

TABLE 1. Differential diagnosis of thrombotic microangiopathies (TMAs)

Diagnostic entity	Pathophysiology	Laboratory Diagnosis
Immune-mediated TMA or Thrombotic thrombocytopenic purpura (TTP)	ADAMTS13 deficiency	ADAMTS13 activity
Complement-mediated hemolytic uremic syndrome (HUS)	Complement dysregulation	No gold standard (several assays suggesting complement activation, ie soluble C5b-9, plus genetic testing)
Infection-associated TMA	Shiga-toxin, Campylobacter jejuni, Streptococcus pneumonia, Human immunodeficiency virus, Cytomegalovirus, Epstein–Barr virus, Parvovirus B19, BK virus, Influenza	Detection of infectious agents
Disseminated intravascular coagulation	Abnormal coagulation, Underlying cause	No gold standard (based on risk scores)
Secondary TMA	Cancer, Transplantation, Systemic lupus erythematosus, Antiphospholipid antibody syndrome, Scleroderma, Vasculitis/glomerulonephritis	Diagnosis of the underlying entity
Malignant hypertension-induced TMA	Malignant hypertension	Hypertension levels
Drug-induced TMA	Calcineurin or mTOR inhibitors, Quinine Estrogen/progesterone, Gemcitabine/mitomycin C. Interferon Vascular endothelial growth factor or proteasome inhibitors, Cocaine	Based on history and exclusion of other entities
Metabolism-associated TMA	Cobalamin responsive methylmalonic acidemia, Diacylglycerolkinase epsilon mutation	Methylmalonic acid, genetic testing
Pregnancy-associated TMA	HELLP (hemolysis, elevated liver enzymes, and low platelets) syndrome, HUS, TTP	According to each syndrome (see above), HELLP diagnostic criteria