Differential diagnosis for primary TMAs: atypical HUS, TTP, and STEC-HUS

Thrombocytopenia
Platelet Count <150,000/mm³ or
>25% Decrease from Baseline

AND

Microangiopathic Hemolysis
Schistocytes and/or
Elevated LDH and/or
Decreased Haptoglobin and/or
Decreased Hemoglobin

Plus one or more of the following:

Neurological Symptoms
Confusion and/or
Seizures and/or
Other Cerebral Abnormalities

Renal Impairment
Elevated Creatinine and/or
Decreased eGFR and/or
Elevated Blood Pressure and/or
Abnormal Urinalysis

Gastrointestinal Symptoms
Diarrhea ± Blood and/or
Nausea/Vomiting and/or
Abdominal Pain and/or
Gastroenteritis

Cardiovascular Symptoms
Myocardial Infarction and/or
Hypertension and/or
Arterial Stenosis and/or
Peripheral Gangrene

Pulmonary Impairment
Dyspnea and/or
Pulmonary Hemorrhage and/or
Pulmonary Edema

Visual Symptoms
Pain and Blurred Vision
Retinal Vessel Occlusion
Ocular Hemorrhage

Evaluate ADAMTS13 Activity and Shiga-toxin/EHEC* Test

While waiting for ADAMTS13 results, a platelet count of >30,000/mm³ and/or a serum creatinine level >150-200 μmol/L almost eliminates a diagnosis of severe ADAMTS13 deficiency (TTP)

≤10% ADAMTS13 Activity

>10% ADAMTS13 Activity

TTP

atypical HUS

Shiga-toxin/EHEC Positive

STEC-HUS*

- Genetic mutations are not identified in 30% to 50% of patients with atypical HUS
- A diagnosis of atypical HUS does not require identification of a mutation

The availability of an effective treatment option warrants a differential diagnosis of atypical HUS.

*Shiga-toxin/EHEC test is warranted in history/presence of GI symptoms.

The information on this page is intended as educational information for healthcare providers. It does not replace a healthcare professional's judgment or clinical diagnosis.

References: