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

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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.

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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

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Comparison of Fixed Cell-based Assay to Radioimmunoprecipitation Assay for Acetylcholine Receptor Antibody Detection in Myasthenia Gravis

Ario Mirian, Michael Nicolle, Adrian Budhram

Objective

To compare specificity and sensitivity of a commercially available fixed cell-based assay (F-CBA) to radioimmunoprecipitation assay (RIPA) for acetylcholine receptor antibody (anti-AChR) detection in myasthenia gravis (MG).

Background

Approximately 50% of ocular and 85% of generalized MG are anti-AChR positive by RIPA, the current gold standard test. Clustered live cell-based assay (L-CBA) can detect low-affinity anti-AChR that are missed by RIPA, but the costly and time-consuming nature of L-CBA has restricted its use to specialized centres. A commercial F-CBA has become available for anti-AChR detection, but its diagnostic performance compared to RIPA requires evaluation.

Design/Methods

In this retrospective diagnostic cohort study we reviewed the clinical information of suspected MG patients evaluated at London Health Sciences Centre MG clinic, who were clinically classified as MG or non-MG and who had anti-AChR RIPA and then F-CBA performed. Classification of each patient as anti-AChR F-CBA-negative/positive, RIPA-negative/positive, and MG/non-MG permitted specificity and sensitivity calculations for each assay.

Results

Six-hundred-eighteen patients were included in study analysis. The median patient age at time of sample collection was 45.8 years (range: 7.5–87.5 years) and 312/618 (50.5%) were female. Of 618 patients, 395 (63.9%) were classified as MG. Specificity of both F-CBA and RIPA was excellent (99.6% vs. 100%, P > 0.99). One F-CBA-positive patient was classified as non-MG, although in retrospect ocular MG with functional overlay was challenging to exclude. Sensitivity of F-CBA was significantly higher than RIPA (76.7% vs. 72.7%, P = 0.002). Overall, 20/97 (21%) otherwise SNMG patients after RIPA evaluation had anti-AChR detected by F-CBA.

Conclusions

In our study anti-AChR F-CBA and RIPA both had excellent specificity, while F-CBA had 4% higher sensitivity for MG and detected anti-AChR in 21% of SNMG patients. Our findings indicate that F-CBA is a viable alternative to RIPA for anti-AChR detection.

Disclosure: Dr. Mirian has nothing to disclose. Dr. Nicolle has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for Alexion. Dr. Budhram has nothing to disclose.

Piloting an Advanced Neuroimmunology Elective for Neurology Residents

Sonia Kaur Singh, Rohini Samudralwar

Objective

To describe the creation of an Advanced Neuroimmunology elective for residents with a special interest in clinical neuroimmunology.

Background

There has been a dramatic change in the landscape of neuroimmune conditions with the discovery of new pathogenic autoantibodies, disease modifying therapies and wider availability of multidisciplinary care systems for patients. Most residencies do offer exposure to multiple sclerosis but with increasing interest in neuroimmunology and autoimmune neurological conditions, there is a gap in resident education to meet needs of this changing landscape.

Design/Methods

A curriculum for advanced neuroimmunology (NI) was developed for residents with special interest in clinical neuroimmunology. This two-week elective consisted of rotations through NI and affiliated multidisciplinary clinics to increase exposure to immune mediated neurological illnesses, appreciate their heterogeneity, and aid multidisciplinary approach. Department experts in various disease states related to neuroimmunology were contacted and based on interest and resident elective time, a schedule was set up for rotations through neuroinfectious diseases, pulmonary sarcoidosis clinic, neuro-oncology, neuropathology and rheumatology. An additional expectation was to work with the fellow on inpatient consults that came in through the 2 weeks. In addition to multiple sclerosis/neuroimmunology division didactics, residents are encouraged to attend other affiliated department conferences as well as present at interdepartmental meetings, such as neuro-rheumatology conference.

Results

The availability of this elective allowed increased exposure to neuroimmunological conditions outside the typical Multiple Sclerosis elective at UTHealth. It also has allowed for additional inter-departmental collaboration clinically. Since the initial pilot elective, more residents have requested this as an elective and will be surveyed about their experience.

Conclusions

There is an unmet need for MS and NI subspecialists. Exposure to the broad spectrum of neuroimmunological conditions through multidisciplinary collaborations during residency is instrumental to ensure future specialists have the foundations to adapt to this rapidly advancing field.

Disclosure: Dr. Singh has nothing to disclose. Dr. Samudralwar has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Sanofi Genzyme. Dr. Samudralwar has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for Biogen. Dr. Samudralwar has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for EMD Serono.

A Case of Pembrolizumab (Anti-PD-1) Induced Encephalitis

Anza Zahid, Meryim Poursheykhi, Mujtaba Saeed, Ivo Tremont

Objective

N/A.

Background

PD-1 Immune checkpoint inhibitors (ICI) have been associated with neurologic immune-related adverse events including meningoencephalitis and limbic encephalitis that can manifest as paraneoplastic syndromes. We present a case of suspected pembrolizumab (anti PD-1) induced limbic encephalitis presenting as episodic aphasia.

Design/Methods

N/A.

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

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