

Water, Water, Everywhere, but not a Drop in the Vessels: A Case of Systemic Capillary Leak Syndrome

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Introduction

- Systemic Capillary Leak Syndrome (SCLS) is exceedingly rare, with less than 150 cases reported worldwide.
- SCLS can be fatal in the initial episode, necessitating early recognition.

Case Overview

HPI: 74-year-old man presented with a 5-day history of malaise, anorexia, abdominal pain, and orthostasis. He was diagnosed with gastroenteritis. Two days later, he returned to the emergency room with shortness of breath.

PMH: Hypertension
Coronary Artery Disease
Type I Diabetes Mellitus
Ankylosing Spondylitis HLA-B27+

Vitals: 99° F, BP 103/58, HR 108, RR 38
O₂ sat 89% on RA

Exam: Faint rhonchi over the right lung base
Soft, nontender, nondistended abdomen
Warm, well-perfused extremities, no edema

Labs:
12.9 $\frac{19}{57}$ 69 Creatinine: 2.0 mg/dL (baseline 1.1)
Albumin: 3.4 g/dL

Imaging: CXR - scattered interstitial infiltrates in right middle and lower lung

Outside Hospital Course: Treatment for community-acquired pneumonia was initiated, but respiratory status declined precipitously. He was intubated and transferred to the intensive care unit.

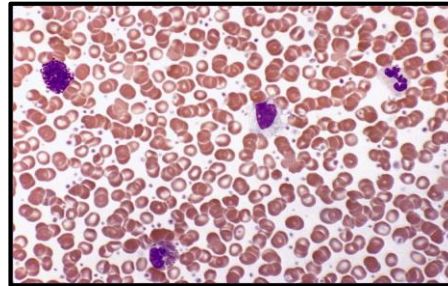
ICU Course: The patient became hypotensive, requiring vasopressor support with norepinephrine, vasopressin, and epinephrine.

39 $\frac{21}{63}$ 97 Creatinine: 2.4 mg/dL
Albumin: <1 g/dL

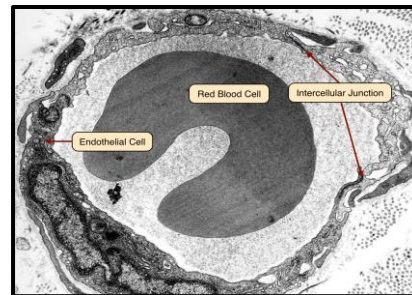
CXR revealed severe pulmonary edema with bilateral pleural effusions. Echocardiography demonstrated a large pericardial effusion.

He was treated for suspected SCLS with aminophylline and terbutaline. Further testing revealed a monoclonal gammopathy.

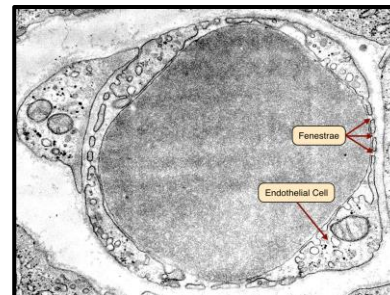
He developed anasarca and multi-organ failure, requiring neuromuscular blockade, three thoracenteses, and hemodialysis. One week later, he was weaned off pressors and extubated. His respiratory status remained tenuous, requiring daily ultrafiltration to maintain oxygenation. He was ultimately reintubated, which prompted the transition to comfort care. The patient died shortly after extubation.



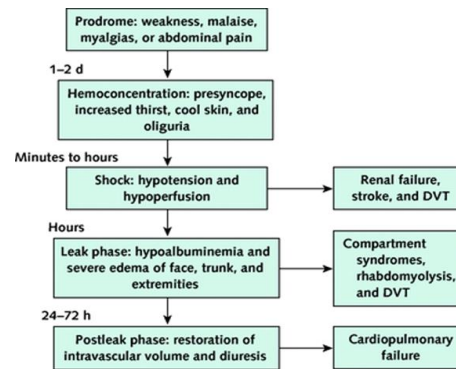
Peripheral blood smear from a patient with polycythemia vera, demonstrating the appearance of hemoconcentration.



Electron microscopy of a continuous capillary.



Electron microscopy of fenestrated capillary.



Discussion

- Systemic Capillary Leak Syndrome is characterized by acute and self-limited profound disruption of the vascular endothelium, resulting in the classic triad of hypotension, hypoalbuminemia, and hemoconcentration.
- SCLS is idiopathic in nature, though the majority of cases are associated with a small monoclonal gammopathy. It is unclear whether these paraproteins incite the syndrome, or represent a secondary effect. Our patient eventually progressed to florid pancytopenia, increasing our suspicion for an underlying hematological process.
- SCLS presents with three phases –
 - Prodromal
 - Extravasation
 - Recovery

The priority in an acute crisis is supportive care through the extravasation phase. Pharmacological therapy has not been shown to shorten the length or change the outcomes of this phase.

- The **cornerstone of management** relies on close monitoring for the abrupt transition to the recovery phase, as the goal of therapy quickly shifts from treatment of intravascular hypovolemia to prevention of life-threatening volume overload.
- Medical therapy primarily targets prevention of recurrent episodes. Currently intravenous immunoglobulin is favored; other options include aminophylline, theophylline, and terbutaline.
- The differential diagnosis of SCLS includes septic shock, toxic shock syndrome, and anaphylaxis. Given the rarity of SCLS, patients are often treated empirically for these conditions initially.
- One should consider SCLS when severe hypoalbuminemia and hemoconcentration are also present, as it requires a unique approach to the management of shock.

References

- Druey KM, Greipp PR. Narrative Review: The Systemic Capillary Leak Syndrome. *Ann Intern Med.* 2010;153:90-98.
 - Guffrey A, Dervieux B, Gravier S, et al. Systemic capillary leak syndrome and autoimmune diseases: A case series. *Seminars in Arthritis and Rheumatism*, 2016.
- Figure 1: University of Minnesota School of Medicine
Figure 2: Systems Cell Biology, Yale University
Figure 3: Druey KM, *Ann Intern Med*