Acute sensory and autonomic neuronopathy (ASANN)

Acute-onset and severe sensory and autonomic deficits with no motor dysfunction, typically preceded by a febrile illness, with poor recovery, and the often fatal outcome are the hallmark features of acute sensory and autonomic neuronopathy (ASANN). Pathologically and electrophysiologically, ASANN is characterized by an extensive ganglionopathy affecting sensory and autonomic ganglia with the preservation of motor neurons. Consequently, patients, usually children or young adult, develop acute-onset profound widespread loss of all sensory modalities resulting in automutilations, as well as autonomic failure causing neurogenic orthostatic hypotension, neurogenic underactive bladder, and gastroparesis and constipation. The diagnosis is clinical with support of nerve conduction studies and autonomic testing, as well as spinal cord magnetic resonance imaging showing characteristic posterior cord hyperintensities. Although the presumed etiology is immune-mediated, further studies are required to clarify the physiopathology of the disease. We here performed a systematic review of the epidemiology, pathophysiology, diagnosis, and management of ASANN, with three representative cases that recently presented at our clinic. All three patients had the typical clinical manifestations of ASANN but in different combinations, illustrating the variable phenotype of the disorder. Immunosuppression is seldom effective. Management options are limited to supportive and symptomatic care with the goal of minimizing complications and preventing death [1].

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Gutierrez J, Palma JA, Kaufmann H. Acute Sensory and Autonomic Neuronopathy: A Devastating Disorder Affecting Sensory and Autonomic Ganglia [published online ahead of print, 2020 Sep 9]. Semin Neurol. 2020;10.1055/s-0040-1713843. doi:10.1055/s-0040-1713843