Polymyositis / Dermatomyositis Diagnosis and Treatment

Polymyositis (PM) and dermatomyositis (DM) are two separate but closely related diseases that are classified as idiopathic inflammatory myopathies. Diagnostic labs include increased creatine kinase (CK), increased aldolase, presence of anti-Jo-1 antibodies, antinuclear (ANA) antibodies, and anti-SRP antibodies, abnormal electromyography (EMG), and muscle biopsy findings. Initial treatment involves immunosuppression with corticosteroids, methotrexate, and azathioprine. For disease cases resistant to initial treatment, additional therapy with rituximab, intravenous immunoglobulin (IVIG), calcineurin inhibitors like cyclosporine and tacrolimus, and cyclophosphamide may be utilized.



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Diagnosis

Increased CK (CPK)
Up-arrow with CK-Model
An elevated serum level of muscle enzymes like creatine kinase (CK) and aldolase are characteristic but not specific to dermatomyositis and polymyositis. However, they are useful initial tests when the diagnoses are suspected.

Increased Aldolase
Up-arrow with Aldo-with-lace
An elevated serum level of muscle enzymes like CK and aldolase are characteristic but not specific to dermatomyositis and polymyositis. However, they are useful initial tests when the diagnoses are suspected.

Anti-JO-1 Antibody
Ant-tie-cup-of-joe and (1) wand
Myositis-specific antibodies like anti-Jo-1 antibody and anti-SRP (signal recognition particle) antibody can be detected in both these diseases. Anti-Jo-1 antibody is a type of antisynthetase antibody, a class of antibody that is related to an overarching condition called antisynthetase syndrome, that some patients with DM or PM may fall under.

Anti-SRP Antibody
Anti-tie-Slurpee
Myositis-specific antibodies like anti-Jo-1 antibody and anti-SRP (signal recognition particle) antibody can be detected in both these diseases. Anti-SRP antibodies are almost always found in PM patients and often indicate severe, treatment-resistant disease progression. Other antibodies that may be identified in these conditions include anti-Mi2, anti-PM-Scl, and anti-Ku.

Antinuclear Antibody
Ant-tie-nuclear
This larger class of antibody is detected in a variety of immune conditions; their presence is common in DM and PM patients and can be seen with immunofluorescent staining.

Abnormal EMG
Abnormal-Electric-Muscle-Lines
Electromyography, or EMG, is a diagnostic test that uses electrical energy to demonstrate muscular activity. Either a surface or needle electrode is placed on or within the muscle to trigger and measure electrical activity produced by a muscle; these measurements are then used to generate an electromyogram. Patients with DM or PM have weak muscle fibers, and therefore abnormal EMG findings, like spontaneous fibrillations and repetitive discharges. EMG serves to differentiate myopathies from neuropathic disorders like myasthenia gravis.

Biopsy
Biopsy-needle
For a definitive diagnosis of PM or DM, a muscle biopsy provides positive confirmation. Biopsy for PM shows CD8+ T-cell infiltration, along with necrotic muscle fibers. Muscle biopsy for DM shows CD4+ T-cell infiltration along with perifascicular atrophy, and a skin biopsy may also support the etiology of

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the cutaneous features.

Treatment

Corticosteroids

Quarter-on-steroids

Corticosteroids suppress the immune system by upregulating anti-inflammatory proteins and downregulating pro-inflammatory proteins within cells. Systemic corticosteroid therapy for a duration of nine months to a year is a common first approach to controlling these myopathic diseases. Either prednisone or methylprednisolone is given in high doses initially, and then tapered off gradually depending on clinical response.

Methotrexate

Moth-T-REX-ate

Methotrexate (MTX) is an immunosuppressive drug that inhibits dihydrofolate reductase. It may be used as an adjunct to corticosteroid therapy, and it decreases the need for high-dose steroids, thereby reducing their side effects.

Azathioprine

Astronaut-police

Azathioprine (AZA) is another immunosuppressive drug that acts as an antimetabolite in order to interfere with DNA synthesis within a cell. Combination with glucocorticoids has been shown to be effective with improved long-term patient outcomes over single agent steroid therapy.

Treatment For Resistant Disease

Intravenous Immunoglobulin (IVIG)

Ivy-gold-goblin

Intravenous immunoglobulin (IVIG) is a blood product used to slow down an aberrant immune system via several, but debated mechanisms. It has been shown to be effective in controlling treatment resistant dermatomyositis, but it is not effective for polymyositis.

Cyclophosphamide

Cyclops-phosphate-P

Classified as an alkylating agent, cyclophosphamide cross-links DNA to trigger cell death. It is used for chemotherapy as well as autoimmune conditions. Side effects include myelosuppression and hemorrhagic cystitis, so its use to treat PM and DM should only be considered after second-line agents have failed.

Rituximab

Red-tux-mob

Monoclonal antibody to the B-cell marker CD20, used as both a chemotherapeutic and immunosuppressive agent. When treatment to corticosteroid therapy plus MTX or AZA fails, rituximab is often used.

Calcineurin Inhibitors

Calcium-cow-urchin with Inhibiting-chains

Cyclosporine and tacrolimus work by inhibiting the calcineurin cellular receptor, which subsequently prevents T-cell activation. These medications are considered in resistant PM and DM, and in cases involving interstitial lung disease.

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