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Diagnostic and Therapeutic Updates of Autoimmune Encephalitis: a Narrative Review

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Abstract:
Autoimmune encephalitis (AE) is a diverse collection of immune-mediated neurological illnesses that cause inflammation of the brain parenchyma by autoantibody-mediated pathways. AE encompasses a group of non-infectious immune-mediated inflammatory disorders of the brain parenchyma often involving the cortical or deep gray matter with or without involvement of the white matter, meninges, or the spinal cord. Suggested mechanisms that may trigger AE include tumors (paraneoplastic), infections (para-infectious), or it may be cryptogenic. This review summarises current knowledge about the pathogenesis, clinical presentation, diagnostic techniques, and treatment methods linked with AE.The wide range of autoantibodies targeting neuronal surface antigens, such as NMDA receptor, LGI1, and CASPR2 antibodies, is highlighted, as is their relationship to specific clinical characteristics. The paper discusses diagnostic techniques that include cerebrospinal fluid (CSF) analysis, magnetic resonance imaging (MRI), and autoantibody detection, as well as new biomarkers. The review emphasises developments in immunotherapy, such as corticosteroids, plasma exchange, intravenous immunoglobulin, and targeted monoclonal antibodies, and their impact on prognosis.Early diagnosis,differential diagnosis, and long-term management challenges are discussed, as well as future research opportunities for unravelling pathogenic pathways and optimising personalised treatment approaches. Understanding the complicated immunopathology of AE is critical for earlier diagnosis and treatment, which ultimately improves neurological outcomes.

Introduction:

Autoimmune encephalitis (AE) is a broad category of inflammatory brain illnesses defined by the immune system's abnormal attack on neuronal cell surface or intracellular antigens.[1] Previously thought to be rare, recent breakthroughs have shed light on its larger clinical spectrum, emphasising the significance of early identification and treatment. AE can cause a variety of neuropsychiatric symptoms, such as seizures, cognitive impairment, behavioural abnormalities, and movement difficulties, which frequently mirror viral or underlying mental diseases[2] . The discovery of particular autoantibodies has transformed understanding of its aetiology, allowing for targeted therapy that greatly improve patient outcomes. Despite these developments, hurdles persist in early detection, differential diagnosis, and comprehension of the underlying mechanisms.[4]This review aims to provide a comprehensive overview of the current knowledge on autoimmune encephalitis, including its epidemiology, pathophysiology, clinical features, diagnostic approaches, and management strategies, emphasising recent developments and ongoing research in this rapidly changing field.[4]

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AE is a group of syndromes with paraneoplastic or immunological etiology, usually characterized by subacute onset of memory impairment, confusion, and frequent seizures[5].An increasing number of types of AE is being identified across a broad clinical spectrum, ranging from epilepsy to movement disorders to psychosis. Autoimmune (immune = mediated) encephalitis includes diseases related to antibodies against neuronal cell-surface synaptic proteins as well as those linked with antibodies against intracellular neuronal proteins.[6] Although both types of AE can present as paraneoplastic symptoms of an underlying cancer, the prevalence of cancer connection differs according to the autoantibodies. Most neural antibodies associated with paraneoplastic encephalitis (e.g., >70 percent of patients have a cancer association) target intracellular neuronal proteins (onconeuronal proteins).[7,]The discovery of new antibodies and associated disorders has led to the development of autoimmune neurology, a rapidly evolving discipline.[8]

AE is a challenging clinical diagnosis since numerous kinds of autoimmune and infectious encephalitis have comparable clinical, imaging, and laboratory findings. Patients usually come to the doctor with a memory impairment lasting days or weeks. Anamnesis and neurological examination may provide some or no clues.[9]

Epidemiology:

In Western nations, the prevalence of encephalitis in adults varies between 0.7 and 12.6 cases per 100,000.[10] Most occurrences of encephalitis are caused by infections.[11] An autoimmune aetiology is suggested by the growing number of individuals in clinical and research settings who have antibodies to neuronal cell-surface proteins.[11] The frequency of AE (0.8 per 100,000) is comparable to that of infective encephalitis (1 per 100,000), per a recent epidemiological research conducted in Olmsted County.[12]

Pathophysiology:

There are several clinical and pathologic subtypes of AE. Classic paraneoplastic diseases brought on by intracellular antigen antibodies, such anti-Hu, fall within the first type.[26] These disorders, which are marked by T-cell responses that target neurones, are closely linked to cancer. Autoantibodies against extracellular epitopes of channels, receptors, and other proteins, including the anti-N-methyl-D-aspartate receptor, fall under the second category[27] Autoantibodies to intracellular synaptic proteins, such those against glutamic acid decarboxylase 65 (GAD65), are indicative of an intermediate category of diseases; this is not entirely obvious because it does not neatly fit into any of these groups.Other forms of AE that do not have particular antigens, like acute disseminated encephalomyelitis or lupus cerebritis, are uncommon neurological disorders that typically develop following exposure to an autoimmune, bacterial, or viral agent. Common symptoms of the last category of acute disseminated encephalomyelitis include headache, disorientation, weakness, and paraesthesia. They produce inflammation in the central nervous system.

Clinical Presentations:

A subacute (days to a few weeks) progressive loss of consciousness, frequently accompanied by fluctuations or a decline in cognitive function, is the usual manifestation of AE [4].Memory, particularly the capacity to remember new information, may be impaired in the early stages of the illness. Psychiatric symptoms, such as psychosis, violence, terror, panic attacks, obsessive behaviours, and improper sexual activity, are very commonly seen in the early stages[5] Seizures and mobility abnormalities are also prevalent in AE. Ataxia and gait abnormalities are caused by a specific kind of cerebellitis, with AE often exhibiting vertigo and nystagmus.

Diagnostic criteria for possible AE

Diagnosis can be made when all 3 of the following criteria have been met:

1. Subacute onset (rapid progression of <3 months) of working memory deficits (short-term memory loss), altered mental status, or psychiatric symptoms
2. At least one of the following:
   - New focal central nervous system findings;
   - Seizures not explained by a previously known seizure disorder;
   - Cerebrospinal fluid pleocytosis (white blood cell count of more than 5 cells per mm3);
   - Magnetic resonance imaging (MRI) features suggestive of encephalitis.
3. Reasonable exclusion of alternative causes.

Classifications:

A. Anatomical classification :

Cortical/subcortical, limbic, diencephalic, striatal, cerebellar, brainstem, encephalomyelitis, meningoencephalitis, and combined.

B. Etiological classification:

Idiopathic, paraneoplastic, post-infectious, iatrogenic (e.g., in the setting of immune checkpoint inhibitors or other immune-modulating agents).

C. Serological classification:

Antibodies to surface antigens and other antigens with high clinical relevance (e.g., GFAP, NMDAR, LGI1, AMPAR, contactin-associated proteinlike 2 [CASPR2], dipeptidyl-peptidase-like protein 6 [DPPX], GABAR A/B, AQP4, glycine receptor, MOG), antibodies to surface antigens with low clinical relevance (e.g. VGCC, VGKC), antibodies to intracellular antigens (classical onconeural antibodies), seronegative AIE.

In most cases of AIE, there is a widespread immune reaction that affects the brain, spinal cord, peripheral nervous system, and meninges, leading to a poly-syndromic presentation that varies based on the location and extent of inflammation [4]. There are several clinical-anatomical syndrome categories in AIE (Table 1).

Table 1: Clinical-anatomical syndrome categories in AE

Cortical/subcortical encephalitis	PCA-2 (MAP1b), NMDAR, GABA A/B R,DPPX, MOG antibodies. The most common presentation iscognitive and seizure presentation
Limbic encephalitis	The Hu, CRMP5/CV2, Ma2, NMDAR,AMPAR, LGI1,CASPR2,GAD65, GABABR, DPPX, mGluR5,AK5, neurexin-3á antibodies. The mostcommon presentation is cognitive,psychiatric, and epileptic presentation
Striatal encephalitis	CRMP5/CV2, DR2, LGI1, PD10A, NMDAR, antibodies. The most common is
Diencephalic encephalitis	movement disorder presentation Ma 1–2, AQP4, DPPX, IgLON5 antibodies. The most common is sleep disorder presentation and autonomic presentation
Brainstem encephalitis	Ri, IgLON5, Ma 1–2, DPPX, KLHL11, MOG, AQP4, and GQ1b antibodies. The most common presentation is craniobulbar presentation, movement disorder presentation, and cognitive presentation.
Cerebellitis or cerebellar degeneration	Hu, VGCC, CASPR2, Ri, KLHL11, Yo, NIF, Tr, mGluR1, GAD65 antibodies. The most common presentation is ataxic presentation.
Meningoencephalitis	It can be seronegative AIE or linked to GFAP antibody. The most common presentation is meningeal presentation, seizure presentation, cognitive presentation
Encephalomyelitis	Amphyphysin, GAD65, PCA-2 (MAP1B), glycine receptor, GABA A/B R, MOG, CRMP5/CV2, DPPX, AQP4, antibodies. The most common presentation is spinal presentation, movement disorder presentation including PERM and SPS, opticospinal presentation
Autonomic neuropathy/ ganglionopathy	Hu, anti-ganglionic AchR, CRMP5 antibodies. The most common is autonomic presentation.
Neuropathy/ neuronopathy	NIF155, Hu, amphiphysin, PCA-2 (MAP1B), CONTACTIN1, CRMP5, CASPR1, CASPR2 antibodies. The most common is sensorimotor and ataxic presentation.
Neuromuscular junction dysfunction	AchR, VGCC antibodies. The most common is myasthenic presentation

NMDAR: N-Methyl D-aspartate receptor, LGI1: Leucine-rich glioma inactivated, AQP4: Anti-aquaporin-4, MOG: Myelin oligodendrocyte glycoprotein, GFAP: Glial fibrillary acidic protein, CASPR2: Contactin-associated protein-like 2, DPPX: Dipeptidyl-peptidase-like protein 6, GAD65: Glutamic acid decarboxylase 65.

Diagnostic Approach:

The first step in diagnosing AIE is to conduct a thorough history and clinical examination. AIE is frequently associated with acute or subacute onset[10]. Chronic manifestations are seen in CASPR2, LGI1, DPPX, and glutamic acid decarboxylase 65-antibody encephalitis[11]. A recurring course may indicate an autoimmune cause. Inadequate treatment or abruptly stopping immunotherapy might lead to AIE relapses. Consider vascular aetiology for hyperacute presentations. Chronic presentations may indicate neurodegenerative illness or other underlying causes. Idiopathic AIE often has a single phase, but paraneoplastic AIE has a progressive course. AIE can be caused by immune-modulating medications such TNFá inhibitors and ICIs, as well as herpes simplex virus encephalitis[1].

Brain imaging and cerebrospinal fluid (CSF) analysis are typically performed after taking a patient's history and clinical assessment. The initial diagnostic phase is determining if there is multifocal or focal brain pathology. MRI is the most prevalent diagnostic technique for this[12].

Electroencephalogram (EEG) is a diagnostic technique for MRI-negative patients, encephalopathy, and seizures. PET, is utilized as a diagnostic technique in cases of ambiguous diagnosis or negative MRI results[12].

The second phase in diagnosing involves blood tests for serum neuronal antibodies and a lumbar puncture. Cerebrospinal fluid should be evaluated for infections, inflammatory markers (IgG index and oligoclonal bands), and occasionally cytology. The second phase in diagnosing involves blood tests for serum neuronal antibodies and a lumbar puncture[13].

Cerebrospinal fluid should be evaluated for infections, inflammatory markers (IgG index and oligoclonal bands), and occasionally cytology. In the second diagnostic phase, instruments are employed to eliminate other possible causes and confirm the inflammatory aetiology[14].

If the diagnosis is still unknown after the first two phases, a brain biopsy may be used to confirm the cause and guide treatment decisions.Biopsy results are rarely definitive in autoimmune cases. Overall, the clinical impact of a biopsy performed for suspected encephalitis is minimal, with only about 8% of cases showing clear benefits[15].

The third diagnostic stage involves screening for related neoplasms.Typically, computed tomography is the initial screening method for neoplasms, with other screening modalities utilised as needed until a neoplasm is identified.If the clinical picture indicates anti-NMDAR encephalitis or a specific tumour, a tailored screening technique may be performed (e.g., pelvic ultrasound and additional diagnostic tests).Neuronal antibodies to intracellular antigens, such as Hu, are more frequently found in cancer patients than in autoimmune disease patients[16] . For instance, in patients with small-cell lung cancer without anti-Hu neurological symptoms, a low-titer serum Hu response is a common diagnosis associated with ovarian teratoma, assessed using pelvic MRI or ultrasonography. Testicular ultrasonography is essential in young men with a diagnosis of Ma2 or suspected cases, while for women with anti-Yo antibodies, mammography or breast MRI, pap smears, and pelvic imaging may be most beneficial[17].

Therapeutically challenges:

Initiating immunotherapy after excluding infectious aetiologies based on CSF data and when AE is strongly suspected is recommended by the 2016 AE clinical criteria and retrospective studies . Early and aggressive immunotherapy improves survival in AE patients, while delaying treatment until a positive antibody confirms AE is risky and impracticable[1,18]. Titulaer et al. studied the long-term outcomes of anti-NMDA receptor encephalitis patients, analysing therapy and prognostic variables.[18] Titulaer et al. conducted a multi-institutional observational cohort study from 2007 to 2012 to assess the presence of NMDAR antibodies in the cerebrospinal fluid (CSF) and serum of encephalitis patients. The study comprised 577 people who tested positive for NMDAR antibodies.[18] The median age was 21 years, with a range of 8 months to 85 years. A total of 211 patients were under the age of 18. The modified Rankin scale was used to examine study participants at 6 time intervals, including symptom onset, months 4, 8, 12, 18, and 24.[19]

Treatment options included first-lineimmuno-therapy (steroid, IVIG, and plasmapheresis), second-line immunotherapy (rituximab and cyclophosphamide), and tumour removal. [20]

Immunotherapy is effective in treating most people with anti-NMDAR encephalitis. When first-line treatments do not work, second-line immunotherapy can be successful. Some patients in this sample required up to 18 months for recovery[21]. There are no clinical trials comparing the acute treatment of AIE with other immunotherapy options. Therefore,

the initial therapy may be chosen based on clinical presentation, comorbidities, and anecdotal evidence.[22]

Current therapeutic options include corticosteroids, intravenous immunoglobulin, plasma exchange, rituximab, and cyclophosphamide[23].

Treatment response and prognosis data for anti-NMDAR encephalitis are generally available[24]. Empirical treatment with intravenous methylprednisolone at a dose of 1 g/day for 3-7 days can provide initial immunosuppressive and anti-inflammatory effects in AE patients[1]. Corticosteroid-responsive patients have FBDS indicating LGI1-antibody encephalitis and MRI showing demyelinating patterns[24].Paraneoplastic AE with classical onconeuronal antibodies responds well to cancer therapy and is resistant to immunosuppression. Corticosteroids are the preferable alternative for immunosuppression over plasma exchange (PLEX) or intravenous immunoglobulin (IVIg) in cases of suspected or confirmed paraneoplastic AE with classical onconeuronal antibodies. Corticosteroids can decrease T-cell overactivity, which is the pathologic hallmark of ICI-associated immunological adverse events. However, in some situations, second-line therapy may be required[21].IVIg at a dose of 2 g/kg over 2-5 days is an alternative for rapid immunomodulation in instances with known antibody-mediated illness (e.g., NMDAR-antibody encephalitis) and when corticosteroids are not recommended[18]. A randomised blinded research found that IVIg was more effective than placebo in reducing seizures in a limited group of patients with CASPR2 andLGI1 antibody AE[13]. Furthermore, IVIg may not be useful for paraneoplastic AIE caused by antibodies against intracellular antigens, as these are cell-mediated rather than antibody-mediated. IVIg can raise thromboembolic risk and aggravate hyponatraemia, potentially leading to cerebral oedema and poor mental state[14].

Plasma exchange PLEX (5-10 sessions every other day) is a viable alternative for acute immunomodulation when corticosteroids are ineffective or contraindicated. Immunomodulation may be faster for patients with severe or fulminant presentations. Abboud et al. found comparable outcomes in their retrospective evaluation of additional antibody-mediated diseases, including NMOSD[25].

DeSena et al. conducted a modest retrospective investigation on 14 patients with NMDAR-antibody encephalitis. This study found that patients who received both PLEX and corticosteroids experienced greater improvement in their modified Rankin score compared to those who only received corticosteroids [25]. In AE instances with NMOSD or central demyelination, PLEX may be especially beneficial.Although there is limited evidence, combination first-line therapy can be utilised from the start for severe instances such as NMDAR-antibody encephalitis, NORSE, and severe dysautonomia. After 2-4 weeks of optimised first-line therapy, second-line medicines are employed if no significant clinical or radiological response is observed. Both rituximab and cyclophosphamide have shown promising effects as second-line rescue therapies for AE[11]. In specialised centres, individuals over 16 years are typically prescribed rituximab (375 mg/m2) weekly for 4 weeks and cyclophosphamide (750 mg/m2) for 6 months[24].

Therapeutic agents used for autoimmune encephalitis.

Treatment	Regimen
First line immunotherapy	
Methylprednisolone	1 g daily, for 3 to 5 days
Intravenous immunoglobulin	2 g/kg over 5 days (400mg/kg/day)
Plasma exchange	1 session ever other day for 5–7 cycles
Second line immunotherapy	
Rituximab	375 mg/m3 weekly IV infusion for 1 week
Cyclophosphamide	750 mg/m3 monthly for 3 to 6 months
Tocilizumab	Initially 4 mg/kg, followed by an increase to 8mg/kg monthly based on clinical response.
Low dose interleukin (IL)-2 2 (aldesleukin)	1.5 million IU/day, 4 SC injections with 3 weeks interval
Steroid sparing agents	
Azathioprine	Initially 1 to 1.5 mg/kg once daily or divided twice daily, target 2–3 mg/kg/day
Mycophenolate mofetil	Initially 500 mg twice daily, target 1000 mg twice daily

Prognosis:

When immunotherapy is started has a significant impact on long-term functional results. Even in cases of severe immunosuppression, autoantibodies that target intracellular antigens usually do not respond, therefore stabilising neurological impairment is frequently seen as a positive accomplishment[12.]In general, patients who have autoantibodies against cell-surface antigens fare better. One of the main causes of death for people with paraneoplastic CNS diseases is the advancement of the underlying cancer[26.]Younger patients should only get rituximab[1].Early treatment leads to improved prognosis[25].

Patients with AE experience varying treatment responses and relapse rates. Half of patients with anti-NMDAR encephalitis do not respond to first immunotherapy and may need second-line treatment, with 12% experiencing relapses.[26] Relapses can occur years after the initial episode in 10% of individuals with anti-CASPR2 encephalitis and 31% with anti-LGI1 encephalitis. Anti-LGI1 encephalitis causes 33% of patients to become handicapped, primarily owing to memory issues.[27]

Conclusion:

AIE can cause a range of symptoms to appear suddenly or gradually. Early detection of underlying autoimmune aetiology is crucial in situations of sudden altered mental state or clinical encephalitis. Physical examination and clinical presentation are crucial for accurately diagnosing AIE.Early therapy improves prognosis and prevents serious consequences. The mechanisms for activation and autoimmune response in the CNS remain unclear. Further research is needed to better understand how immune systems impact nervous system activities. AIE patients may be seronegative, hence relying just on antibody testing is not sufficient for diagnosis. The treatment can be adjusted and analysed based on antibody results.

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