Dermatomyositis & Polymyositis

Proximal weakness with a rash, or without one · the antibody decides the lung risk, the cancer risk and the prognosis

Screen for cancer *and for ILD*

RECOGNIZE IT *weakness that is proximal, symmetric and usually painless*

THE WEAKNESS

Proximal, symmetric, progressive over weeks to months. Trouble rising from a chair, climbing stairs, washing hair. **Usually painless**, which surprises people. **Ask about dysphagia**: pharyngeal involvement predicts aspiration and is a reason to admit.

THE RASH, IF PRESENT

Gottron papules over the knuckles and **heliotrope rash** on the eyelids are close to pathognomonic. Also the **shawl and V signs**, **mechanic's hands**, nailfold capillary changes and calcinosis. **The rash can precede the weakness by months.**

WHAT IT IS NOT

Distal weakness, asymmetry, or finger flexor and quadriceps wasting in an older man suggests inclusion body myositis, which is slowly progressive and **does not respond to immunosuppression**. Getting this wrong means years of futile steroids.

THE WORKUP *and what each test is actually for*

Test	What it adds
CK, aldolase, LDH	CK is usually **markedly raised**, often in the thousands, and tracks disease activity. **A normal CK does not exclude it**, particularly in amyopathic dermatomyositis and in longstanding disease with muscle atrophy
Myositis antibody panel	**The highest-yield test on this list**, because the specific antibody predicts the complications better than the muscle biopsy does. See the table below
MRI of thigh muscles	Shows **edema in active inflammation** and, importantly, **guides where to biopsy**, which reduces the sampling error that causes false-negative biopsies
EMG and biopsy	EMG shows an irritable myopathic pattern. **Biopsy remains the reference standard** and distinguishes the subtypes, including inclusion body myositis
Always exclude the mimics	**Check TSH** (hypothyroid myopathy), review **statins and steroids**, and consider alcohol, electrolyte disturbance and muscular dystrophies. **Steroid myopathy is a common confounder in a patient already being treated**, and it has a normal CK

THE ANTIBODY CHANGES THE PLAN *this is the most useful table on the sheet*

Antibody	Phenotype	What it obliges you to do
Anti-Jo-1 and other anti-synthetases	**Antisynthetase syndrome**: myositis, **ILD**, mechanic's hands, Raynaud, arthritis, fever	**Screen the lungs actively** with high-resolution CT and pulmonary function tests. The ILD, not the muscle disease, usually determines outcome
Anti-MDA5	**Clinically amyopathic** dermatomyositis with skin ulceration, often with a **normal or near-normal CK**	**Rapidly progressive ILD with high mortality.** This is the one to treat urgently and aggressively, and a normal CK must not be reassuring here
Anti-TIF1-gamma, **anti-NXP2**	Dermatomyositis in adults, often with prominent skin disease	**Strongly associated with underlying malignancy.** These findings justify a thorough, and if needed repeated, cancer search
Anti-SRP	**Immune-mediated necrotizing myopathy**: severe weakness, very high CK, little inflammation on biopsy	Often **refractory to steroids alone**; usually needs early combination immunosuppression, and IVIG is frequently used
Anti-HMGCR	**Statin-associated necrotizing myopathy**	**It does NOT resolve on stopping the statin**, unlike ordinary statin myalgia, and it **requires immunosuppression**. Persistent weakness and a high CK weeks after stopping a statin should prompt this test

MALIGNANCY SCREENING *a defining obligation in adult dermatomyositis*

⚠ DERMATOMYOSITIS IS A PARANEOPLASTIC DISEASE UNTIL PROVEN OTHERWISE

Adult dermatomyositis carries a **substantially increased cancer risk, concentrated in the first years around diagnosis**. Do an **age and sex appropriate search**: CT of chest, abdomen and pelvis, mammography and pelvic assessment in women, and colonoscopy per age. **Risk is highest with anti-TIF1-gamma and anti-NXP2**, and lower in antisynthetase syndrome and juvenile disease. **A negative initial search does not close the question**: repeat surveillance over the following years, especially in refractory disease, since **myositis that will not respond is a classic reason to look again for an occult tumor**.

THE ORGANS BEYOND MUSCLE *each one is missed by examining only strength*

Organ	What happens	What to do
Lung the main killer	**ILD**, plus ventilatory failure from respiratory muscle weakness	**High-resolution CT and pulmonary function tests at diagnosis** in anyone with an antisynthetase or MDA5 antibody, cough or breathlessness. Mycophenolate or rituximab are commonly used where ILD dominates
Swallow	Pharyngeal and upper esophageal weakness	**Ask every patient about choking and nasal regurgitation.** Formal swallow assessment, and consider nil by mouth with alternative feeding while treatment takes effect
Heart	Myocarditis, conduction disease and arrhythmia, usually subclinical	**ECG in everyone.** A raised troponin may reflect the myositis itself rather than the heart, so interpret it alongside imaging rather than reflexively treating for ACS
Skin	Photosensitive rash, and **calcinosis**, which is more common in juvenile disease and is difficult to treat	**Rigorous photoprotection**, since ultraviolet light drives the rash. **Hydroxychloroquine helps the skin but not the muscle**, and the skin disease often persists after the weakness has resolved

TREATMENT & PEARLS

- **High-dose glucocorticoids are first-line**, with a **steroid-sparing agent started early rather than later** (methotrexate or azathioprine, mycophenolate where there is ILD). IVIG is effective and is favored in severe, refractory or necrotizing disease and where infection risk limits immunosuppression.
- **Track response with strength and function, not the CK alone.** The CK often falls before strength improves, and a rising CK in a recovering patient is not automatically a relapse.
- **Add PJP prophylaxis** with prolonged high-dose steroids or combination immunosuppression, plus bone protection and gastric protection. **If a treated patient gets weaker with a normal CK, suspect steroid myopathy** and reduce rather than escalate.
- **Assess swallow and respiratory muscles at presentation.** Dysphagia predicts aspiration, and diaphragmatic weakness can cause **ventilatory failure with a normal chest radiograph**. Both change disposition immediately.

References: EULAR/ACR classification criteria for idiopathic inflammatory myopathies · published literature on myositis-specific autoantibodies and their phenotypes · cohort studies of malignancy risk in dermatomyositis.

Educational aid; verify against current guidance, local formulary and patient factors.