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118787

Safety of eculizumab in NMOSD and MG: Analysis of the phase 3 studies prevent and regain, and their extensions

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Background and aims

Eculizumab (a terminal complement inhibitor) demonstrated efficacy in reducing relapse risk and eliciting clinical improvements in the phase 3, randomised, double-blind PREVENT (NCT01892345) and REGAIN (NCT01997229) studies and their open-label extensions (NCT02003144 and NCT02301624, respectively) in aquaporin-4 immunoglobulin G-positive neuromyelitis optica spectrum disorder (AQP4+ NMOSD) and acetylcholine receptor antibody-positive generalised myasthenia gravis (AChR+ gMG), respectively. The aim of this analysis was to compare infection rates for eculizumab vs placebo according to number of concomitant immunosuppressive therapies (ISTs) during these studies. Eculizumab is not reimbursed for neurology indications in Italy as of April 2021.

Methods

Patients were randomised to eculizumab or placebo. Post hoc analysis examined infection rates overall and by number of baseline ISTs.

Results

Infection rates/100 patient-years for eculizumab vs placebo in NMOSD and gMG, respectively, were: no IST, 176.1 vs 192.2 and 236.8 vs 305.6; 1 IST, 171.5 vs 154.1 and 228.8 vs 253.1; 2 ISTs, 186.7 vs 238.2 and 170.5 vs 192.5; ≥3 ISTs (gMG only), 97.5 vs 100.1. Serious infection rates/100 patient-years were: no IST, 2.3 vs 8.0 and none observed; 1 IST, 11.2 vs 7.0 and 16.2 vs 34.5; 2 ISTs, 14.8 vs 47.6 and 13.4 vs 24.1; ≥3 ISTs (gMG only), 13.9 vs 0.0. One patient with gMG (2 ISTs) had meningococcal meningitis that resolved with antibiotics and eculizumab was resumed.

Conclusions

In AQP4+ NMOSD and AChR+ gMG, infection rates were similar in eculizumab and placebo groups, regardless of concomitant IST, and were consistent with eculizumab's established safety profile. This study was funded by Alexion Pharmaceuticals, Inc.

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Severe Hashimoto's encephalopathy debuted with delusion of COVID-19 contamination: A case report

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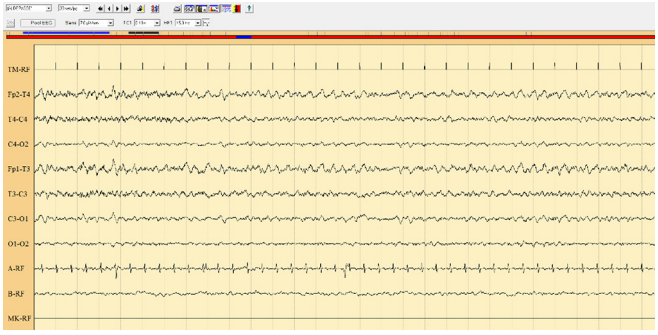
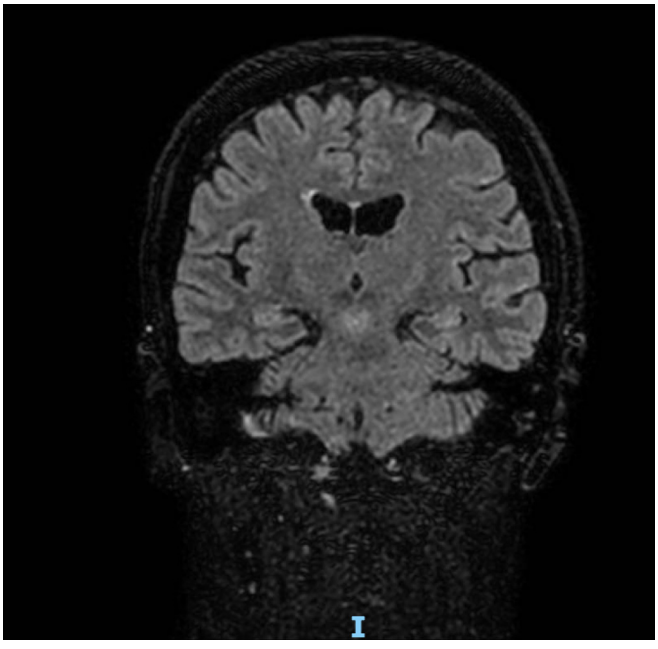
Background and aims

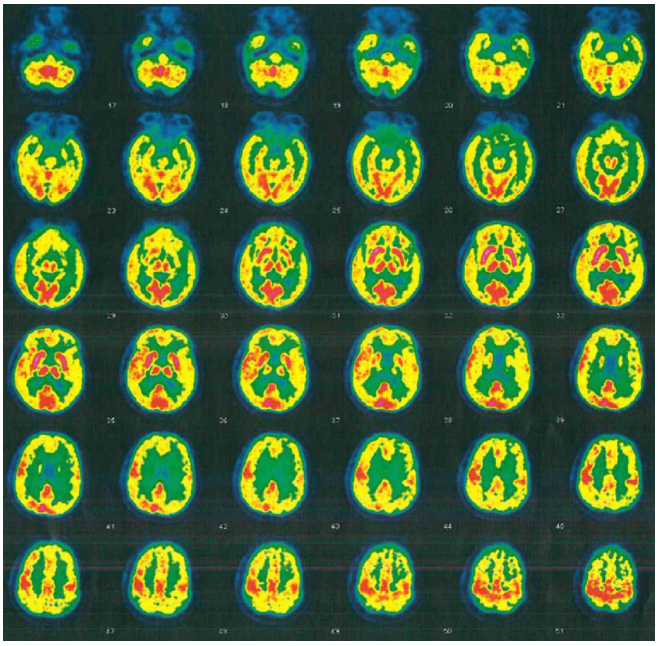
Hashimoto's Encephalopathy (HE), is a rare autoimmune disorder to be considered in patients with neuropsychiatric symptoms accompanied by normal or nonspecific brain MRI, LCR findings and thyroid function or mild hypothyroidism, increased serum levels of thyroid peroxidase antibodies (TPOAb), and response to steroids. We describe the case of a 68-years old caucasian female without previous psychiatric history conducted to Emergency Care in February 2020 for recently debuted confusion, visual hallucinations and cleanliness delusions in response to SARS-COV2 pandemic.

Methods

We found TSH elevation (38,05 μUI/mL) with negative brain CT. EEG presented diffuse 2–4 Hz activity. LCR findings were negative for infections. Brain MRI was inconsistent. We started levothyroxine replacement. At EEG reappeared an alpha activity. TSH began to descend. TPOAb were at a very high titer (>600 U/mL). We started oral prednisone with initial neuropsychiatric improvement. Patient rapidly got again worse in psychiatric aspects and presented also extrapiramidal signs. Antibodies against cell-surface, synaptic and onconeural proteins were absent. CT total body was inconsistent. 18PDG-PET was negative. Levels of beta amyloid, 14–3-3protein, tau and phospho-tau in LCR were normal. Brain PET demonstrated an hypometabolism in frontotemporoparietal bilateral regions. At psychometric tests patient presented a multi-domain disorder. We treated her with high dose intravenous corticosteroids followed by an immunoglobulins cycle.







Results

At follow-up extrapiramidal and psychiatric disorders got slowly better. At control psychometric tests and cerebral PET patient improved. She is performing 18PDG- PET every 6 months to exclude an underlying paraneoplastic syndrome.

Conclusions

Hashimoto's Encephalopathy seems to respond to steroids and immunoglobulins therapy.

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Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (clippers) after COVID-19

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Background and aims

SARS-CoV-2 infection is now known to be associated with a wide spectrum of neurological autoimmune syndromes, in some cases responding to immunotherapies, arising during or after the infection. Whether molecular mimicry or other immune stimulation may induce an aberrant delayed autoimmune response is still to be established.

Methods

Case Report.

Results

A 71 year-old man with no previous medical history apart from mild COVID-19 pneumonia 3 month earlier, sought medical attention for a subacute onset of diplopia in left gaze, general malaise and fatigue. MRI was characterized by bilateral FLAIR hyperintensities with punctate, perivascular and confluent post-gadolinium enhancement in the pons, mesencephalon, hypothalamus, internal capsules and right hippocampus. Repeated cerebrospinal fluid analysis were normal (2 cells/μL), with no evidence of oligoclonal bands or atypical cells. Screening panel for autoimmune and infectious aetiologies was negative. Whole-body contrast-enhanced CT was unremarkable. Stereotactic temporal lobe brain biopsy showed aspecific chronic lymphocytic perivascular inflammation. Partial spontaneous remission of symptoms occurred within few weeks. He was then treated with intravenous high-dose methylprednisolone with almost complete enhancement regression on MRI. Collected data were suggestive of CLIPPERS with diffuse bilateral supratentorial involvement. The patient started daily oral steroid tapering and monthly cycles of intravenous cyclophosphamide with persistent clinical and neuroradiological stability.

Conclusions

CLIPPERS is a rare diagnosis and to the best of our knowledge, this is the first time it was reported after COVID-19 disease. Even though a case report is not enough to suggest a causal link, future reports could support this possibility.

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A rare cause of drug induced mononeuropathy

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Background and aims

Drug-induced mononeuropathy is a common and painful condition seen with use of chemotherapeutic agents, antimicrobials and anticonvulsants. It takes weeks to months to set in. Churg Strauss Syndrome (CSS) is primarily a disease of middle age and its presentation in childhood is rare. We hereby present a rare case of an adolescent female presenting with mononeuropathy which later turned out to be a part of eosinophilic granulomatosis with polyangiitis or CSS precipitated by prolonged use of leukotriene receptor antagonist.

Methods

16 year old female child presented with complaints of rash and swelling over legs and arthralgia for last 7 days. She also had severe pain and numbness in right foot. Child had been diagnosed with bronchial asthma one year backfor which she took montelukast. A nerve conduction velocity study was done which showed evidence of sensory neuropathy in right sural nerve. The complete blood counts consistently showed more than 70% eosinophils. Total IgE levels were elevated and HRCT chest showed bronchiectasis. Skin biopsy results revealed leukocytoclastic vasculitis with perivascular eosinophils. With the diagnosis of CSS child was started on oral steroids and montelukast was discontinued. pANCA and cANCA levels were negative.