Thrombo-Guide
vom Labor zur Diagnose

Thrombotic Microangiopathies (TMA
Thrombotic Thrombocytopenic Purpura (TTP
Hemolytic Uremic Syndrome (HUS)

Description:

Thrombotic microangiopathies comprise acute, life-threatening syndromes characterized by thrombocytopenia, microangiopathic hemolytic anemia (non-immune mediated), and diffuse multi-organ dysfunction. Pathophysiologically, they are classified as follows:

- **Thrombotic Thrombocytopenic Purpura (TTP):** Severe deficiency of ADAMTS13 activity, either immune-mediated (acquired) or congenital.
- **Complement-Mediated TMA:** (Historically termed atypical hemolytic uremic syndrome [aHUS]).
- Infection-Associated TMA: Driven by gastrointestinal infections presenting with diarrhea (STEC-HUS), viral infections, parasites, etc.
- **Pregnancy-Associated TMA:** HELLP syndrome, gestational toxicosis, preeclampsia/eclampsia, etc.
- **Secondary TMAs:** Triggered by medications, malignancies, systemic autoimmune diseases, malignant hypertension, hematopoietic stem cell or solid organ transplantation, etc.

Diagnostics:

When a TMA is suspected, a systematic differential diagnostic evaluation must be initiated immediately (refer to specific guidelines). Rapid determination of ADAMTS13 activity is critical to guide emergent therapeutic decision-making.

Therapeutic Options:

- Immune TTP (severe ADAMTS13 deficiency accompanied by a detectable inhibitor) is managed with caplacizumab, rituximab, and therapeutic plasma exchange (TPE) (consult specific literature).
- Congenital TTP (severe ADAMTS13 deficiency caused by genetic mutations, without an inhibitor) is treated with recombinant ADAMTS13 concentrate.
- Complement-mediated TMA is treated with complement inhibition (eculizumab, ravulizumab, or novel agents) alongside renal replacement therapy as clinically indicated.
- In Secondary TMAs, management focuses primarily on treating the underlying primary medical condition.

References:

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