Polymyositis / Dermatomyositis Disease

Polymyositis (PM) and dermatomyositis (DM) are two separate but closely related diseases that are classified as idiopathic inflammatory myopathies. These diseases are more common in women in the fourth and fifth decade of life. The underlying immunology of polymyositis involves endomysial inflammation mediated by CD8+ T cells, while dermatomyositis involves perimysial inflammation mediated by CD4+ T cells. Both diseases present with symmetric proximal muscle weakness, dysphagia, dysphonia and cardiopulmonary involvement. Dermatomyositis has unique cutaneous findings that include a malar rash, Gottron papules, heliotrope rash, and the shawl and V signs.



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Pathophysiology

Idiopathic Inflammatory Myopathy
Idiot-hat In-flames with Muscle
Both polymyositis and dermatomyositis are acquired, noninfectious diseases that involve inflammation of muscle tissue due to an aberrant immunological response. The exact cause of either disease is unknown, however research has shown associations to different cancers, most notably lung, gastric, and genitourinary carcinomas.

More Common in Women
Female Sex-symbol
These diseases appear more often in women than men and during the fourth and fifth decade of life, however children may still be affected by either disease.

Endomysial Inflammation with CD8+ Cells (Polymyositis)
CD with (8) Ball-inside
Within a muscle, the connective tissue layer that surrounds an individual muscle fiber is called the endomysium. It ensheaths and protects the muscle fiber, but when CD8+ T cells (cytotoxic T cells) attack and inflame this layer, muscle fibers weaken and symptoms of PM manifest.

Perimysial Inflammation with CD4+ Cells (Dermatomyositis)
CD with (4) Fork
Individual muscle fibers are grouped into bundles called fascicles; these bundles are bound together and protected by a connective tissue layer called the perimysium. Inflammation of this layer mediated by CD4+ T cells (helper T cells) and antibody activation causes muscle fiber weakness and manifestation of DM symptoms.

Signs and Symptoms (Both)

Symmetric Proximal Muscle Weakness
P-roximal Muscles
Proximal muscle weakness is the most common clinical symptom of these diseases; weakness will be bilateral and affect hip and shoulder strength to the greatest degree. Patients may complain of difficulty climbing stairs, rising from a seated position, trouble combing their hair, and reaching for high objects.

Dysphagia
Dice-fajita
As the disease progresses, weakness of esophageal muscle in the upper third (striated, voluntary muscle) of the tract will present with dysphagia, and possible aspiration of solids and liquids. These patients are at increased risk for the development of aspiration pneumonia.

Lung and Cardiac Involvement
Lungs & Heart Exposed
These patients are at higher risk for developing interstitial lung disease due to pulmonary inflammation, as well as respiratory failure due to diaphragm and intercostal muscle weakness. They are also at greater risk for congestive heart failure, myocardial infarction, and myocarditis leading to cardiac arrhythmias

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and conduction defects.

Signs and Symptoms (Dermatomyositis)

Malar Rash

Monarch-face-butterfly

Classically associated with systemic lupus erythematosus, a malar or butterfly rash may also occur in DM. Appearing bilaterally on the cheeks in a symmetric distribution, the rash is reddish-purple, scaly, and non-pruritic. Of particular note, the facial rash seen in polymyositis and dermatomyositis includes the nasolabial folds.

Shawl-like Shoulder Rash

Shawl-like-rash on Shoulders

Both these signs refer to the characteristic distribution of erythema and poikiloderma (mixed hyperpigmentation and hypopigmentation). The rash appears in the areas a shawl would cover when worn, specifically the upper back and arms, hence the name shawl sign. V-sign refers to appearance of a rash on the upper chest and anterior neck.

Gottron Papules on Knuckles

Gauntlet with Papules on Knuckles

Erythematous and scaling eruptions on the extensor surfaces of the knuckles are known as Gottron papules; if found on other extensor surfaces, like the elbows and knees, they are called Gottron sign, and are considered pathognomonic for DM.

Heliotrope Rash

Heliotrope-flowers

A heliotrope rash is a violaceous eruption on the upper eyelids with surrounding periorbital edema, and it is sometimes called raccoon eyes.

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