KSN 2021
FULLY VIRTUAL MEETING
September 02 (Thu) - 05 (Sun)

Abstract Submission No. : 9146

Experience with Thrombotic Microangiopathy over the past 20 years in one institution

Yeong-Jin Choi
The Catholic University of Korea, Seoul St. Mary's Hospital, Korea

Thrombotic microangiopathy (TMA) is the pathologic term for a condition characterized by microvascular changes including thrombosis, microangiopathic hemolytic anemia (MAHA) and thrombocytopenia.

The pathogenesis of TMA is explained by uncontrolled C' activation, platelet- rich and fibrin-rich thrombi formation together with platelet consumption, endothelial swelling, vascular occlusion which lead to mechanical non-immune intravascular hemolysis and finally Ischemic organ injury.

The diagnosis of TMA can be established by clinical findings and/or pathologic findings. TMA can present clinically as the heterogeneous diseases with a wide range of symptoms and conditions due to its systemic nature. The classification of TMA is quite complex, and the clinical recognition of various disease categories at the onset of the disease is often difficult or impossible. Limited clinical experience and confidence due to the rarity of TMA cases, TMA may be diagnosed first by pathologist with renal biopsies, and it could be a big clue for clinicians starting to suspect TMA and closely monitor the patient. Therefore, an accurate and active pathological diagnosis of TMA is required.

The basic pathologic abnormalities of TMA are endothelial damage, intimal thickening, coagulation both within the vessel wall and the vascular lumina. Histological confirmation is not mandatory to establish a diagnosis of TMA, since histological findings of TMA are not pathognomonic for diagnosing each etiology. However histological confirmation is useful, when clinical presentation is uncommon, as well as to determine prognosis. The most recommended biopsy site is the skin, gastrointestinal tract and gingiva, regardless of the presence of visible lesion. A kidney biopsy is the gold standard for diagnosing TMA in the kidney. When you diagnose the kidney biopsy slides, the recognition of TMA is often difficult, especially cases without thrombi formation.

When TMA is suspected, evaluation for etiological diagnosis should be initiated. Recently the evaluation for the complement regulatory proteins is frequently required. Testing for

mutations of the complement related factors or the presence of autoantibodies are necessary. Due to the introduction of new antibody treatment modality including Eculizumab, the early implementation of etiological treatment can change the natural course of serious conditions.

Here, we analyzed and evaluated the pathologic and clinical findings of TMA cases experienced at our institution over the past 20 years. We would like to discuss in particular the pathological aspects of heterogeneous cases of TMA and TMA associated with other renal diseases.