Conclusions

To our knowledge, hyperintensity within the medulla and spinal trigeminal tract in zoster opthalmicus is a rare imaging finding. We have coined this radiologic finding the "trigeminal tract sign" due to the anatomic structure affected. A thorough radiologic/laboratory workup should be considered in patients with similar presentations; however, a high index of suspicion for zoster opthalmicus should be considered in cases with the aforementioned imaging findings. Whether this radiological finding predicts disease severity or the likelihood of multiple cranial nerve involvement (as was seen in all patients in this series) requires further study.

Disclosure: Dr. Yong has nothing to disclose. Dr. Wallace has nothing to disclose. Dr. Kapadia has nothing to disclose.

Characterization of Clinical and Paraclinical Features Associated With TS-HDS Autoantibody Seropositivity

Mohamed Rezk, Igal Mirman, Sarah Berini, Pitcha Chompoopong, Christopher Klein, John R. Mills, Divyanshu Dubey

Objective

To evaluate neuropathy phenotypes and clinical outcomes associated with trisulfated heparin disaccharide (TS-HDS) autoantibodies.

Background

TS-HDS autoantibody has been reported as a biomarker of immune-mediated neuropathy. However, studies evaluating the clinical associations of this autoantibody are limited.

Design/Methods

Electronic medical records were reviewed to identify TS-HDS autoantibody seropositive patients and characterize their clinical and electrodiagnostic findings.

Results

Seventy-seven TS-HDS-IgM seropositive (titer range 9000-350,000) patients were identified (50 females; median age of onset was 48 years (range 9-83 years). Eleven patients were also positive for FGFR3-IgG (titer range 4000-19,000). 70% (54/77) had clinical/paraclinical evidence of neuropathy (54/77, 70% of TS-HDS-IgM alone; 10/11, 91% of TS-HDS-IgM with coexisting FGFR3-IgG). The managing physician characterized an immune-mediated neuropathy in 30% (23/77) and 54% (6/11) of the TS-HDS-IgM only and TS-HDS-IgM with coexisting FGFR3-IgG seropositive patients, respectively. Small fiber neuropathy presented in 58% (45/77) and 63% (7/11) of TS-HDS-IgM only, and both antibodies seropositive patients, respectively. Length-dependent neuropathy was the most common neuropathy phenotype amongst TS-HDS IgM (43/54, 80%) and dual seropositive cases (7/11, 63%). Forty-one (53%) patients received immunotherapy, predominantly: IVIG (n = 37), IV solumedrol (n = 7), oral prednisone (n = 14), and mycophenolate mofetil (n = 12). Among these, 43% (15/35) with TS-HDS-IgM seropositivity alone had improvement in inflammatory neuropathy cause and treatment (INCAT) disability score or modified Rankin Scale (mRS), while 33% (2/6) of patients with dual seropositivity had INCAT and mRS improvement. TS-HDS-IgM titers had low discriminative ability to identify immunotherapy response with an AUC of 0.621.

Conclusions

Neuropathy associations and clinical phenotypes amongst TS-HDS-IgM seropositive cases are variable. Furthermore, only a minority of cases are immunotherapy responsive, limiting the value of this biomarker in identifying immune-mediated neuropathies.

Disclosure: Dr. Rezk has nothing to disclose. Dr. Mirman has nothing to disclose. Dr. Berini has nothing to disclose. Dr. Chompoopong has nothing to disclose. Dr. Klein has a non-compensated relationship as a Klein with Neurology Journal that is relevant to AAN interests or activities. Dr. Mills has received intellectual property interests from a discovery or technology relating to health care. The institution of Dr. Dubey has received personal compensation in the range of $500-$4,999 for serving as a Consultant for UCB. The institution of Dr. Dubey has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Astellas. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for AGRIMS. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Advances in Neurology. Dr. Dubey has received personal compensation in the range of $0-$499 for serving on a Speakers Bureau for Moffit Cancer Center. Dr. Dubey has received research support from Department of Defense. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care. Dr. Dubey has received intellectual property interests from a discovery or technology relating to health care.

BNT162b2 mRNA COVID-19 Vaccine Three-Dose Safety and Risk of COVID-19 in Patients With Myasthenia Gravis

Alon Doron, Yoav Piura, Ifat Vigiser, Hadar Kolb, Keren Regev, Nahum Nesher, Arnon Karni

Objective

To study the COVID-19 vaccine three-dose safety and risk of COVID-19 in patients with myasthenia gravis

Background

Various vaccines, including those against SARS-CoV-2, were reported to trigger or exacerbate myasthenia gravis (MG). As COVID-19 may potentially contribute to the tendency of MG patients to develop respiratory failure, it is important to study the safety of vaccines against SARS-CoV-2 and assess the risk of COVID-19 in MG patients.

Design/Methods

Among 215 MG patients treated in Tel Aviv Medical Center, 160 were interviewed about their response to the three-dose BNT162b2 mRNA vaccine. We assessed exacerbation rate and safety in a period of up to 6 weeks from each vaccine dose, as well as patient morbidity and mortality during COVID-19 compared to the general population.

Results

430 vaccine doses were administered across 150 patients. Thirteen patients (8.7%) complained of exacerbation within 6 weeks (risk period) of each vaccine dose, 8 (5.3%) confirmed by physician report. The exacerbation rates were similar during the risk period (5.6%) compared to corresponding period the previous year (4.8%). MG onset rates during the vaccination period were unaffected compared to previous years. Exacerbation rate among 15 patients who had COVID-19 was significantly higher (40%) compared to rate in the risk period following vaccination, with higher severe or lethal COVID-19 (26.7%) compared to the general population (0.96%), occurring in unvaccinated, steroid-treated, generalized MG patients.

Conclusions

Three-dose BNT162b2 vaccination is neither associated with exacerbation nor the new onset of MG, whereas COVID-19 is associated with severe disease and death in unvaccinated, steroid-treated generalized MG patients. Hence, it is strongly recommended for generalized MG patients to get vaccinated

Disclosure: Mr. Doron has nothing to disclose. Yoav Piura has received personal compensation in the range of $0-$499 for serving as a Consultant for Merck. Yoav Piura has received personal compensation in the range of $0-$499 for serving as a Consultant for Novartis. Ifat Vigiser has nothing to disclose. Hadar Kolb has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Novartis. Hadar Kolb has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Marck. Dr. Regev has nothing to disclose. Steve Barash has received personal compensation for serving as an employee of Teva. Steve Barash has received stock or an ownership interest

Characterization of Clinical and Paraclinical Features Associated With TS-HDS Autoantibody Seropositivity

Mohamed Rezk, Igal Mirman, Sarah Berini, et al.

Neurology 2022;99;S71
DOI 10.1212/01.wnl.0000903568.79179.7c