Is it thrombotic microangiopathy (TMA)?

Consider TMA in lupus patients with thrombocytopenia, microangiopathic hemolytic anemia, and organ involvement[1-5]

TMA can be associated with numerous conditions and triggers[1]

Definition of TMA[1,2]

Thrombocytopenia

AND

Microangiopathic hemolytic anemia

Schistocytes and elevated LDH and decreased haptoglobin and decreased hemoglobin

Plus 1 or more of the following signs and symptoms:

Organ manifestations

Common signs and symptoms

- **Neurological symptoms**
- **Renal impairment**
- **GI symptoms**

Other

- **CV symptoms**
- **Pulmonary symptoms**
- **Visual symptoms**

Evaluate ADAMTS13 activity and Shiga toxin/EHEC test[a]

While ADAMTS13 results are awaited, a platelet count >30 x 10^9/L and/or sCr >1.7 to 2.3 mg/dL almost eliminates a diagnosis of severe ADAMTS13 deficiency (TTP)

≤5%[b] ADAMTS13 activity	>5% ADAMTS13 activity	Shiga toxin/EHEC positive
TTP	Atypical-HUS[c]	STEC-HUS

[a]Shiga toxin/EHEC test is warranted with history/presence of GI symptoms. [b]Range found in published literature is <5%-10%. [c]Where TTP and STEC-HUS have been ruled out, the TMA can be attributed to complement dysregulation. Atypical-HUS is a complement-mediated TMA (CM-TMA) that has been more broadly or narrowly defined in different contexts, based on such factors as the presence and nature of a trigger and/or identification of an underlying genetic mutation. [d]Many other conditions may trigger atypical-HUS.

TMA in patients with lupus—it could be atypical-HUS

If TMA is suspected, time is of the essence

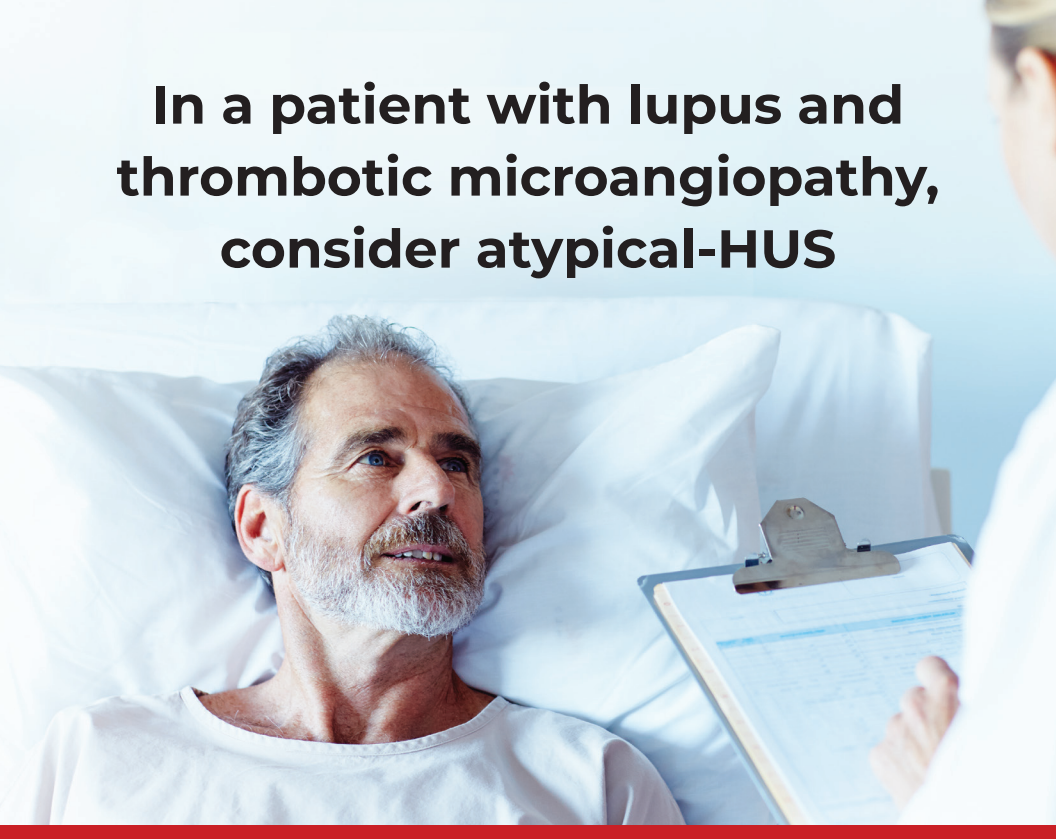
Atypical-HUS:

- is a serious, life-threatening medical disorder that can manifest with or be triggered[d] by SLE[1,3,6-9]
- is associated with complement dysregulation that leads to microvascular thrombosis, hemolysis, and ischemic organ injury[1,2,10,11]
- may be a complication of lupus[2,7,9]

Differential diagnosis of TMA[1,5,10-12]
SIGNS OF TMA
- Platelet count, blood smear showing schistocytes, signs of organ involvement, elevated LDH
TRIGGERS OF TMA
- Autoimmune disorders (eg, SLE, catastrophic antiphospholipid syndrome) Associated tests: antiphospholipid, LAC
- Cobalamin C deficiency Associated tests: B12, homocysteine, methylmalonic acid/methionine testing, *MMACHC* mutation screening
- DIC Associated tests: PT/aPTT, D-dimer
- Infection Associated tests: Shiga toxin panel, COVID-19, other infection panels
- TTP Associated test: ADAMTS13 activity level
- Other triggers (eg, pregnancy, genetic mutation)
- Atypical-HUS There is no single definitive test. Diagnosis is exclusionary

ADAMTS13=a disintegrin and metalloproteinase with a thrombospondin type 1 motif member 13; CV=cardiovascular; DIC=disseminated intravascular coagulation; EHEC=enterohemorrhagic *Escherichia coli*; GI=gastrointestinal; HUS=hemolytic uremic syndrome; LAC=lupus anticoagulant; LDH=lactate dehydrogenase; *MMACHC*=methylmalonic aciduria and homocystinuria type C; PT/aPTT=prothrombin time/activated partial thromboplastin time; sCr=serum creatinine; SLE=systemic lupus erythematosus; STEC=Shiga toxin–producing *Escherichia coli*; TMA=thrombotic microangiopathy; TTP=thrombotic thrombocytopenic purpura.

In a patient with lupus and thrombotic microangiopathy, consider atypical-HUS



In patients with lupus and persistent TMA, a timely diagnosis of atypical-HUS can help with necessary and specialized atypical-HUS disease management.[1,13]

Learn more about TMA and atypical-HUS at **aHUSSource.com/physician**

References

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