A. Polymyositis, Dermatomyositis, Inclusion-Body Myositis

The clinical picture of the various forms of myositis can vary greatly. The diagnostic criteria are presented in (**4**). Polymyositis (**1**) is characterized by proximal muscle weakness, especially in the shoulders, upper arms, and thighs. The patients experience difficulties in getting up out of a chair and climbing stairs. Dermatomyositis (**2**) is additionally characterized by the occurrence of skin manifestations, especially on areas of the skin exposed to light. One of the most common manifestations is the so-called heliotrope rash (**5**). Characteristic papuloid skin changes called Gottron's sign develop on the knuckles (**6**). A concomitant tumor is often present in many dermatomyositis patients over 50 years of age. Hence, selective investigations for carcinomas must be made, especially in the region of the breasts, lungs, and gastrointestinal tract. The autoantibody findings are generally uncharacteristic; low titers of anti-nuclear antibodies are occasionally found. Conventional blood tests reveal high concentrations of creatine kinase (CK) and myoglobulin. The presence of anti-proteasome antibodies was also recently demonstrated.

Anti-synthetase syndrome (**2**) is an independent disease entity characterized by the presence of Jo-1 antibodies, which are directed against histidyl synthetase. The typical clinical features of the disease include interstitial lung disease, arthritis, Raynaud's phenomenon, skin manifestations, fever, and muscular weakness.

Inclusion-body myositis (**3**) is another distinct disease entity. Unlike the aforementioned diseases, it has a predilection for distal muscles. For the most part, this entity has no characteristic laboratory features.

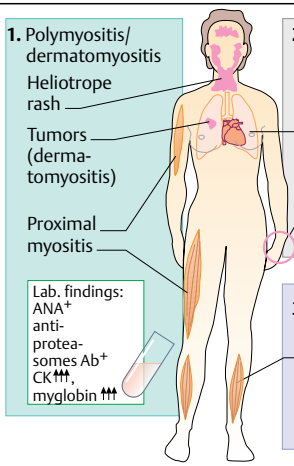
B. Histology of Inflammatory Myopathies

The histological pictures of polymyositis and dermatomyositis vary greatly. In polymyositis, $CD8^+$ T cells directly infiltrate the individual muscle cells within the muscle fibers (see also **A.7**). Dermatomyositis, on the other hand, is characterized by vasculitis with concomitant perivascular inflammation under the influence of $CD4^+$ T cells. In this case, myocyte death occurs as a secondary effect of vessel lesions.

C. Pathogenesis

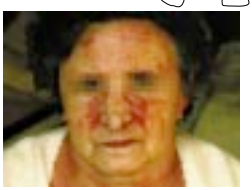
Like their histology, the pathogenesis of polymyositis (PM) and dermatomyositis (DM) also differs. Class I MHC-mediated mechanisms play a predominant role in polymyositis. In polymyositis, aberrant expression of class I HLA antigens occurs on the surface of muscle cells, which are normally HLA-negative (only a few other cells in the body are HLA-negative), due to a genetic predisposition and unknown (viral?) factors. Cytotoxic T cells then recognize the altered myocytes as "foreign" and destroy them. This is the histological basis of the aforementioned characteristic processes of cell death within the muscle bundle. Similar pathogenetic mechanisms are suspected in inclusion-body myositis (IBM). The mechanism of development of amyloid-containing cell inclusions in IBM is still unclear.

In dermatomyositis, the inflammation arises from the perimysial vessels. Hence, blood vessel damage is a central element in the pathogenesis of the disease. The resulting tissue ischemia leads to secondary death of the muscle cells, characteristically with pronounced perifascicular involvement.

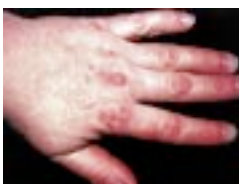


a) Typical clinical findings
b) Increased creatine kinase activity
c) Multifunctional changes in electron micrographs
d) Bioptic proof of myositis
e) Myositis-associated antibodies

4. Diagnostic criteria



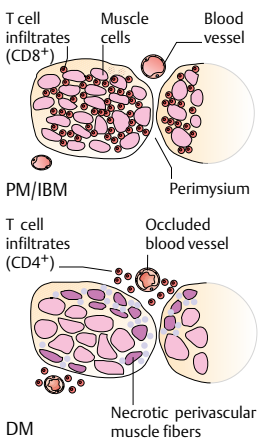
5. Heliotropic exanthema



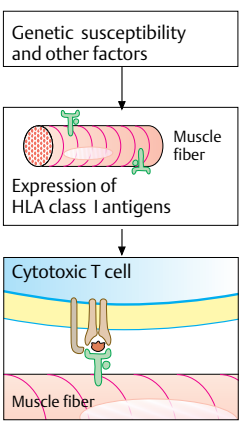
6. Gottron's sign

7. Histology of polymyositis

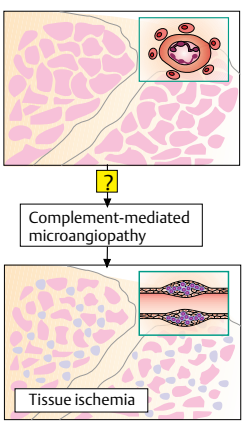
A. Polymyositis/dermatomyositis/inclusion body myositis



B. Histology of myositis



1. PM/IBM



2. Dermatomyositis

C. Pathogenesis