A Refractory Case of Catastrophic Antiphospholipid Syndrome (CAPS)

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Background
- Catastrophic Antiphospholipid Syndrome (CAPS) is a rare, life-threatening form of Antiphospholipid Syndrome (APS) characterized by severe thrombotic complications of the microvasculature & large vessels.
- CAPS is typically treated with a combination of anticoagulation, glucocorticoids, & Therapeutic Plasma Exchange (TPE) or Intravenous Immune Globulin (IVIG).

Refractory cases and treatment
- Rituximab (RIT) or Eculizumab
- Anti-CD 38 trials use ongoing (availability is a limiting factor).

Uniqueness
- Can be refractory to all the available treatment options.
- DDx: VEXAS (Vacuoles, E1 enzyme, X-linked Autoinflammatory, Somatic) syndrome, relatively new.

Case Description
- 40-year-old male presented with calf pain, chest pain and dyspnea.
- Imaging - DVT of the right lower extremity and Pulmonary Embolism (PE).
- Treated with Apixaban (stopped due to rash) and switched to Enoxaparin (EN).
- Initial hypercoagulable workup – indeterminate Lupus Anticoagulant (LA) and everything else normal. Repeat LA + so started on Warfarin.
- 2 months later developed new DVT & PE while therapeutic on Warfarin, so switched back to EN.

3 months later and serious sequence of events afterwards
- Hospitalization for new PE. Heparin & then Rivaroxaban.
- Readmitted within a day for multi-territorial acute brain infarct. Treated with Fondaparinux, Prednisone, Hydroxychloroquine, TPE, & RIT.
- Did respond to TPE initially.
- Readmitted after 1 month with loss of vision due to ischemic strokes involving bilateral occipital lobes (Fig. 2). Treated with Fondaparinux, IVIG, TPE, Prednisone, Mycophenolate Mofetil (MMF), Eculizumab.
- Developed more multifocal embolic strokes, splenic/renal infarcts, RV thrombus, severe thrombocytopenia PLT <30 (PF4-), low fibrinogen <50.
- Fondaparinux temporarily held.
- Romiplostim used to improve PLT counts.
- Cryoprecipitate given once to improve fibrinogen level in preparation to re-start anticoagulation (Argatroban).

Outcomes
- Within 2 days of starting Argatroban developed globular hemorrhagic transformation (Fig. 3) of prior stroke and inferior STEMI.
- Timely initiation of heparin was challenging.
- Heparin started after 2 days.
- Respiratory status progressively worsened.
- Family decided to transition to comfort care.
- Passed away within 6 months of diagnosis.

Discussion/Conclusion
- More research is needed to explore treatment options for refractory cases of CAPS.
- Evaluation for possible triggers require multidisciplinary approach.

Challenges:
- Minimally invasive workup (to evaluate possible trigger) can be difficult due to concomitant coagulopathy.
- Rare disease so lack of data.

References