Section: General Medicine

Case Report

AN UNUSUAL CASE OF VIRAL POLYMYOSITISCOMPLICATED BY CAPILLARY LEAK SYNDROME

Received : 24/01/2025
Received in revised form : 15/03/2025
Accepted : 01/04/2025

Keywords: capillary permeability, compartment syndrome, hypotension, hypoalbuminemia, haemoconcentration, hypotension, hypovolemic shock, renal failure, SCLS

Corresponding Author:
Dr. Bhaskar Balasundaram
Email: shaynabansal1997@gmail.com

DOI: 10.47009/jamp.2025.7.2.206

Source of Support: Nil,
Conflict of Interest: None declared

Int J Acad Med Pharm
2025; 7 (2); 1028-1031

Shayna Bansal[1], Vandhana B.S[1], Sandeep Maruti Karimani[1],Bhaskar Balasundaram[2]

[1]Junior Resident,Department of General Medicine, Sapthagiri Institute of Medical Sciences and Research Centre, Bangalore, Karnataka, India
[2]Professor, Department of General Medicine, Sapthagiri Institute of Medical Sciences and Research Centre, Bangalore, Karnataka, India

Abstract

Background: The vascular endothelial barrier maintains intravascular volume and metabolic homeostasis but systemic vascular leakage increases in critical illness associated with sepsis, burns and trauma, certain drugs or toxin exposures. Systemic capillary leak syndrome (SCLS) results in the leakage of fluid and proteins from capillaries into surrounding tissues, potentially causing hypotension, hypoalbuminemia, and haemoconcentration. Complications can encompass overall swelling, compartment syndrome, renal failure, and stroke. Here is a descriptive case report of middle aged man presenting to a tertiary care center with viral prodrome later presenting with complication of capillary leak and wide ranging symptoms. **Conclusion:** SCLS is a life threatening condition characterised by wide ranging complications. Further investigation is essential to understand the connection between inflammatory myositis and systemic capillary leak syndrome (SCLS).

INTRODUCTION

The vascular endothelial barrier maintains intravascular volume and metabolic homeostasis. Although plasma fluids and proteins extravasate continuously from tissue microvasculature (capillaries, post-capillary venules), systemic vascular leakage increases in critical illness associated with sepsis, burns and trauma, among others, or in association with certain drugs or toxin exposures.[1] Systemic capillary leak syndrome (SCLS) results in the leakage of fluid and proteins from capillaries into surrounding tissues, potentially causing hypotension, hypoalbuminemia, and haemoconcentration.[2] Initial symptoms may include fatigue, nausea, abdominal discomfort, extreme thirst, and sudden weight gain. Complications can encompass overall swelling, compartment syndrome, renal failure, and stroke. SCLS manifests in episodic occurrences, varying in frequency; some individuals may experience a single episode throughout their lifetime, while others may have multiple episodes annually. The severity also fluctuates, with the condition posing a risk of fatality. In numerous cases, the underlying cause remains unknown, termed idiopathic SCLS (Clarkson Disease).[3] This disorder is exceedingly rare, affecting fewer than 500 individuals globally.

Incidence: SCLS occurs sporadically and has been described most commonly in middle-aged, Caucasian adults (median age at diagnosis 48; age range newborn to 85 years). There is no sex predominance (52% were female). SCLS has also been reported in fourteen children during the past six years.[3]

Clinical presentation: SCLS flares are not triggered by allergy as such.In the maximum cases reported so far, patients had a readily active and easily identifiable trigger in the form of infections which included predominantly upper respiratory infections such as influenza, respiratory syncytial virus, and West Nile virus. Other factors included increased physical activity, exposure to Sunlight, travel etc.[3]

Many patients have SCLS episodes as “severe” based on the presence of one or more of the following:

1) systolic blood pressure < 60 mm Hg;
2) mean blood pressure < 65 mm Hg;
3) loss of consciousness;
4) admission to the ICU.

While in some other patients, it is characterized by a milder events like smaller drop in blood pressure, fatigue, weakness, and/or dizziness; increased thirst; decreased urinary output; and mild edema of the extremities that may only manifest as muscle tightness or discomfort. And third subset may experience unexplained, non-cyclical peripheral edema but not acute hypotensive episodes, which we call as “chronic” SCLS.[3]

ISSN (O): 2687-5365; ISSN (P): 2753-6556

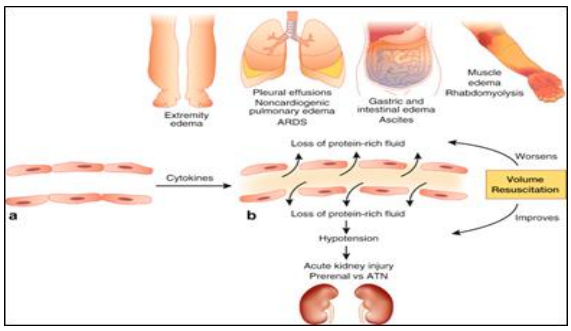

Figure 1: Capillary leak syndrome: etiologies, pathophysiology and management

CASE REPORT

A 38-year-old male who is diagnosed with type 2 diabetes mellitus 1 year back, on treatment and history of chronic alcohol consumption for 20 years, presented with generalised weakness, myalgia and painful swelling of all four limbs since 15 days(distal weakness >proximal weakness). Patient also had high-grade fever, malaise, and reduced appetite for one week.

Vitals on presentation: BP- not recordable
PR- 108 bpm, regular rhythm, feeble
Spo2- 88% under Room Air
Temp-99 degree F
RR- 25 cpm
Early Warning Score-8
General Physical Examination- Patient was conscious, oriented to time, place ,person.
Generalised edema present.
Blanchable Rash with petechia present
Atrophic tongue papilla
No signs of pallor, icterus, clubbing, cyanosis, lymphadenopathy.

Systemic Examination

Central nervous system: Conscious, oriented
Motor System Examination- Power in all 4 limbs- 3/5
Absent deep tendon reflexes
Bilateral plantar-flexor
Muscle tenderness present
Cardiovascular System: S1,S2 present. No Murmurs
Respiratory System: Normal vesicular breath sounds, no added sounds
Per Abdomen: Soft, non tender, no organomegaly felt, Bowel Sounds present
Patient was hemodynamically stabilised. Immediate IV fluid bolus was administered patient was admitted to the ICU.

Table 1: Relevant investigations done as below.

Lab parameters	
Hemoglobin	20.3
Hematocrit	61.4
MCV	89.7
MCH	29.7
Total Leucocyte Count	13800
Platelet Count	1.01 Lac
Serum Urea	85
Serum Creatinine	0.84
Serum Sodium	115
Serum Potassium	7.69
Total Bilirubin	1.82
Serum Albumin	2.9
Aspartate Aminotransferase	2092
Alanine Aminotransferase	559
Lactate Dehydrogenase	4007
INR	1.04
Dengue Serology	Non Reactive
Total Creatine Phosphokinase	71869
HbA1c	10.4
Thyroid Stimulating Hormone(TSH)	10.78

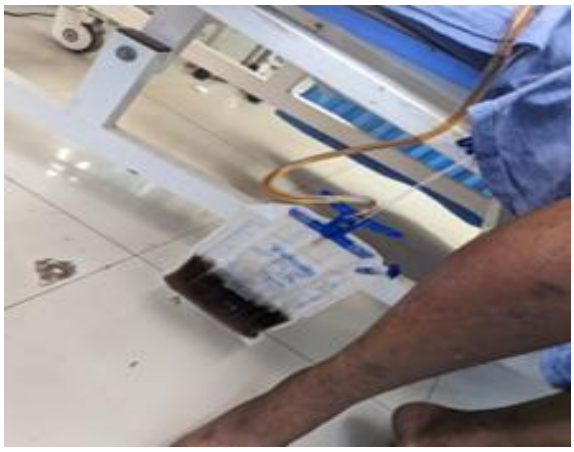

Figure 2: Patient's urobag showing hematuria

Figure 3: Bilateral upper limbs showing edema due to capillary leak

ISSN (O): 2687-5365; ISSN (P): 2753-6556

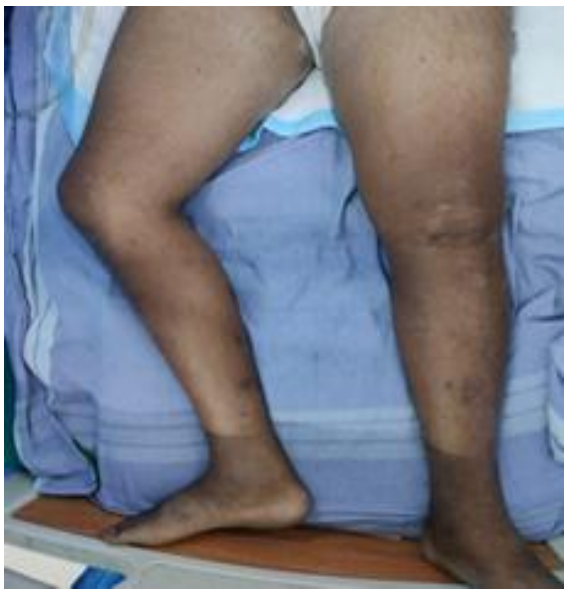

Figure 4: Bilateral lower limbs showing edema and blanchable rashes with petechia

Based on clinical examination, lab investigations, and other relevant investigations, a provisional diagnosis of viral polymyositis was made. Muscle biopsy showed inflammatory myositis overlap myopathy, mostly attributed to a viral aetiology. Further lab investigations showed hypoalbuminemia, raised haematocrit, elevated hemoglobin with raised LDH, serum potassium, and significant myoglobinuria. Persistent low blood pressure was managed with aggressive fluid therapy. The patient was diagnosed with viral polymyositis complicated by a rare syndrome of systemic capillary leak (in view of triad of hypotension, hypoalbuminemia, hemoconcentration) and started on methylprednisolone 1 gram IV as pulse therapy for 5 days, which showed significant improvement in clinical and biochemical parameters. Furthermore, the patient was discharged with combination therapy of azathioprine at 2.5 mg/kg/day with prednisolone 80 mg. After 2 weeks of therapy, the patient's edema reduced by 50% from baseline and is doing well with regular follow-up.

DISCUSSION

SCLS is a disease characterized by spontaneous recurrent episodes of hypovolaemic shock due to extravasation of plasma resulting from altered capillary permeability.an episode may be generally divided into two phases, the capillary leak phase which presents as acute hypovolaemia, and the recruitment phase, when fluid returns to the intravascular space, and may cause pulmonary oedema In the capillary leak phase, albumin and plasma are rapidly transferred from the vascular compartment to the extravascular interstitial compartment . Extravasation, Haemoconcentration, leucocytosis, and a decrease in concentration of albumin are characteristic during acute SCLS attacks. Compartment syndrome and rhabdomyolysis are recognized complications of the capillary leak Renal failure due to acute tubular necrosis secondary to hypovolaemic shock or rhabdomyolysis may also occur.[4,5] Treatment during the capillary leak phase is supportive and dominated by fluid resuscitation, although it is likely that the fluid resuscitation may itself worsen the oedema and muscle damage with resultant compartment syndrome. These patients therefore require intensive or high dependency care or critical monitoring patients with SCLS should be considered during resuscitation The recruitment phase is characterised by abrupt recruitment of the extravasated fluid, which includes the native plasma and the parenteral fluids Significant diuresis may occur but impaired kidneys may be unable to compensate. This may result in pulmonary oedema which is the most frequent cause of mortality in these patients. SCLS is a life threatening condition characterized by severe hypovolaemia, hypotension, haemoconcentration, generalized oedema and typically paraproteinaemia. A compartment syndrome may complicate acute attacks and appropriate monitoring must be considered.

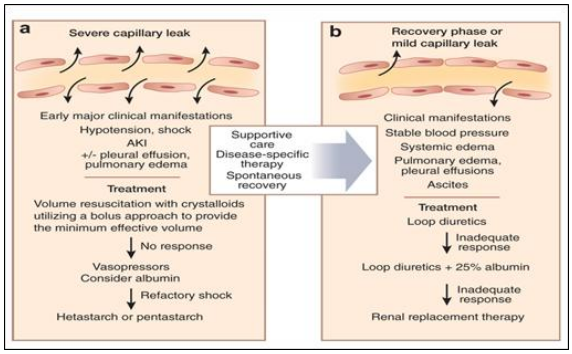

Figure 5: Capillary leak syndrome: an overview

CONCLUSION

Further investigation is essential to understand the connection between inflammatory myositis and systemic capillary leak syndrome (SCLS). There exists a significant gap in our understanding regarding the pathophysiology and common immunomechanisms linking inflammatory myositis with SCLS. It is imperative to conduct more research to bridge this knowledge gap. Notably, SCLS is a rare disorder, impacting fewer than 500 individuals worldwide.

REFERENCES

1. Druey KM, Arnaud L, Parikh SM. Systemic capillary leak syndrome. Nat Rev Dis Primer. 2024 Nov 14;10(1):86.
2. Dolberg-Stolik OC, Putterman C, Rubinow A, Rivkind AI, Sprung CL. Idiopathic Capillary Leak Syndrome Complicated by Massive Rhabdomyolysis. Chest. 1993 Jul;104(1):123–6.
3. Druey KM, Parikh SM. Idiopathic systemic capillary leak syndrome (Clarkson disease). J Allergy Clin Immunol. 2017 Sep;140(3):663–70.

ISSN (O): 2687-5365; ISSN (P): 2753-6556

4. Siddall E, Khatri M, Radhakrishnan J. Capillary leak syndrome: etiologies, pathophysiology, and management. Kidney Int. 2017 Jul;92(1):37–46.

5. Guidet B, Guerin B, Maury E, Offenstadt G, Amstutz P. Capillary leakage complicated by compartment syndrome necessitating surgery. Intensive Care Med. 1990 May;16(5):332–3.

ISSN (O): 2687-5365; ISSN (P): 2753-6556