infradiaphragmatic hypermetabolic adenopathies. Biopsy of an adenopathy was performed objecting non-caseating granulomas consistent with the diagnosis of probable neurosarcoidosis. Moreover, the patient explained weakness and muscle fatigue since she was 18 years old, which was observed at proximal limb muscles. Acetylcholine receptor antibodies were detected and electromyographic study showed a decremental response to repeated nerve stimulation.

Conclusions

It is well known that autoimmune disorders may coexist in some patients. Neurosarcoidosis and Myasthenia Gravis are two rare diseases with different pathogenesis. Although their coexistance could be coincidental it may also suggest immunologic mechanisms triggering the occurrence of these diagnoses together.

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Autoimmune Encephalitis Misdiagnosis: A Review of Reported Cases

Maria Daniela Orellana Zambrano, Gregory Day, Elia Sechi, Alfonso Lopez

Objective

To identify autoimmune encephalitis (AE) mimics and clinical features reported in the literature.

Background

Recent evidence suggesting that AE is as frequent as infectious encephalitis has increased awareness and testing for immune-mediated causes of neurological impairment. Consistent with this theme, several publications have focused on patients in whom a diagnosis of AE was initially overlooked. On the contrary, AE remains a rare diagnosis in clinical practice, opening up the possibility for symptoms, signs, and test findings associated with other etiologies to be misattributed to AE.

Design/Methods

Case reports published in PubMed in English language before 04/2022 were reviewed. Cases in whom AE was clearly suspected during the diagnostic work-up or misdiagnosed were included.

Results

A total of forty-five patients with a final diagnosis different from AE were included from 40 reports. Median age was 52 (range 5-86) years; 30/45 (67%) were male. Twenty-eight patients fulfilled the criteria for possible AE (62%), five for definite AE (11%), and twelve neither (27%). Features suggestive of AE were acute/subacute altered mental status (ranging from abnormal behavior to coma), (82%); new-onset refractory status epilepticus, (7%); CSF pleocytosis (42%) or oligoclonal bands (9%), and apparent response to immunotherapy (38%). In 26 cases, imaging corresponded to the anatomical classification of limbic encephalitis, 15 had one or more cortical/subcortical T2-abnormalities, one meningeal involvement, one brainstem involvement, and two had normal MRI. In 12 patients, clinically not relevant neural autoantibodies were detected in serum and/or CSF, including GAD, Anti-Zic4, CASPR2, VGKC, anti-N-type calcium channel antibody, anti-LGI1, and GQ1B. We identified four common AE mimic categories: neoplasms (15 patients), infectious diseases (9 patients), genetic diseases (9 patients), and neurodegenerative diseases (7 patients). Five patients had other etiologies.

Conclusions

Despite well-defined clinical diagnostic criteria, the misdiagnosis of AE encompasses atypical presentation of common disorders and less likely rare diagnoses.

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The Complicated Course of a Patient With Faciobrachial Dystonic Seizures Associated With LGI1-Antibody Limbic Encephalitis

Shirin Sadeghpour, Dilasha Neupane, Mariam Mouti, Jeffrey Clark

Objective

Highlighting diagnostic and treatment challenges of Faciobrachial Dystonic Seizures (FBDS) associated with LGI1-antibody limbic encephalitis (LE)

Background

Anti-LGI1 LE presents with FDBS as its hallmark: brief, recurrent, contractions of facial and upper limb muscles. Patients have associated cognitive decline and psychiatric disturbance. Temporal lobe involvement is often found on MRI.

Design/Methods

NA.

Results

This 85-year-old female presented with a 2-week history of involuntary "twitching" in the face and arms.Family reported that she hadalsobeen uncharacteristically quiet.Neurological exam and a CT headwere normal.rEEG showed no epileptiform activity.One monthafter onset, episodes became longer and more frequent.Observation during outpatient evaluation led to consideration ofFDBS based on semiology. MRI revealedT2/flairhyperintensity in medial temporal lobes consistent with LE.InpatientcEEG was obtained: 26 episodes were marked, with no ictal EEGcorrelate seen.IVIG and methylprednisolone were started.InitialCSF studies were unremarkable andencephalitis/meningitis panelwas negative. Autoimmuneand paraneoplastic encephalopathy panel later revealed LGI-1antibodiesin the CSF. Chest and abdominopelvic CT were unrevealing of underlying malignancy. Whilereceiving methylprednisolone and IVIG, she developed impaired orientation, hallucinations, and agitation. A 5-day course was completed but with worsening mentation and limited improvement in FBDS.Thus,a decision was made to initiate PLEX (now 1.5 months fromsymptomonset). FBDS episodes resolved withPLEX and mentation improved, butshe development bleeding and retroperitoneal hematoma requiring transfusion, delaying completion of PLEX. Her course was further

complicated by a UTI and delirium. PLEX was restarted afteroneweek andhermental status improved, reportedly 80% back to baseline after five sessions.

Conclusions

The main diagnostic challenge for LE is recognizing its unique hallmark, FBDS, often associated with insidious decline in cognition. FDBS may have no ictal correlate on EEG. Early diagnosis and management prevents long term disability - mainly cognitive. LE is commonly treated with immunotherapy, corticosteroids and PLEX. Monitoring response is challenging as patients are commonly older with multiple comorbidities: steroid-induced psychosis and delirium may complicate evaluation and treatment side effects limit options.

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Autoimmune Encephalitis-A Diagnostic Challenge for the Neurologist

Laxmi Khanna, Mandaville Gourie-Devi, Ritu Verma

Objective

Autoimmune encephalitis, is a clinically challenging entity with varied neurological presentations. As autoimmune serology is negative in over 50% cases, our objective was to prioritise the use of electroencephalography supported by MRI Brain or PET–CT imaging to make a definitive diagnosis of autoimmune encephalitis.

Background

Autoimmune encephalitis is the consequence of an antibody mediated neuronal damage caused by cell surface antigens. However, antibody assays can be negative in early stages of the disease. Hence, we suggest the use of electroencephalogram along with MRI brain imaging or PET-CT scans to avoid diagnostic delays.

Design/Methods

During a span of four years [2018–2022] a retrospective review of the case records of 50 patients of autoimmune encephalitis and 50 patients of non-autoimmune encephalitis were compared. Besides clinical examination, serum and cerebrospinal fluid viral and autoimmune antibody assay, electroencephalogram, Magnetic Resonance and FDG-PET- CT scans were used to confirm the diagnosis.

Results

60% patients were seronegative and 40 % were seropositive in the autoimmune group while 90% were seropositive and 10% seronegative in the non-autoimmune encephalitis group. Electroencephalography was abnormal in all cases of autoimmune encephalitis 100% [50/50] and in 80% [40/50] cases of viral encephalitis. In autoimmune encephalitis, MRI Brain revealed evidence of limbic encephalitis in 80% cases and FDG PET-CT scans were abnormal in the remaining 20%. In non-autoimmune encephalitis, MRI Brain was abnormal in 60% cases and FDG PET-CT was abnormal in 10%.

Conclusions

In seronegative autoimmune encephalitis, electroencephalographic abnormalities supported by MRI Brain imaging and PET-CT scans enabled early diagnosis in 100% cases [p value <0.001]. While in non-autoimmune encephalitis serology with electroencephalography, MRI Brain and PET-CT scans were diagnostic in 80% cases [p value <0.001]. However, as the sample size is small further studies are needed to confirm these findings.

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Brains on Fire: Patient Outcomes and Quality of Life Following Autoimmune Encephalitis

Ava Easton

Objective

To present and improve understanding of patient outcomes and quality of life post-autoimmune encephalitis.

Background

Patient outcome following encephalitis and in particular following autoimmune encephalitis is not well understood. It is only over the last 15 years that we knew, became able to test for, and identify some of these autoimmune causes of encephalitis and so there currently only a small and emerging literature about patient outcomes. Many papers that talk about or refer to patient outcome look quite early on in the patient pathway and often when they talk about outcome they mean immediate clinical outcome and not how patients are, if they survive, several months or years down the line. Therefore we don't actually have a good handle yet on autoimmune patient outcomes nor their quality of life. Yet, most papers agree that much more needs to be done to assess long-term outcome and quality of life in autoimmune encephalitis patients. One further point of importance is that some complications of AE might not appear until several months or years down the line.

Design/Methods

Review of the literature accompanied by first-hand patient video testimony providing rich insight into long term outcomes and impact on quality of life post-autoimmune encephalitis.

Results

Patient outcomes post-AE can be life-changing and in some instances may occur several months and years post-acute illness. These outcomes and quality of life influence how patients understand and make sense of their experience; how they engage with recovery and rehabilitation. A range of poorly understood factors influence patient quality of life post-AE. These include a lack of easily understood index event, lack of community and collective understanding, fears of relapse, post-ICU PTSD, and a lack of accurate information and understanding of their outcomes pre and post-discharge.

Conclusions

Improved understanding of patient outcomes post autoimmune encephalitis can improve patient care and engagement.

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