AUTOIMMUNE CONNECTIVE TISSUE DISEASES

SYSTEMIC CAPILLARY LEAK SYNDROME IN DERMATOMYOSITIS: A REPORT OF 2 CASES

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Background: Systemic capillary leak syndrome (SCLS) is a rare, often fatal, condition characterized by hemoconcentration, hypoalbuminemia and hypotension. Its association with other disease entities remains unclear. In this case report, we describe two diagnosed cases of Dermatomyositis, an idiopathic inflammatory myopathy, complicated with SCLS.

Observation: The first case is of a 41 –year-old, male and the second case is of a 61-year-old female. Both cases were admitted for the management of dermatomyositis and presented with characteristic cutaneous lesions of the condition, proximal muscle weakness, elevated muscle enzymes, positive ANA, abnormal results in their electromyogram and nerve conduction studies as well as supporting skin punch biopsy results.

Suspicion of SCLS arose when both patients, while admitted, were observed to have dyspnea, abdominal pain, sudden rapid weight gain, anasarca and persistent hypotension. Laboratory examinations of both patients revealed leukocytosis, elevated hematocrit concentration, and hypoalbuminemia.

Multidisciplinary management, involving Rheumatology, Nephrology, Hematology, Cardiology, Pulmonology, Endocrinology, and Surgery, for both cases were done. To address SCLS, intravenous immunoglobulin (IVIG) coupled with Albumin, Aminophylline and steroid infusion were used for the first case. The second case was managed mainly with high-dose intravenous corticosteroids and diuretics.

Key message: This report explores the link between SCLS and Dermatomyositis. It emphasizes the significance of early recognition of SCLS in the background of Dermatomyositis and validates the role of both IVIG and corticosteroids in preventing the mortality associated with SCLS.