Vasculopathy in Dermatomyositis

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Inflammatory myopathy is frequently observed in various types of collagen diseases and related disorders, and among them, polymyositis (PM) and dermatomyositis (DM) show the most prominent muscular symptoms. Previously, both diseases were considered simply to be varieties of the identical disease, and sometimes PM/DM syndrome was used as an inclusive term. But today, the difference between the two is clear immunologically as well as pathologically. Although striated muscle is the main target of the autoimmune mechanism in both, in DM, the intramuscular small blood vessels are more dominant effector sites, while in PM, the muscle fibers are more direct targets. Occlusions of arterioles and the resultant necrosis of the muscle fibers, immune complexes of immunoglobulins and complement C3 in the perimysial venules, and MAC (membrane attack complex of complement) neoantigen have been reported. And more recently, changes in the vascular components were proven to be the primary lesion in this disease because microtubular inclusions and microvacuoles in endomysial capillaries and MAC deposits with an anti-MAC antibody were recognized even in otherwise normal muscle of patients with DM (1, 2).

It is well known that complications in organs or tissues other than muscle and skin are frequent in this disease, and such lesions have also been attributed to vascular pathology. In this issue of the Journal, Matsuda et al reported a case with DM accompanied by splenic and renal infarction (3).

As the authors indicated, these complications have been considered to be very rare especially in adults, but this combination is not likely coincidental even though administration of adrenal cortical steroids played some role in evoking such infarctions. Such cases might suggest a possibility that a more generalized autoimmune mechanism involving the vascular system is present in DM. The demonstration of circulating antibody to human umbilical vein endothelial cells in patients with DM associated with interstitial lung disease (4) might be one of support for such possibility.

The reason why blood vessels in the muscular and cutaneous tissues are selectively involved in this disease is of course obscure. The complexity of the clinical features of autoimmune disorders might be explained by the specificity of autoantigens together with genetic factors of the individual patient. For example, in autoimmune peripheral neuropathies, motor, sensory, or autonomic nerves are independently attacked. In addition, such selectivities are not only correlated with types of nerve fibers, but sometimes localizations are also specific. In some types, cranial nerves are involved, and in other types brachial plexus fibers are involved. Corresponding to such clinical differences, specific autoantibodies have been found in certain subtypes of such disorders.

More frequent routine usage of MRI, CT or other methods may disclose the more frequent presence of such cases as described by Matsuda et al (3), and further studies should focus on investigations of the specificity of antigenic polypeptides in individual patients.

See also p 512.

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References