frequency of subsequent IVIg treatment and associated cost during the first 12 months post-IVIg initiation were analyzed. Usage patterns of concomitant gMG treatments during the year preceding and 3 years post-IVIg initiation were compared.

Results

Among 1225 patients with gMG who initiated IVIg treatment, 706 patients (57.6%) received 1 to 5 IVIg treatment courses (intermittent IVIg users), and 519 patients (42.4%) received = 6 IVIg treatment courses (chronic IVIg users) within the subsequent year. Mean annual medical cost per patient was nearly 2.5-fold higher for chronic vs intermittent IVIg users ($161,478 vs $64,888, $p < 0.001$). Usage frequency of other immunotherapy treatments did not decrease over the follow-up period, even for patients who continued annual chronic IVIg for 3 consecutive years post-initiation.

Conclusions

Nearly half of patients with gMG received chronic and multiple IVIg treatment courses within the first year once initiating IVIg treatment, indicating higher usage than expected. For all IVIg initiators, usage frequency of other immunotherapy treatments did not decrease over 3 years despite IVIg initiation.

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Neurological Immune-Related Adverse Events After COVID-19 Vaccination: A Systematic Review

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Objective

The goal of this study is to compile published data reporting neurological immune-related adverse events following COVID-19 vaccination, not including those relating to hematologic abnormalities such as thrombosis or hemorrhage.

Background

COVID-19 vaccination has been repeatedly shown to reduce the incidence and severity of COVID-19 infection. The expedited timeline of these vaccines has given rise to many discussions pertaining to their safety. Many neurological and non-neurological adverse events have been linked to COVID-19 vaccination including acute MI, anaphylaxis, appendicitis, Bell's palsy, deep vein thrombosis, disseminated intravascular coagulation, encephalomyelitis, transverse myelitis, and many others.

Design/Methods

The following databases were searched in April 2021 using different keywords: PubMed, Medline, Embase, Scopus, Web of Science, Science, Direct, MedRxiv, and Lens.org. Studies were included if they reported any adverse immune-related neurological events secondary to COVID-19 vaccination. Studies were excluded if they were not in English, included self-reported events only, or did not report primary data. Screening and extraction were conducted by 2 different reviewers using Covidence.

Results

The search strategy yielded 18 studies which reported a total of 61 patients who had received a COVID-19 vaccination and experienced = 1 neurological adverse events. Most reported adverse events were facial nerve palsy (52.5%), reactivation of herpes zoster (11.5%), Guillian-Barré syndrome (6.6%), demyelinating disease (6.6%), and neuropathy (11.5%). Other reported adverse effects were delirium, periauricular vesicular rash, bilateral sensorineural hearing loss, visual disturbance, gait disturbance, serotonin syndrome, and vestibular ataxia (16.4%).

Conclusions

The symptoms were time-limited and self-resolving in nature. In addition, the incidence of the reported events following COVID-19 vaccination compared to the general population is similar. Hence, there is little to no evidence suggesting a causal relationship between COVID-19 vaccination and neurological adverse events.

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Post COVID-19 Vaccination Associated Acquired Neuromyotonia

Shivangi Shivangi, Amit Shankar Singh, Jeenendra Prakash Singhvi, Namit Bansal, Rajat Gupta

Objective

Acquired neuromyotonia or Isaacs' syndrome is an immune mediated inflammatory disorder characterized by involuntary continuous muscle fiber activity manifesting as twitching and stiffness along with autonomic dysfunctions like hyperhidrosis and/or tachycardia. Here we report a young male who developed acquired neuromyotonia following COVID-19 vaccination.

Background

A 20-year-old male presented in our clinic with gradually progressive pain and numbness in bilateral lower limbs, tremors in both hands, shivering while walking, excessive sweating and difficulty in micturition for last 15 days. He also noticed twitching of muscles in calf and thigh muscles along with these symptoms. According to patient, these symptoms started after he took his first dose of COVID-19 vaccination (Covishield- Oxford-AstraZeneca viral vector vaccine) 10 days back. There was no history of fever or backache. He had no chronic illness and was not on any medications. Examination revealed hyperhidrosis, mild proximal muscle weakness in both lower limbs with twitching in muscles suggestive of myokymia. There were quivering and rippling movements of intrinsic muscles of both hands resembling polyminimyoclonus. In view of the above findings, possibility of acquired neuromyotonia possibly following COVID-19 vaccination was kept and further evaluation was done.

Design/Methods

Routine blood investigations, thyroid function test, anti-thyroid peroxidase antibodies and anti-nuclear antibodies were normal. Cerebrospinal fluid analysis was normal. Anti-VGKC antibodies were detected in serum with strongly positive anti-CASPR and weakly positive anti-LGI1 antibodies confirming diagnosis of acquired neuromyotonia.

Results

Pulse dose of intravenous methylprednisolone for 5 days was given which resulted in visible improvement in pain, twitching, hyperhidrosis and urinary symptoms. He was continued on oral steroids and complete resolution of his symptoms was noted over a period of 2 months.

Conclusions

COVID-19 vector vaccine associated acquired neuromyotonia is a rare condition, but its early recognition and treatment is the key for a favorable prognosis.

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Medusa-head Antibodies-More than Cerebellar Ataxia: Report of Two Cases

Lei Liu, Jingxiao Zhang

Objective

To report an anti-metabotropic glutamate receptor 1 (mGluR1) encephalitis case present with cerebellar ataxia and a paraneoplastic retinopathy and optic neuropathy case with anti-inositol 1,4,5-trisphosphate receptor type 1 (ITPR1) antibodies.

Background

Medusa-head antibodies refer to a group of autoantibodies with a staining pattern resembling a Gorgon's head, caused by binding of IgG to Purkinje cell somata and dendrites in tissue-based assay. Although these antibodies often related with cerebellar ataxia, increasing cases presented or accompanied with encephalopathy or peripheral neuropathy have been reported.

Design/Methods

Cases report.

Results

Case 1: A 48-year-old woman presented with acute dizziness and unsteady gait developed head titubation, repeated language and calculation impairment. Her neural antibodies were negative with commercial kits. Further investigation showed serum and CSF "Medusa head" staining pattern of Purkinje cell in rat cerebellum. An in-house cell-based assay proved the existence of anti-mGluR1 antibodies. One-year follow-up revealed serum antibodies titers dramatically decreased and CSF antibodies negative after using steroids and intravenous immunoglobulin, but still left prominent cerebellum atrophy and severe cerebellar deficits. Case 2: Two years ago, a 61-year-old man presented with painless nasal defects of right eye, photopsia and progressed to 1 m/FC within 3 days. A similar episode happened 7 months ago in his left eye. Prior malignancy including renal carcinoma and lung adenocarcinoma discovered seven and four months ago, respectively. On admission, visual acuities were OD 50 cm/FC and OS 1 m/FC. There was a right RAPD. MRI showed bilateral optic nerves atrophy. Serum AQP4, MOG and paraneoplastic antibodies were all negative. ITPR1 antibodies were proved by tissue-based assay and Euroimmun cell-based assay. He didn't accept immunotherapies and was discharged.

Conclusions

Besides cerebellar ataxia, anti-mGluR1 antibodies may also cause encephalopathy symptoms. Our findings indicate a possible role of autoimmunity to ITPR1 in the pathogenesis of paraneoplastic retinopathy and optic neuropathy and expand the panel of diagnostic markers for this condition.

Disclosure: Dr. Liu has nothing to disclose. Dr. Zhang has nothing to disclose.

Testing for N-type Voltage Gated Calcium Channel Antibody has Limited Utility in Evaluating Patients With Suspected Lambert-Eaton Myasthenic Syndrome

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Objective

To determine if testing for N-type voltage-gated calcium channel antibody improves diagnostic performance of serological testing for Lambert-Eaton Myasthenic Syndrome.

Background

Patients with suspected autoimmune neuromuscular junction transmission disorders are commonly tested for both P/Q-type (VGCC-P/Q) and N-type (VGCC-N) voltage-gated calcium channel antibodies to aid the diagnosis of Lambert-Eaton Myasthenic Syndrome (LEMS).

Design/Methods

This retrospective cross-sectional study included 93 patients diagnosed at Mayo Clinic with LEMS based on electrodiagnostic findings, clinical presentation and positive serology for VGCC-P/Q and/or VGCC-N.

Results

Forty-five patients (48.4%) were female, of median age 61 years (range 11- 99). Twenty-five (26.9%) were positive for both VGCC antibody types (reference interval <0.04 nmol/L [VGCC-N] and <0.03 nmol/L [VGCC-P/Q]); 67 (72.0%) were positive for VGCC-P/Q only and one (1.1%) was positive for VGCC-N only. The single VGCC-N positive only result [VGCC-N = 0.10 nmol/L; VGCC-PQ = 0.02 nmol/L] was from a patient with classic LEMS presentation and no evidence of malignancy by Chest CT. The prevalence of VGCC-N only positivity was lower than in the healthy population or other disease control populations. VGCC-P/Q antibody titers were higher in patients who had detectable co-existing VGCC-N antibody (median titer 1.58 versus 0.39 nmol/L, P = 0.02) and there was a positive correlation (r = 0.762, P < 0.001) between VGCC-N and VGCC-P/Q titers in double positive cases. Dual positivity did not significantly increase the likelihood of an underlying cancer (40.9% versus 30.1%, P = 0.33).

Conclusions

Testing for VGCC-P/Q antibodies alone is sufficient in the serological evaluation of suspected LEMS cases. Inclusion of VGCC-N antibody testing does not improve diagnostic performance. A positive VGCC-N antibody did not significantly increase the risk of paraneoplastic LEMS.

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