

Stiff-person syndrome and other myelopathies constitute paraneoplastic neurological syndromes

Source

Department of Neurology, Majors of Medical Sciences, Graduate School of Comprehensive Human Sciences, University of Tsukuba, 1-1-1 Tennodai, Tsukuba, Ibaraki 305-8575, Japan.

Abstract

Stiff-person syndrome (SPS) is an autoimmune neurological disorder characterized by rigidity of the trunk and proximal limb muscles, intermittent superimposed spasms, and increased sensitivity to external stimuli. It has been more than 50 years since Moersch and Woltman reported the first 14 cases with this syndrome. During the last half century, many autoantibodies discovered, such as anti-glutamic acid decarboxylase (GAD), anti-amphiphysin, anti-gephyrin, and anti-gamma-aminobutyric acid A receptor-associated protein (GABARAP) antibodies. There is strong evidence that in SPS, GABAergic neurotransmission is impaired by these pathogenic autoantibodies; however, the exact antigenic target remains unknown. This chapter focuses on the recent advances in the diagnosis, immunopathogenesis, and treatment of paraneoplastic SPS. Paraneoplastic SPS accounts for approximately 5% of all cases of SPS, and is associated with anti-amphiphysin, anti-gephyrin, and anti-Ri antibodies. In addition, author has reported cases of patients with SPS who were positive for anti-GAD antibodies and subsequently developed cancer. Because SPS often develops before the diagnosis of cancer, patients diagnosed with SPS should be monitored for the development of cancer. The treatment of SPS includes the administration of GABA enhancing and antispasmodic drugs and immunomodulating therapies such as the administration of intravenous immunoglobulin (IVIG). Treatments for cancer occasionally produce symptomatic improvement in patients with paraneoplastic SPS. Although the understanding and treatment of SPS have evolved, the disease remains underdiagnosed. In the past, some patients with SPS have been diagnosed with psychiatric disorders. Therefore, it is important to increase awareness of SPS among practicing physicians.

PMID:

20420178

[PubMed - indexed for MEDLINE]