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Weird and wonderful ICU cases: Unusual causes of shock

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ABSTRACT

During their practice, intensivists are ought to face challenging cases that are rare. Intensivists need to be aware of the rare causes of shock beyond common presentations. In each category of shock, there are rare causes that require prompt identification and management. Certain clues in the patient's presentation might point to those rare causes.

Classically shock is classified into: distributive, hypovolemic, cardiogenic, and obstructive. In this era of bedside point-of-care ultrasound, intensivists are able to promptly identify the cause of shock and institute a resuscitation plan. However, there are cases when the diagnosis is still obscure and the cause of shock is not easily identified. For example, in a study of patients admitted with presumed septic shock, 7.4% had no identified cause of shock and 11% had sepsis mimickers.[1]

Hypovolemic shock occurs secondary to a reduction in the effective circulating volume secondary to fluid loss or third spacing. A rare cause of hypovolemic shock is idiopathic capillary leak syndrome (Clarkson Syndrome).[2] The syndrome is characterized by recurrent episodes of rapidly progressive generalized edema, shock, renal failure and high hematocrit. The episode usually resolves

in 3-7 days where the capillary leak resolves and a phase of pulmonary edema occurs. Several treatment options such as intravenous immunoglobulin (IVIG) and aminophylline were used in case reports.[3]

Vasodilatory shock occurs secondary to peripheral vasodilation and decrease in blood flow. It occurs as part of the systemic inflammatory response syndrome for which sepsis, acute pancreatitis, acute liver failure, and major trauma are common causes. Rare causes that need to be considered include: hemophagocytic lymphohistiocytosis (HLH), systemic mastocytosis, and toxic shock syndrome.

Hemophagocytic lymphohistiocytosis (HLH) is a hyperinflammatory syndrome characterized by macrophage activation and engulfment of hemopoetic cells which leads to pancytopenia. It is also characterized by cytokines storm that lead to a vasodilatory shock, multi-organ failure, and acute respiratory distress syndrome (ARDS). The most common triggers are infection, malignancy, and autoimmune diseases. Pointers to this diagnosis in the intensive care unit include: pancytopenias, hypofibrinogenemia, high triglycerides, and high ferritin. Treatment necessitates treating the underlying cause as well as using immune modifying therapies.[4]

Systemic mastocytosis is a rare cause of recurrent anaphylaxis shock. It results from the accumulation of mast cells in tissues and can present with anaphylaxis and vascular collapse. An important clue to the diagnosis is the presence of urticarial pigmentosa and the absence of an allergen history.[5]

Toxic shock syndrome is a unique cause of sepsis. It is caused by a pre-formed toxin produced by Staphylococcus aureus and Streptococcus pyrogenes. The clue to the diagnosis include the rapid onset after the precipitating factor, erythroderma, and skin desquamation. Treatment includes IVIG and Clindamycin.[6]

Keywords: rare cases, ICU, Clarkson syndrome, hemophagocytic lymphohistiocytosis, systemic mastocytosis

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