Editorial

Anticentromere Antibodies and Vascular Diseases

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Anti-centromere antibodies (ACA) were discovered by Moroi et al (1) in 1980 as a specific antibody for CREST syndrome, a variant of systemic sclerosis (SSc) characterized by calcinosis, Raynaud’s phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia. ACA are autoantibodies to proteins that localize to a distinct chromosomal domain known as the centromere. In addition to the three major centromere proteins (CENPs A, B and C) recognized by most sera with ACA, recently, a centromere kinesin-like protein, CENP-E, was recognized as an antigenic target in SSc (2). CENP B fusion proteins are now used in an enzyme-linked immunosorbent test for the detection of ACA. Subsequently, it was found that these antibodies are occasionally present in a number of other rheumatic diseases, including rheumatoid arthritis, systemic lupus erythematosus (SLE) and primary Sjögren’s syndrome, and primary biliary cirrhosis in the context of CREST syndrome (3).

Furthermore, recent reports have described the presence of ACA in the sera of patients with idiopathic Raynaud’s phenomenon, gangrene in the extremities with mild or no scleroderma (4–6) and primary pulmonary hypertension (7). These findings suggest a strong correlation between ACA and Raynaud’s phenomenon (8), and an association of ACA with severe vascular damage.

In this issue of the Journal (9), Kitamura et al report an interesting case of phlebosclerosis of the colon with positive ACA and Raynaud’s phenomenon. Phlebosclerosis is a very rare disease characterized by ileus and the remarkable calcification of the sclerotic walls of the colic veins.

See also p 416.

In the available literature, all patients of such disease have been Japanese. The histological feature of the colon was a marked increase of collagen in the submucosa and fibrous replacement in the muscle layers of the colon, which were similar to those of SSc. The authors speculated that the pathogenesis of the phlebosclerosis of colon is related to CREST syndrome, based on the presence of Raynaud’s phenomenon and ACA. Immunological studies were not done in the other case reports of this disease. But a fibrosis colon may be the result of ischemic colitis due to occlusion of veins (10).

The disease association of ACA is not only with CREST syndrome, but also with vascular diseases including Raynaud’s phenomenon. Phlebosclerosis of the colon may be a new disease association of this antibody.

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References