNA synthetase (a/w ILD)
- Dermatomyositis
 - Perivascular infiltrate (**B-cell mediated**) that causes a microangiopathy with anti-endothelial abx → perifascicular ischemia/atrophy
 - **Rash:** violaceous [eyes/hands (papules)]
- Complications: ILD, Malignancy
- Key Diff Dx: PMR