AUTOIMMUNE CONNECTIVE TISSUE DISEASES

VASCULAR ABNORMALITIES IN SYSTEMIC SCLEROSIS

Yoshihide Asano [(1)]

University of Tokyo Graduate School of Medicine, Dermatology, Tokyo, Japan [(1)]

Systemic sclerosis (SSc) is a multisystem autoimmune disease characterized by vasculopathy and fibrosis of the skin and various internal organs. Although the pathogenesis of SSc still remains elusive, it is generally accepted that initial vascular injury due to autoimmunity and/or environmental factors causes structural and functional abnormalities of vasculature which eventually result in the constitutive activation of fibroblasts in various organs. Structural alterations consist of destructive vasculopathy (loss of small vessels) and proliferative obliterative vasculopathy (occlusion of arterioles and small arteries with fibro-proliferative change) caused by impaired compensatory vasculogenesis and angiogenesis. Impaired function of SSc vasculature includes the altered expression of cell adhesion molecules predominantly inducing Th2 and Th17 cell infiltration, endothelial dysfunction primarily due to the low availability of nitric oxide, the activated endothelial-to-mesenchymal transition leading to fibro-proliferative vascular change and tissue fibrosis, and the impaired coagulation/fibrinolysis system promoting the formation of intravascular fibrin deposits. Clinically, the evaluation of vascular structure and peripheral circulation by nailfold video capillaroscopy is useful to evaluate the severity and activity of SSc as well as the efficacy of treatments. In my talk, I would like to overview the pathogenesis of SSc vasculopathy and its significance in the clinical setting.