Catastrophic Antiphospholipid Syndrome Complicated with Hepatic Budd-Chiari Syndrome and Inferior Vena Cava Occlusion. Case report of successful management with endovascular sharp IVC recanalization and direct intrahepatic porto-systemic shunting

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Disclosure

I do not have any potential conflict of interest
Purpose

To describe combined sharp needle-assisted recanalization of the IVC and Direct intrahepatic portosystemic shunt treatment for a case of Bud Chiari syndrome with IVC occlusion in a complicated catastrophic antiphospholipid syndrome.
Catastrophic Antiphospholipid Syndrome

- Fatal variant of antiphospholipid syndrome (50%).

- Systemic autoimmune disease characterized by life threatening multiorgan thromboses developing over a short period.
Catastrophic Antiphospholipid Syndrome

Severe form of APS characterized by:

- Multiple organ involvement over a short period of time.
- Histopathologic evidence of small vessel involvement.
- Laboratory confirmation of aPL.

Our Patient

- 14 year old female.
- She was diagnosed on April 2017 with Catastrophic Antiphospholipid syndrome.
Previous Workup

• Mild Pericardial Effusion and elevated Troponin level.
• Positive Lupus Anticoagulant Beta 2 Glycoprotein and Inflammatory Markers.
• Liver biopsy: ischemic changes (infarction) and intravascular thrombi.

Enoxaparin (LMWH) 50 mg S/C every 12 hours
Hospital admission

- Presented on Jun 2018 with Epigastric pain of 1 day duration referred to right upper quadrant.
  - Progressive pain with increase intensity stabbing nature (aggravated by eating).
  - Off medication by parent since 3 months.
Problems

✓ Short segment of “hepatic” IVC occlusion.

✓ Hepatic vein occlusion.
Next day
Follow on November 2018

✓ No Pain.
✓ No vomiting.
✓ No tenderness.
✓ QoL of the patient improves.
✓ Comparing his previous symptomatic issues there is remarkable improvement.
Thank you
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