Conclusions

Neuronal uptake of normal and paraneoplastic IgGs requires the interaction of the Fc portion of the IgG molecule with previously uncharacterized neuronal FcγRI receptors. Our study provides a mechanism through which antibodies reactive with intracellular neuronal proteins could gain access to their target antigens to cause neuronal injury and neurological disease. The observation that neuronal antibody uptake can be blocked by normal IgG has possible implications for patient treatment.

Disclosure: Dr. Smith has nothing to disclose. The institution of an immediate family member of Dr. Liu has received research support from NIH. The institution of Mr. Carlson has received research support from Biogen. Mr. Carlson has received personal compensation in the range of $100,000-$499,999 for serving as a Employee as a Researcher with Veteran Affairs. Dr. Clardy has received personal compensation for serving as an employee of Veterans Health Administration (VHA). Dr. Clardy has received personal compensation for serving as an employee of University of Utah Health. Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for Clarion. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for ExpertConnect. The institution of Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for VielaBio. The institution of Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for Genentech. The institution of Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Alexion. The institution of Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for GuidePoint. Dr. Clardy has received personal compensation in the range of $10,000-$49,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Neurology/AAN Publications. The institution of Dr. Clardy has received research support from Alexion Pharma. The institution of Dr. Clardy has received research support from Sumaira Foundation for NMO. The institution of Dr. Clardy has received research support from Immune Deficiency Foundation. The institution of Dr. Clardy has received research support from Western Institute for Veteran Research. The institution of Dr. Clardy has received research support from NIH/NINDS. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a AAN Summer Meeting CoDirector Travel and Lodging with AAN. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Grand Rounds Travel and Lodging with U of Iowa. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Speaker Honoraria for Grand Rounds with Barrow Neurological Institute. Dr. Greenlee has received personal compensation in the range of $500-$4,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Medlink. Dr. Greenlee has received publishing royalties from a publication relating to health care. Dr. Greenlee has received publishing royalties from a publication relating to health care.

Anti-Tr/DNER Paraneoplastic Cerebellar Degeneration with Marked Cerebellar and Psychological Symptoms Responsive to Plasma Exchange

Paul Crane, DonRaphael Pratt Wynn, Dana DeWitt, John Greenlee

Objective

We present a patient who developed cerebellar degeneration and severe psychological symptoms leading to the diagnosis of Hodgkin's disease and detection of anti-Tr/DNER antibodies. The patient failed to respond methylprednisolone intravenous immunoglobulin G, rituximab, and tumor treatment but had significant improvement with plasma exchange (PLEX).

Background

Paraneoplastic cerebellar degeneration accompanying Hodgkin's disease may have its onset prior to detection of the underlying malignancy, during its course, or following treatment. The associated autoantibody, anti-Tr, is reactive with neuronal delta/notch-like epidermal growth factor-related receptors (DNER), an autoantibody not included in all paraneoplastic testing screens. The condition characterized by progressive cerebellar injury, and response to immunosuppressive therapy and tumor treatment is generally poor.

Design/Methods

Case Presentation: A 60-year-old male presented with diplopia, progressive loss of balance, and ataxia, with impaired short-term memory, confusion, and anger outbursts. Initial commercial screen for paraneoplastic autoantibodies was negative. Two months following his initial presentation he developed inguinal lymphadenopathy. He was diagnosed as having Hodgkin's Lymphoma Stage 1B and found by a second laboratory to have anti-TR/DNER antibodies (Titer 1:3480; Reference range <1:240), an antibody not included in the initial testing panel. CSF analysis was notable for a protein of 92 mg/dL. MRI demonstrated normal findings for age.

Results

Treatment with Doxorubicin-Bleomycin-Vinblastine-Dacarbazine (ABVD), pulse methylprednisolone, and intravenous immunoglobulin did not affect disease progression. Plasma exchange PLEX resulted in marked improvement. Symptoms worsened during subsequent treatment with intravenous immunoglobulins and rituximab but improved with further plasma exchange.

Conclusions

Although Hodgkin's disease is an important malignancy in paraneoplastic cerebellar degeneration, its associated autoantibody is not necessarily included in commercial paraneoplastic autoantibody screens, potentially leading to delay in diagnosis. Our patient's dramatic improvement with PLEX suggests that PLEX should be considered early in treatment, or where there is poor response to other treatment modalities.

Disclosure: Dr. Crane has nothing to disclose. Dr. Wynn has nothing to disclose. Dr. DeWitt has received personal compensation in the range of $5,000-$9,999 for serving on a Speakers Bureau for Biogen. Dr. Greenlee has received personal compensation in the range of $500-$4,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Medlink. Dr. Greenlee has received publishing royalties from a publication relating to health care. Dr. Greenlee has received publishing royalties from a publication relating to health care.

CASPR-2 Antibody Associated Autoimmunity in the Setting of COVID-19 (Infection, Vaccination, or Both?) and Chronic Lymphocytic Leukemia: Case Report and Review of the Literature

Neda Sattarnezhad, Jamie McDonald, Anna Tomczak, Julia Sumera, Jacob Loeffler, May Han

Objective

To report a case of Anti-Contactin-Associated Protein-like2 (CASPR-2) autoimmunity in a patient with low-grade Chronic Lymphocytic Leukemia (CLL) following COVID-19 vaccination and infection.

Background

Anti-CASPR2 antibody disorder has been associated with neoplastic disorders like thymoma. Recent reports enlist COVID-19 as apotential trigger of CASPR2 autoimmunity. While the clinical presentations are similar, management differs based on the underlying etiology.

Design/Methods

We review a case of anti-CASPR2-antibody associated disorder with concurrent low grade CLL and recent history of COVID-19 vaccination and infection. Additionally, we review the literature and discuss the therapeutic challenges.

Results

A 73-years old male presented with five months of progressive fatigue, weight loss, diffuse sweating, muscle cramps, and neuropathic pain. He eventually developed bilateral upper and lower facial weakness. Patient

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Paul Crane, DonRaphael Pratt Wynn, Dana DeWitt, et al.

Neurology 2022;99;S67
DOI 10.1212/01.wnl.0000903536.25426.aa