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Aortic thrombi are important because potentially it can cause the central and peripheral embolizations. Aortic thrombi can occur anywhere in the aorta but extremely rare in ascending aorta without atherosclerosis, aneurysm, or cardiosurgical or traumatic state. Systemic sclerosis (SSc) is an autoimmune disorder of connective tissue and it can involve multisystem. Enhanced coagulation pathways, decreased fibrinolysis, and endothelial dysfunction probably contribute to vascular events in SSc. Especially Raynaud phenomenon and microcirculatory abnormalities are well recognized but macrovascular disease is not common. We report a case of a highly mobile thrombus in the ascending aorta, presented with an acute embolic stroke in the patient with SSc. Surgical removal was performed because it was refractory to anticoagulation. In histopathological examination, a organized thrombus without evidence of infectious organism or malignancy was revealed. The wall of the ascending aorta demonstrated no evidence of atherosclerosis or inflammatory change.

