Thrombotic microangiopathy (TM) is characterized by the presence of thrombocytopenia and microangiopathic hemolytic anemia. There are different pathologies of various etiologies, hereditary or acquired, which causes said microangiopathy. A prompt diagnosis is of the utmost importance in order to begin treatment immediately, since this pathology is considered a medical emergency.

Classic TM, thrombocytopenic thrombotic purpura (TTP), is caused by a severe ADAMTS-13 deficiency (disintegrin and metalproteinase), acquired mostly by autoantibodies, this enzyme normally proteolizes native von Willebrand factor immediately after being synthesized in the vascular endothelium generating the normal multimers, which are the physiological active molecules leading to platelet adherence and aggregation as part of normal thrombus formation. Atypical hemolytic uremic syndrome (aHUS) is very similar to TTP, although it is linked to genetic defects.\(^1\) In the last decade, management, diagnosis and treatment of these pathologies have evolved notably. Téllez-Hinojosa et al.\(^2\) elegantly analyzes classic TM etiopathogenesis, diagnosis and treatment in this issue of Medicina Universitaria. It is very important to insist that a timely diagnosis is mandatory in order to proceed to a rapid therapeutic intervention (plasma exchange), which can be life-saving. In this respect, the examination of a well-stained peripheral blood slide by an experimented hematologist is critical in establishing the presence of this hematological emergency, requiring a multidisciplinary therapeutic approach.

Refractory TTP treatment has evolved considerably; the role of splenectomy in patients who do not respond to rituximab has been described with a relapse-free survival rate of 70% at 10 years. The use of bortezomib, cyclosporine, cyclophosphamide, vincristine, eculizumab and N-acetylcysteine have been reported in some refractory cases as multiple first and second line therapies, with different responses.\(^3\)

TTP diagnosis will have to be more precise. ADAMTS-13 activity determination has been relevant in the study of TTP pathogenesis; however, it does not identify all patients who benefit from plasma exchange.\(^4\) The treatment will have to be more effective, more accessible and safer. Survival rate has not improved substantially since the introduction of plasmapheresis, which is technically complex, expensive and may cause complications. Significant advances have been made in the study of TTP; nevertheless, there is still much more to be accomplished.

References


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