Shockingly, It’s Not Just Shock!

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Learning Objectives

1. Identify atypical shock presentations that may indicate underlying Capillary Leak Syndrome (CLS).
2. Explain the potential complications of aggressive fluid resuscitation in undifferentiated shock and justify the need for a cautious approach to fluid management.

Conflicts of Interest

None to declare

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Patient Presentation

30 yo female presenting in undifferentiated shock

- PMHx: from home alone, healthy, no prior medical history, no home meds
- HPI: 24h of altered LOC, severe vomitting, hypothermia (34.4C), hypotensive (BP 59/42), in refractory shock
- Investigations: Metabolic acidosis (pH 6.98, lactate 11), leukocytosis (32.8), with elevated Hgb (226, HCT 70%), low albumin (19)
- Initial management: Empirically started on antibiotics (Piperacillin-Tazobactam, Vancomycin, Clindamycin) for suspected septic shock, aggressive fluid resuscitation (10L ringers lactate), brought to ICU requiring 3 vasopressors

Hospital Course

Prolonged hospital course:

- Developed significant peripheral edema from third spacing
- Resulted in occlusive thrombus of the right cephalic vein and bilateral lower extremity compartment syndrome, requiring emergency bilateral fasciotomies and initiation of therapeutic anticoagulation with enoxaparin
- 10 days later developed new skin ecchymosis, thrombocytopenia, and left leg DVT, confirmed on HIT assay to have Heparin Induced Thrombocytopenia
- Extended hospitalization for stabilization and rehabilitation (~1.5 months)

Work-up

Refractory shock in the absence of a true source?

- Given her refractory hypotension in the absence of a true septic source, unusually elevated Hgb, and unusual sequelae of events post aggressive fluid resuscitation, the team looked into alternative etiologies for her condition
- In the context of her **hypotension, hypoalbuminemia,** and **hemoconcentration,** she was diagnosed with **Capillary Leak Syndrome (CLS):** a rare disorder characterized by increased vascular permeability, leading to plasma leakage into interstitial spaces.
- Hematology was consulted, and she underwent testing for triggers of CLS including:
  - Sepsis/toxic shock: covid and influenza positive but blood, urine, vaginal cultures all negative.
  - HLH: no bicytopenia, hepatosplenomegaly, or excessive ferritin elevation
  - Genetic: JAK2 negative, hereditary angioedema (C1 esterase negative, complements normal)
  - Monoclonal gammopathy: immunoglobulins low - started on IVIG, SPEP normal
  - Hematologic malignancy: BCR-ABL negative, no lymphadenopathy
  - Autoimmune: dsDNA, ANCA, RF, myositis panel all normal
- Thus, it was felt that her CLS was triggered virally by Covid and Influenza vs idiopathic (Clarkson’s Disease)

Management

3 Phases of Treatment

- Acute phase: fluid resuscitation to counter initial hypovolemia. Patient initially received 10L of ringers lactate + Vasopressors over the course of 3 days

- Recovery Phase: Aggressive Diuresis to remove excess fluid. Patient was maintained on regular Furosemide 40mg IV daily x 1 week to remove excess fluid

- Prophylaxis: Patient was found to have low immunoglobulins and hence was given IVIG.

Discussion

Learning Points

- This case highlights the critical importance of recognizing CLS, particularly in patients who present with atypical/unexplained presentations of hypotension.

- Aggressive fluid resuscitation, while standard of practice for hypotension, can exacerbate and lead to severe complications such as compartment syndrome.

- Optimal fluid resuscitation in CLS requires a tailored approach to volume replacement, close hemodynamic/ volume monitoring, and adjunctive therapies.