

Acquired Thrombotic Thrombocytopenic Purpura in Pregnancy : A Case Report

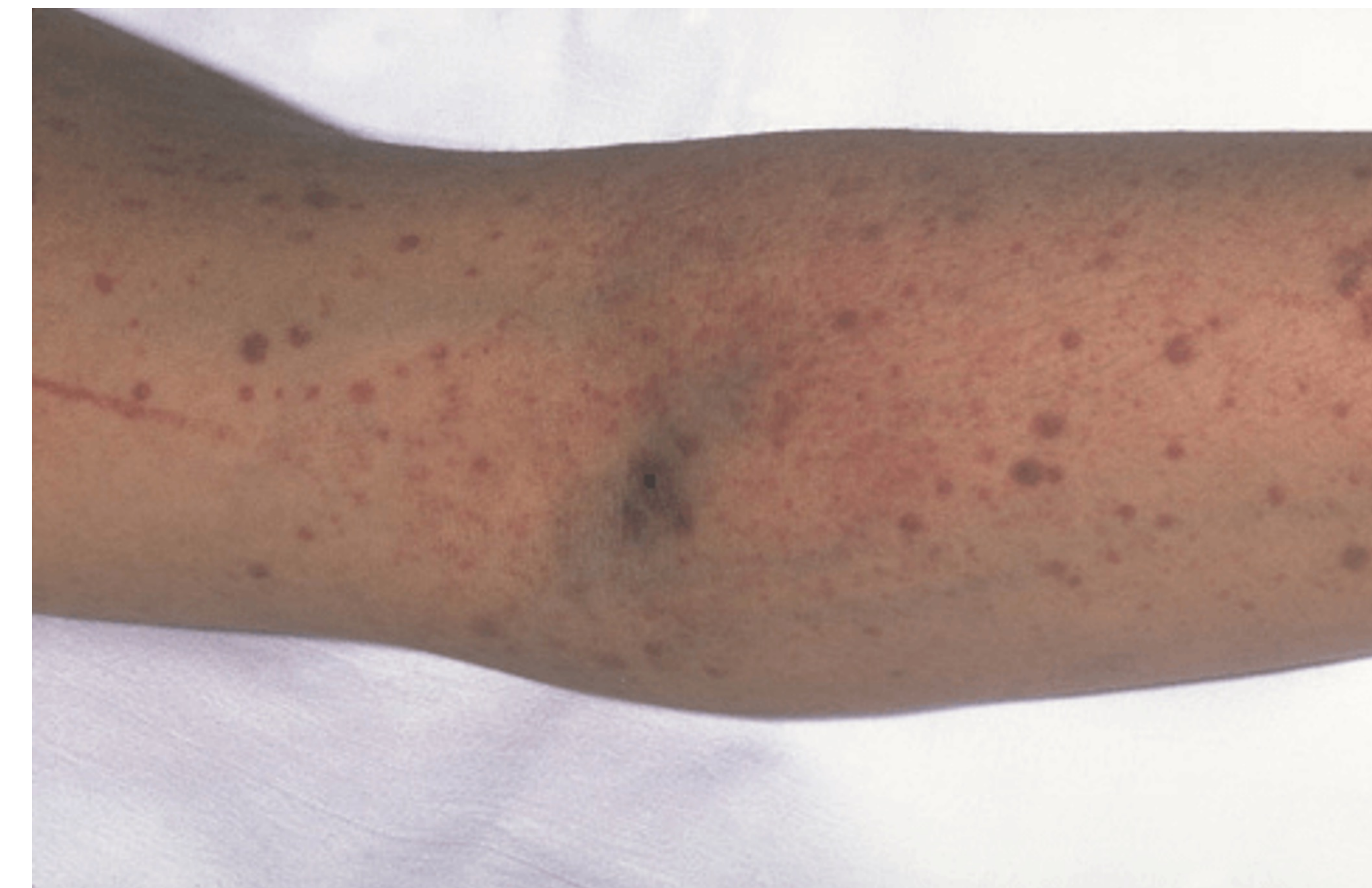
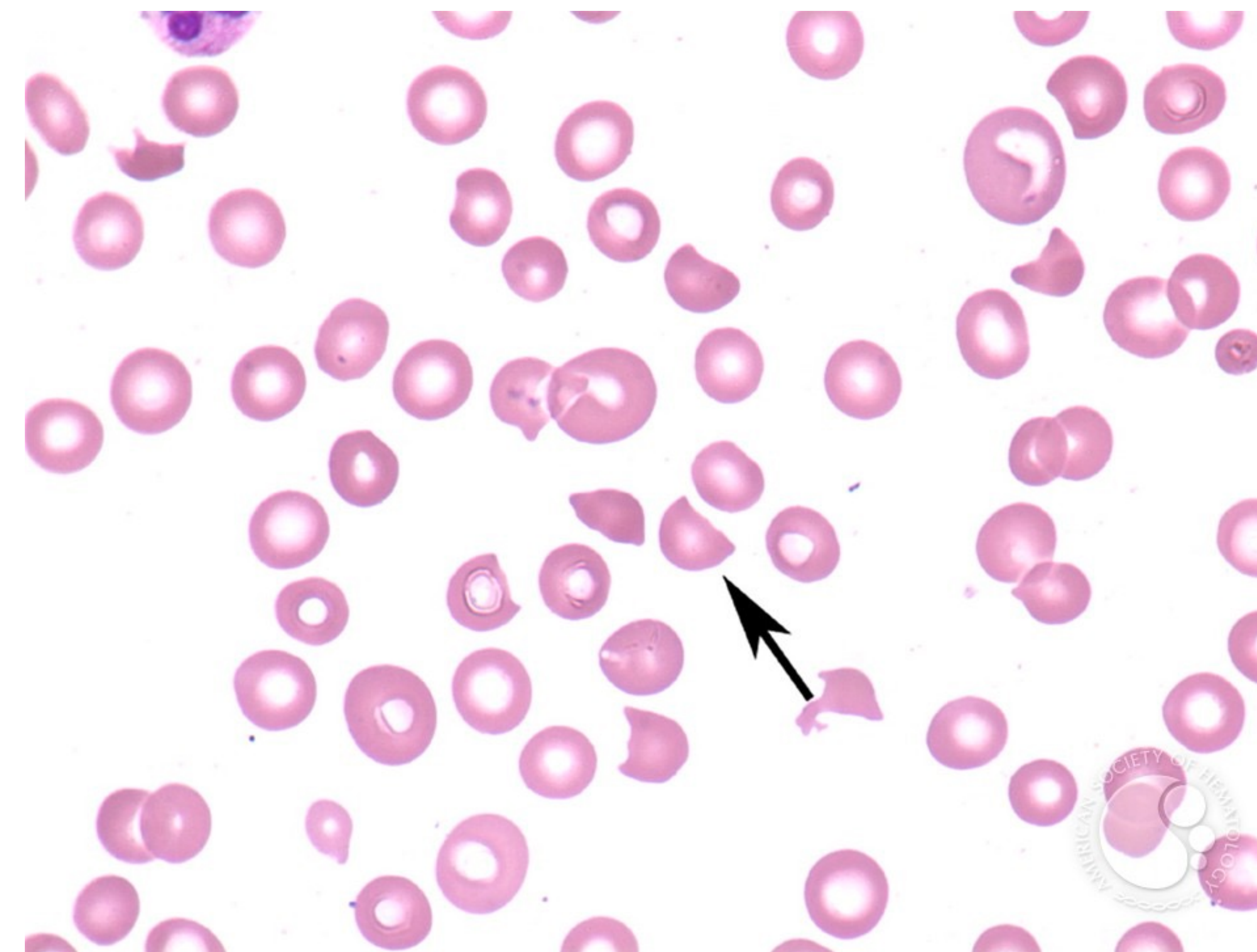
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Background

- Thrombocytopenia affects 10% of parturients¹
- Causes include gestational thrombocytopenia, HELLP syndrome, thrombotic microangiopathies
- Acquired TTP is a thrombotic microangiopathy and a rare cause of thrombocytopenia in pregnancy²
- aTTP is caused by an auto-antibody against ADAMTS13 leading to large vWF multimers increasing platelet aggregation and thrombosis

Presenting Signs and Symptoms

- Skin changes
- Acute renal failure
- Thrombocytopenia
- Fever
- Neurologic abnormality



Case Presentation

30-year-old G3P2002 at 28w0d presented with facial numbness and intermittent paresthesias

28w0d

Labs:
Hgb 7.0, Plts 36k
Elevated LDH
Schistocytes on smear

aTTP suspected, PLEX and steroids initiated

28w6d

Plts <10k and she was deemed refractory to standard therapy

aTTP confirmed with ADAMTS13 activity <5%,

Patient agrees to trial of caplacizumab

29w2d

Plts 165k

33w0d

Non-reassuring fetal status prompted urgent cesarean delivery

Plts 418k
Theoretical increased bleeding risk with caplacizumab

PPD3

Cesarean delivery under GA

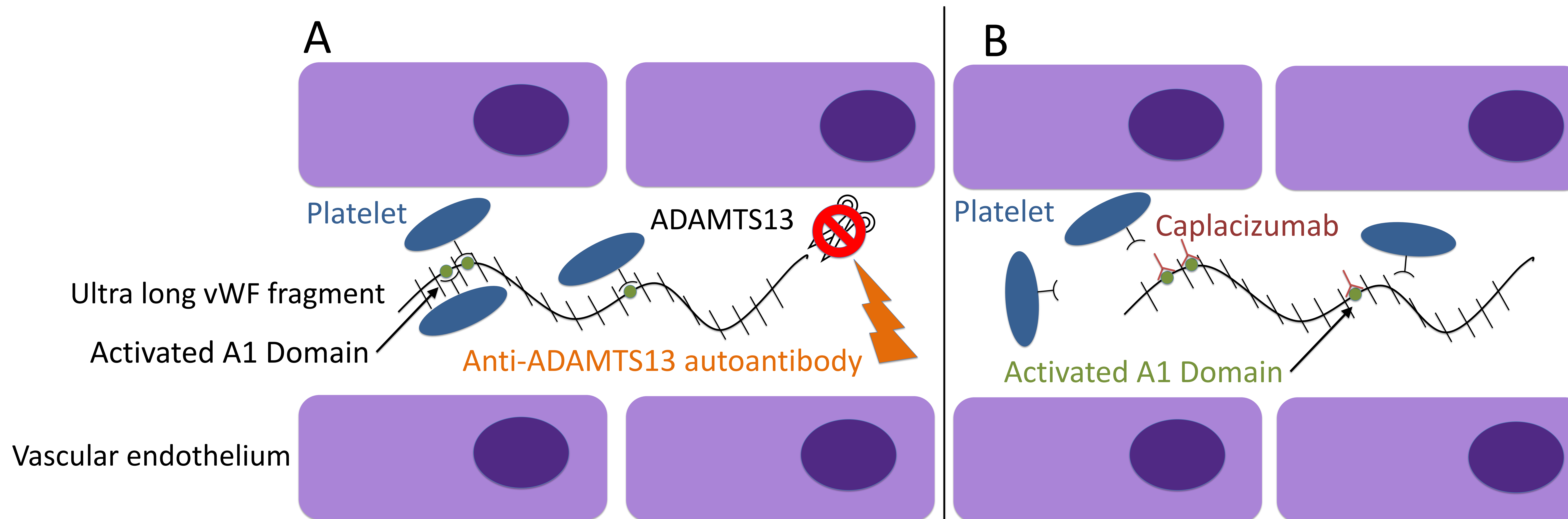
No deleterious effects to mother or fetus were apparent

Discussion

- Standard aTTP treatment in pregnancy is replenishment of ADAMTS13 with immune modulation including plasma exchange and steroids³
- If refractory, Caplacizumab, a humanized bivalent immunoglobulin fragment can be considered

Caplacizumab

- Targets the A1 domain of VWF
- Blocks interaction between VWF and platelet glycoprotein Ib-IX-V preventing platelet aggregation and thrombosis
- Shown to be efficacious in aTTP refractory to standard treatment and severe disease⁴
- Minimal data on use in pregnancy⁵



References

1. Reese (2018) NEJM 379:32-43
2. Brown (2022) Ann Hem 101:2159-2167
3. Shah (2013) Br J Haem 163:514-519
4. Dutt (2021) Blood 137:1731-1740
5. Kühne (2022) Br J Haem 196:e30-e33