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Diagnosis and management of TMA syndrome

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Thrombotic microangiopathy is a syndrome characterized by the presence of microangiopathic hemolytic anemia, thrombocytopenia, and organ injury. Kidney is the most frequently injured organ in this syndrome. In this lecture, we will review the clinical presentation and pathophysiology, and diagnostic approach and empirical and disease specific treatment strategies of TMA syndrome. Firstly, we will preview the cases of two patients who presented with TMA syndrome. This was a case of a patient with typical HUS and secondary TMA by anti-phospholipid syndrome. Peering at actual patients' cases, we will see clinical presentation of TMA and treatment process. In perspective of pathophysiologic view, organ injuries by TMA syndrome result from endothelial dysfunction followed by intravascular thrombosis. Subsequent tissue ischemia and organ injury can be induced. Complement over-activation is thought to play an important role in pathophysiology. TMA syndrome can be classified based on primary and secondary causes. Primary TMA syndromes comprise thrombotic thrombocytopenic purpura (TTP), typical HUS caused by shiga toxin-producing Escherichia coli (STEC-HUS), and atypical HUS (aHUS). We will discuss how to reach a diagnosis and treatment strategies for each category. Especially in completed mediated atypical HUS, targeted therapy for C5 inhibitor, eculizumab and ravulizumab, has been introduced. We can meet TMA syndrome in various life-threatening conditions. Much more investigations should be done to clarify the pathogenesis, but endothelial injury and complement dysregulation is central features of TMA syndromes. Prompt diagnosis and specific management is crucial for adopting highly targeted therapy.

Keywords: TMA syndrome, endothelial injury, hemolytic uremic syndrome