Why so Shocky?? A case of refractory shock

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Why so Shocky??
A case of refractory shock

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Written consent was obtained from patient representative and PSJF HRPP has determined that this case report, as submitted, does not meet the definition of research and does not require IRB review as defined in the federal regulations.
69-year-old female who was transferred to the ICU for shock following several hospitalizations for hypotension, fevers, weakness, fatigue, loose stools and night sweats.
Prior workup

- Unexplained hyponatremia
- Cholestatic transaminitis
- Pituitary microadenoma
- Euthyroid sick syndrome

- Negative for adrenal insufficiency
- Negative extensive infectious workup
- Negative for amyloid, leptomeningeal carcinomatosis, paraneoplastic and rheumatological processes
ICU work up

- Pulmonary artery catheter evaluation
  - normal cardiac function
  - highly fluid responsive state despite aggressive resuscitation
Case continued...

- Progressive lactic acidosis and refractory shock despite multiple vasoconstrictive medications, empiric antibiotics and trial of pulse dose steroids for potential inflammatory process
A multidisciplinary subspecialty evaluation including Infectious Disease, Endocrinology, General Surgery, Neurology, Rheumatology, Pathology, Hematology and Oncology yielded no definitive answers...
Tragically our patient’s diagnosis was not confirmed until postmortem evaluation.
Microscopic evaluation on autopsy revealed a large B-cell neoplasm involving the intravascular spaces within nearly all the organ systems.
Intravascular large cell lymphoma (ILCL)

- Rare subtype of large cell lymphoma AKA
  - intravascular lymphomatositis
  - angiotropic large cell lymphoma
  - malignant angioendotheliomatosis

- Characterized by lymphoma cell proliferation within the lumina of capillaries and post-capillary venules.
Typically requires deep incisional skin biopsies with immunophenotyping pathology evaluation but is often a postmortem diagnosis.

As the disease progresses, capillary system involvement can produce a profound distributive shock and end organ involvement refractory to typical therapies.
Intravascular large cell lymphoma is a rare disease with high mortality requiring increased recognition.

Importance of maintaining a broad differential in complex multidisciplinary cases.

Tissue biopsy may be required for final diagnosis of unclear presentations.
References

