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Decrypt the link of the ongoing leak: A case of idiopathic systemic capillary leak syndrome (Clarkson's Disease): A case report

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Introduction: Idiopathic systemic capillary leak syndrome (ISCLS) or Clarkson's disease is an extremely rare and fatal condition characterized by episodic attacks of capillary leakage of plasma from the intravascular to the interstitial space resulting in hypotension, hemoconcentration, and hypoalbuminemia. If not diagnosed early, it has a high morbidity and mortality rate. Treatment is supportive, focusing on aggressive but cautious fluid resuscitation together with pharmacologic treatment to control capillary leakage. Each attack varies in severity and can be life features threatening due possible organ failure secondary to poor perfusion. This study aims to raise awareness of the main presentation of ISCLS, to explain the possible pathophysiology, clinical course of the disease, to differentiate with other causes of distributive shock and to present the latest recommendations on treatment and prevention based on limited evidences available.

Case Presentation: Our case is a 38 year old male who initially experienced flu-like symptoms such as body malaise, headache and generalized weakness. He was previously treated as a case of community-acquired pneumonia, high risk, admitted at the intensive care unit. He claimed to have a history of allergy to seafood and medications such as paracetamol and antibiotics. At the emergency room, patient was hypotensive and was managed as a case of anaphylactic shock. He was hydrated, started on inotropic agents and corticosteroid. Work-up tests revealed severe hemoconcentration, hypoalbuminemia, metabolic acidosis, and hyperkalemia. No focus of infection was documented. He remained stable with negative fluid balance until the fourth hospital day, when he suddenly developed pulmonary edema. Patient was managed with diuretics, airway support and inotropics. Patient condition improved and was discharged on 10th hospital day.

Conclusion: ISCLS is a rare and fatal disease that has a high mortality if not detected early. Therefore, prompt recognition is important in the effective management of the disease and its complications.

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