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Search details
Non-paraneoplastic limbic encephalitis

Resources searched
NHS Evidence; TRIP Database; Cochrane Library; CINAHL; EMBASE; MEDLINE; Google Scholar

Database search terms: “non-paraneoplastic limbic encephalitis”; LIMBIC ENCEPHALITIS; “paraneoplastic limbic encephalitis”; “nonparaneoplastic limbic encephalitis”; “non paraneoplastic limbic encephalitis”; paraneoplastic; PARANEOPLASTIC SYNDROMES; limbic adj2 encephalitis; non-paraneoplastic; nonparaneoplastic;

Google search string: (nonparaneoplastic OR non-paraneoplastic) limbic encephalitis; (“without paraneoplastic” OR "not paraneoplastic") limbic encephalitis

Summary
There has been a lot of research on non-paraneoplastic limbic encephalitis published in the last twelve years. I have concentrated on the term “non-paraneoplastic limbic encephalitis” rather than looking for types of limbic encephalitis that are not paraneoplastic. If you require a broader search to cover this research, please let me know.

Guidelines
Encephalitis Society
Limbic Encephalitis 2007

Evidence-based reviews
None found.
1. Autoimmune-mediated encephalitis.

Author(s): Demaerel P, Van Dessel W, Van Paesschen W, Vandenberghe R, Van Laere K, Linn J

Citation: Neuroradiology, November 2011, vol./is. 53/11(837-51), 0028-3940;1432-1920 (2011 Nov)

Publication Date: November 2011

Abstract: Autoimmune-mediated encephalitis may occur as a paraneoplastic or as a non-paraneoplastic condition. The role of neuroimaging in autoimmune-mediated encephalitis has changed in the last decade partly due to improvements in sequence optimisation and higher field strength and partly due to the discovery of an increasing number of antibodies to neuronal cell and cell membrane antigens. Imaging is important since it can support the clinical diagnosis particularly in the absence of antibodies. Structural imaging findings can be subtle and are usually best seen on FLAIR images. A progressive as well as a relapsing-remitting course can be observed. Autoimmune-mediated encephalitis is classically linked to involvement of the hippocampus and amygdala, but extensive changes in the temporal cortex, basal ganglia, hypothalamus, brain stem, frontal and parietal cortex are not unusual. This report is based on a review of the literature (except the literature in Japanese) and own findings in patients with autoimmune-mediated encephalitis.

Source: MEDLINE

Full Text: Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

2. Limbic encephalitis presenting as a post-partum psychiatric condition

Author(s): Gotkine M., Ben-Hur T., Vincent A., Vaknin-Dembinsky A.

Citation: Journal of the Neurological Sciences, September 2011, vol./is. 308/1-2(152-154), 0022-510X;1878-5883 (15 Sep 2011)

Publication Date: September 2011

Abstract: Objective: We describe a woman who presented with a psychiatric disorder post-partum and subsequently developed seizures and cognitive dysfunction prompting further investigation. A diagnosis of limbic encephalitis (LE) was made and antibodies to voltage-gated potassium channel complex (VGKC) detected. These antibodies are found in many non-paraneoplastic patients with LE. Although antibody-mediated conditions tend to present or relapse post-partum, VGKC-LE in the post-partum period has not been described. Design: Case report. Results: Clinical and imaging data were consistent with limbic encephalitis. High titres of anti-VGKC-complex antibodies confirmed the diagnosis of VGKC-LE. Conclusion: The similarities between the psychiatric symptomatology of VGKC-LE and post-partum psychiatric disorders raise the possibility that some instances of post-partum psychiatric conditions are manifestations of immune-mediated, non-paraneoplastic LE. 2011 Elsevier B.V. All rights reserved.

Source: EMBASE

Full Text: Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

3. Non-paraneoplastic limbic encephalitis with antibodies to glutamic acid decarboxylase. Long-term treatment with plasma exchange and steroids

Author(s): Markakis I., Alexopoulos H., Katsiva V., Lyrakos G., Gkekas G., Dalakas M.

Citation: European Journal of Neurology, September 2011, vol./is. 18/(578), 1351-5101 (September 2011)
Abstract: Introduction: Non-paraneoplastic limbic encephalitis (NPLE) has been associated with auto-antibodies against various neuronal antigens, mainly voltage-gated potassium channels, N-methyl-D-aspartate, kainate and gamma-amino- butyric acid receptors. Recently, glutamic acid decarboxylase (GAD), an enzyme catalyzing the conversion of glutamic acid to gamma-aminobutyric acid, has also been identified as a target of humoral autoimmunity in a small subgroup of these patients. We present a case of NPLE with anti-GAD antibodies, which showed a positive response to immunotherapy.

Case report: A 48-year-old female was admitted with a 2-year history of anterograde amnesia, depression and generalized seizures. At previous hospitalizations, MRI scans had revealed bilateral lesions of medial temporal lobes, mamillary bodies and cingulate gyri. Extensive investigation for occult malignancy was negative. Screening for anti-neuronal antibodies was also negative. However, enzyme-linked immunosorbent assay revealed high titres of anti-GAD antibodies in serum (37,550U/ml) and CSF (15,400U/ml), with evidence of intrathecal antibody synthesis. The patient received treatment with prednisolone and long-term plasma exchange. During a 12-month followup period she exhibited complete seizure remission and a moderate improvement in memory and visuo-spatial skills. Serum anti-GAD titres showed a parallel decline, falling to 9,600U/ml, 10 months after treatment onset. Conclusion: Our case offers additional evidence that nonparaneoplastic LE should be included in the spectrum of anti-GAD-associated neurological disorders. Although the direct pathogenetic role of these antibodies remains unclear, their early detection is of clinical relevance. They serve as a useful marker to identify a subset of LE patients that may respond to immunoregulatory treatment.

Source: EMBASE

Full Text: Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

4. Immunotherapy-responsive status epilepticus with anti-glycine receptor antibody


Citation: European Journal of Neurology, September 2011, vol./is. 18/(50), 1351-5101 (September 2011)

Publication Date: September 2011

Abstract: Limbic encephalitis (LE) is mainly a non-paraneoplastic immunotherapy responsive disorder that harbours antibodies targeting neuronal surface proteins. Antibodies against glycine receptor (GlyR-ab) have been described in few cases of encephalomyelitis with rigidity. Here we describe a case of GlyR-ab-associated LE characterized by drug-resistant seizures that was responsive to treatment. Case report: A 25-year-old Indian man was hospitalized after presenting with convulsive seizures without full recovery of consciousness between them. A few days before, he had displayed some behavioural changes (apathy and lack of appetite). On admission, EEG showed diffuse slow abnormalities while brain MRI and CSF analysis were negative. Intense sedation was required and antiviral and bacterial drugs were administered. One week later he still had seizures, EEG disclosed right temporal lobe epileptic activity but the MRI was still negative. Once we had excluded other causes and a thorough search for onconeural, NMDAR and VGKC-complex antibodies was negative, a course of high dose steroids was performed followed by IVIG with dramatic benefit on epileptic activity. A brain MRI performed 45 days after admission did show a T2-hyperintense signal in the right hippocampus, perhaps linked to the seizures. A wider autoantibody screening subsequently demonstrated the presence of GlyR-ab in our patient’s serum. Conclusion: This first report of immunotherapy-responsive LE associated with GlyR-ab raises the possibility that this antibody can affect limbic areas, as GlyRs, although prominent in the brainstem and spinal cord, are also expressed in the hippocampus. Further studies will determine GlyR-Ab contribution to the diagnosis of autoimmune encephalitis.

Source: EMBASE
5. Autoantibodies associated with diseases of the CNS: New developments and future challenges

Author(s): Vincent A., Bien C.G., Irani S.R., Waters P.

Citation: The Lancet Neurology, August 2011, vol./is. 10/8(759-772), 1474-4422;1474-4465 (August 2011)

Publication Date: August 2011

Abstract: Several CNS disorders associated with specific antibodies to ion channels, receptors, and other synaptic proteins have been recognised over the past 10 years, and can be often successfully treated with immunotherapies. Antibodies to components of voltage-gated potassium channel complexes (VGKCs), NMDA receptors (NMDARs), AMPA receptors (AMPARs), GABA type B receptors (GABA\textsubscript{B}Rs), and glycine receptors (GlyRs) can be identified in patients and are associated with various clinical presentations, such as limbic encephalitis and complex and diffuse encephalopathies. These diseases can be associated with tumours, but they are more often non-paraneoplastic, and antibody assays can help with diagnosis. The new specialty of immunotherapy-responsive CNS disorders is likely to expand further as more antibody targets are discovered. Recent findings raise many questions about the classification of these diseases, the relation between antibodies and specific clinical phenotypes, the relative pathological roles of serum and intrathecal antibodies, the mechanisms of autoantibody generation, and the development of optimum treatment strategies. 2011 Elsevier Ltd.

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the hippocampal enhancement at MRI. During the following weeks, frequent seizures re-emerged with snorting followed by tonic contractions of facial muscles and bilateral piloerection on upper limbs; these symptoms were associated with generalized fear, anxiety and grimaces. Ictal EEG showed rhythmic spiking in the right temporal area with contralateral diffusion. After re-assessment of the antiepileptic drugs, emotional and pilomotor seizures gradually subsided. Subsequently, the patient started monthly ivcyclophosphamide and showed a progressive improvement of memory impairment.

Discussion: In our patient, VGKC limbic encephalitis was associated with emotional and pilomotor seizures. Ictal pilomotor erection is typically but not exclusively associated with temporal lobe epilepsy and can be evoked by stimulation of insula, hippocampus, amygdala or hypothalamus. Given the prominence of fear and grimaces, a central role of amygdala in the clinical presentation of patient's seizure could be suspected.

Source: EMBASE

Full Text:
Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

7. Autoantibodies to neuronal surface antigens in thyroid antibody-positive and -negative limbic encephalitis.

Author(s): Tuzun E, Erdag E, Durmus H, Brenner T, Turkoglu R, Kurtuncu M, Lang B, Akman-Demir G, Eraksoy M, Vincent A

Citation: Neurology India, January 2011, vol./is. 59/1(47-50), 0028-3886;0028-3886 (2011 Jan-Feb)

Publication Date: January 2011

Abstract: BACKGROUND: Thyroid antibodies (Thy-Abs) are frequently detected in various autoimmune disorders in coexistence with other systemic autoantibodies. In association with an encephalopathy, they are often taken as evidence of Hashimoto’s encephalitis (HE). However, the presence of Thy-Abs in a cohort of limbic encephalitis (LE) patients and their association with anti-neuronal autoimmunity has not been explored. PATIENTS AND METHODS: We investigated thyroid and anti-neuronal antibodies in the sera of 24 LE patients without identified tumors by cell-based assay and radioimmunoassay and evaluated their clinical features. RESULTS: There was a female predominance in Thy-Ab-positive LE patients. Five of the eight Thy-Ab-positive patients and six of the 16 Thy-Ab-negative patients had antibodies to voltage-gated potassium channel (VGKC), N-methyl-D-aspartate receptor (NMDAR) or undefined surface antigens on cultured hippocampal neurons. There were trends towards fewer VGKC antibodies (1/8 vs. 5/16, P = 0.159) and more NMDAR antibodies (2/8 vs. 1/16, P = 0.095) among the Thy-Ab-positive LE patients; antibodies to undefined surface antigens were only identified in Thy-Ab-positive patients (2/8 vs. 0/16, P = 0.018). There were no distinguishing clinical features between Thy-Ab-positive patients with and without neuronal antibodies. However, patients with anti-neuronal antibodies showed a better treatment response. CONCLUSION: Thy-Ab can be found in a high proportion of patients with non-paraneoplastic LE, often in association with antibodies to specific or as yet undefined neuronal surface antigens. These results suggest that acute idiopathic encephalitis patients with Thy-Ab should be closely monitored for ion-channel antibodies and it should not be assumed that they have HE.

Source: MEDLINE

Full Text:
Available in fulltext at EBSCOhost
Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.


Author(s): Pellkofer HL, Kuempfel T, Jacobson L, Vincent A, Derfuss T

Citation: Journal of Neurology, Neurosurgery & Psychiatry, December 2010, vol./is. 81/12(1407-8), 0022-3050;1468-330X (2010 Dec)

Publication Date: December 2010

Source: MEDLINE

Full Text:
Available in fulltext at Highwire Press
Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

Abstract: Purpose: Chronic limbic encephalitis, usually associated with cancer as a paraneoplastic syndrome, may rarely occur without evidence of underlying malignancy. We present MR imaging and clinical workup in 11 patients with nonparaneoplastic, nonherpetic limbic encephalitis. Methods and Materials: Between 1999 and 2009, 11 patients (7 women and 4 men, mean age 41.1 yrs; range 10-63 yrs) presented with symptoms of hippocampal dysfunction (memory deficits, changing mental status, temporal lobe epilepsy). Extensive clinical, laboratory and whole-body imaging work-up did not reveal underlying malignancy. Results: Increased titers of antibodies to antineuronal antibodies (anti-hu) were detected in 5 of 11 patients (45%), in the other patients these titers were not available. MR imaging (39 investigations, range 2-6/patient), performed on a 1.5 and 3 T superconducting system showed bilateral T1-weighted hypointense and T2-weighted and on the FLAIR sequence hyperintense signals in the hippocampi and amygdalae of all patients. Changes were symmetrical in two and asymmetrical in nine cases. Diffusionweighted source images (performed in 19 examinations) revealed mild hyperintense areas in the gyrus cinguli in two patients. Postcontrast images showed no significant enhancement of the corresponding areas. Singlevoxel spectroscopy (TE=136ms) was performed in 3 patients and demonstrated elevated Choline with respect to Creatine and NAA, reduced NAA/Creatine peak and no lactate peak in all patients. Long-term follow-up MRI in seven patients demonstrated complete resolution of the previous hippocampal T2 signal in three patients followed by hippocampal atrophy, partial resolution of abnormalities in three patient and progression in one patient with lethal course. (Figure presented) Conclusion: In patients with clinical symptoms of hippocampal dysfunction without signs of acute illness and MR pattern of limbic involvement, chronic limbic encephalitis must be considered radiologically, even in absence of a malignant disease. The potential clinical improvement with new immunotherapy concepts in these patients emphasizes the importance of a correct
diagnosis.

Source: EMBASE

Full Text:
Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

11. Guest lecture

Author(s): Vincent A.

Citation: Journal of Neurology, Neurosurgery and Psychiatry, October 2010, vol./is. 81/10(e10-e11), 0022-3050 (October 2010)

Publication Date: October 2010

Abstract: It is well recognised that diseases of the peripheral nervous system can be caused by autoantibodies and respond well to immunotherapies associated with a fall in antibody levels. The best examples are, of course, myasthenia gravis and the Lambert Eaton myasthenic syndrome with antibodies to the acetylcholine receptor and voltage gated calcium channels respectively. In addition, Guillain Barre syndrome and Miller Fisher syndrome are associated with antibodies to gangliosides such as GM1 or GQ1b, although these conditions are not necessarily exclusively antibody-mediated. Over the last decade it has become clear that there are some CNS diseases strongly associated with specific antibodies to ion channels or receptors. Antibodies to GluR3 were first reported in Rasmussen's encephalitis, a rare but devastating form of childhood epilepsy for which the treatment is often hemispherectomy. In fact, there are now some doubts about how frequently these antibodies are found in Rasmussen's (Watson et al Neurology 2005) and they are measured routinely in very few centres. The general conception is that Rasmussen's is a T cell mediated inflammatory disorder and that the antibodies, if present, may not be the primary pathological entity. Antibodies to voltage-gated potassium channels, of the shaker-type that binds the snake toxin dendrotoxin, were first identified in patients with an acquired peripheral nerve hyperexcitability syndrome called Isaac's syndrome or neuromyotonia. This condition causes muscle twitching, fasciculations and cramps and is very uncomfortable but not life-threatening. It responds well to anti-epileptic drugs such as phenytoin, and immunotherapies are seldom required. Sometimes, however, neuromyotonia is associated with autonomic dysfunction, sleep disorders, and cognitive impairment. This triad is usually referred to as Morvan's syndrome (Liguori et al Brain 2001). Although rare, it is highly interesting since the patients can present with such a range of symptoms, and some of their abnormalities reflect psychiatric disorders. A recent case reported by Spinazzi et al (Neurology 2008) illustrates this point. A 64 year old patient exhibited prominent compulsive behaviour with increased catecholamine and serotonin secretion as well as epileptic seizures and circadian rhythm suppression. Brain F-FDG-PET demonstrated markedly increased activity in the basal ganglia. Although the presence of multiple neurological signs suggested an organic disease, the history was complex and the diagnosis not clear for some time. Subsequently it was found that VGKC antibodies were clearly raised at 2000 pM, and indeed most of the symptoms and signs reversed following immunosuppressive treatment with a marked fall in VGKC antibodies. Basal ganglia hypermetabolism had not previously been reported with VGKC antibodies but is found in compulsive and psychotic disorders. Thus some of the features of Morvan's syndrome can mimic psychiatric disease. Much more common is VGKC-antibody associated limbic encephalitis (VGKC-LE) which is now a well recognised, most often non-paraneoplastic, form of LE. The VGKC antibodies are usually >400 pM often >3000 pM and fall dramatically following successful immunotherapies. These may include iv steroids, plasma exchange or intravenous immunoglobulins, and long-term high dose oral steroids that can be tapered to nil following clinical improvement. Most patients present with amnesia and seizures with personality change, but psychiatric presentations are not uncommon (Vincent et al 2004; Harrower et al 2006). High signal on MRI in the mesial temporal lobes, often restricted to the hippocampus, is found in the majority of patients. The cerebrospinal fluid is often unremarkable without oligoclonal bands or increased lymphocytes although protein may be slightly raised. Although the majority of patients do well following treatment, some relapse and anecdotal reports suggest that this may be due to lack of compliance or intolerance to steroids. On the other hand in some patients steroids clearly increases psychotic features. No clear guidelines to alternative therapies
exist at this time. A new development is the finding that many of the antibodies to VGKCs are actually directed at other proteins that are complexed with the VGKCs in vivo. These new findings and their clinical associations will be described. Antibodies to glutamic acid decarboxylase are typically found at very high titre in stiff person syndrome. The antibodies themselves may not be pathogenic as GAD is an intracellular enzyme rather than a membrane protein. There may be other antibodies to neuronal cell surface membranes in these patients that are the pathogenic entity. Nevertheless, GAD antibodies are proving to be an important marker of immunemediated neurological disease and found in an increasing number of patients with cerebellar ataxia, epilepsy and other neurological syndromes. Although not yet reported specifically in psychotic syndromes, it is well known that stiff person syndrome may be complicated by psychiatric features and it would not be surprising if some patients with psychosis turned out to have this antibody. Moreover, GAD antibodies can be associated with a form of epilepsy and limbic encephalitis, mainly in young females (Malter et al 2009). A newly described antibody to glycine receptors (GlyR) has been reported in one patient with an exaggerated startle response progressing to encephalomyelitis with rigidity and myoclonus (Hutchinson et Neurology 2008). Although only reported in a single case so far, this antibody has recently been found in other patients, mainly with a form of stiff person syndrome but some of whom exhibit psychiatric features (Leite, Vincent unpublished). One patient was diagnosed as psychogenic until the antibody was detected and has since been treated successfully with immunosuppression (unpublished results). Finally, the most exciting development is a form of autoimmune encephalitis that seems to present frequently with psychiatric features, although it then usually progresses to a full-blown encephalitis with marked movement disorders, mutism, catatonia, seizures and hypothalamic disturbance. NMDAR antibodies are associated with both paraneoplastic (ovarian tumours in young women) and non-paraneoplastic (both sexes but mainly younger patients so far) conditions. Both do well after treatments (removal of the tumour if relevant) although the long-term prognosis may not be so good in the non-paraneoplastic form (Dalmau et al Ann Neurol 2007; Lancet Neurology 2008). New data from our own cases will be presented. VGKC, GAD and NMDAR antibodies are now being searched in patients with various form of neuro-psychiatric disorders and in a study of first episode psychosis (with Drs Brenda Lennox and Michael Zandi in Cambridge). This and further studies will determine how frequently these antibodies can be responsible for psychiatric disorders with obvious treatment and prognostic implications.

**Source:** EMBASE

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12. **Anti-SOX1 antibodies in patients with paraneoplastic and non-paraneoplastic neuropathy**

**Author(s):** Tschernatsch M., Singh P., Gross O., Gerriets T., Kneifel N., Probst C., Malas S., Kaps M., Blaes F.

**Citation:** Journal of Neuroimmunology, September 2010, vol./is. 226/1-2(177-180), 0165-5728 (September 2010)

**Publication Date:** September 2010

**Abstract:** Anti-SOX1 antibodies have been described to be positive in patients with paraneoplastic Lambert-Eaton myasthenic syndrome and, in a lower amount, in patients with anti-Hu positive paraneoplastic neurological syndromes, and with SCLC alone, respectively. We found 5/32 patients with paraneoplastic neuropathy and, surprisingly, 4/22 patients with neuropathy of unknown origin positive for anti-SOX1 antibodies, whereas no patient with inflammatory neuropathy and no healthy controls showed any reactivity (p = 0.007). All patients with neuropathy of unknown origin where followed up for four years without diagnosis of a tumour so far. Anti-SOX1 antibodies are associated with paraneoplastic neuropathies and may define another group of non-paraneoplastic, immune-mediated neuropathies. 2010 Elsevier B.V.

**Source:** EMBASE
13. Case report non-paraneoplastic limbic encephalitis characterized by mesio-temporal seizures and extratemporal lesions: A case report


Citation: Seizure, September 2010, vol./is. 19/7(446-449), 1059-1311 (September 2010)

Publication Date: September 2010

Abstract: Limbic encephalitis (LE) can be either paraneoplastic or a non-paraneoplastic autoimmune disorder. Magnetic resonance imaging (MRI) of the brain on T2-weighted fluid-attenuated inversion recovery (FLAIR) classically shows hyperintensities of the temporal structures, but multifocal involvement of extratemporal cortex has also been described in paraneoplastic LE. Here we describe a 27-year-old woman whose idiopathic autoimmune (glutamic acid decarboxylase-antibody positive) LE debuted with multiple daily mesio-temporal seizures, amnesia and multifocal extratemporal cortical MRI abnormalities. Mesio-temporal MRI signal increase was found after 20 days. This case report highlights that early diagnosis of non-paraneoplastic LE may be considered in patients with multiple daily mesiotemporal seizures and amnesia even in the absence of early typical MRI abnormalities. 2010 British Epilepsy Association. Published by Elsevier Ltd. All rights reserved.

Source: EMBASE

Full Text: Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.


Citation: Seizure, September 2010, vol./is. 19/7(446-9), 1059-1311;1532-2688 (2010 Sep)

Publication Date: September 2010

Abstract: Limbic encephalitis (LE) can be either paraneoplastic or a non-paraneoplastic autoimmune disorder. Magnetic resonance imaging (MRI) of the brain on T2-weighted fluid-attenuated inversion recovery (FLAIR) classically shows hyperintensities of the temporal structures, but multifocal involvement of extratemporal cortex has also been described in paraneoplastic LE. Here we describe a 27-year-old woman whose idiopathic autoimmune (glutamic acid decarboxylase-antibody positive) LE debuted with multiple daily mesio-temporal seizures, amnesia and multifocal extratemporal cortical MRI abnormalities. Mesio-temporal MRI signal increase was found after 20 days. This case report highlights that early diagnosis of non-paraneoplastic LE may be considered in patients with multiple daily mesiotemporal seizures and amnesia even in the absence of early typical MRI abnormalities. 2010 British Epilepsy Association. Published by Elsevier Ltd. All rights reserved.

Source: MEDLINE

Full Text: Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

15. Antibodies to glutamate receptor in limbic encephalitis
**Author(s):** Takahashi Y., Mogami Y., Takayama R., Ikeda H., Imai K.

**Citation:** Brain and Nerve, August 2010, vol./is. 62/8(827-837), 1881-6096 (August 2010)

**Publication Date:** August 2010

**Abstract:** N-methyl-D-aspartate (NMDA)-type glutamate receptor (GluR), is an important molecule, which contributes to the pathophysiological processes of various neurological diseases by various molecular mechanisms. Antibodies against NMDA-type GluR (NR) are detected by immunoblot analysis, enzyme-linked immunosorbent assay (ELISA), or immunocytochemical analysis (Dalmau's method). Immunoblot method uses whole molecules of GluR2 (NR2B), which are synthesized in NIH3T3 cells by using tetracycline system as antigens. In ELISA, synthesized peptides of each domain of GluR2 and GluR1 (NR1) are used. Immunocytochemical method uses human embryonic kidney (HEK) cells transfected by expression vectors for NR1+NR2B/2A. In non-paraneoplastic, non-herpetic acute limbic encephalitis (NHALE), serum antibodies to GluR2 (NR2B) were detected in approximately 60% of the patients from acute to chronic stages further, these antibodies in the cerebrospinal fluid (CSF) were detected in 50% (acute stage), 40% (recovery stage), and 30% (chronic stage) of the patients. The antibodies against GluR2 seemed to increase in the sera after infection and infiltrate the central nervous system through the blood-brain barrier (BBB), which was damaged by cytokines, etc. The antibodies against GluR2 recognize broad regions of GluR2 as epitopes. In NHALE patients with ovarian teratoma, antibodies to NR recognize epitopes on GluR2 (NR2B) and GluR1 (NR1). Pathophysiology of antibodies against NR is estimated to cause internalization of NR on surface of neurons, resulting in inhibition of NR function.

**Source:** EMBASE

**Full Text:**
Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

16. Antibodies to ion channels and receptors; The widening spectrum

**Author(s):** Vincent A.

**Citation:** Journal of Neurology, June 2010, vol./is. 257/(S3-S4), 0340-5354 (June 2010)

**Publication Date:** June 2010

**Abstract:** In myasthenia gravis there are antibodies to the acetylcholine receptor (AChR) in 80% and to muscle specific kinase (Musk) in a proportion of the remaining 20%. The frequency of MuSK antibodies appears to differ with latitude suggesting an environmental influence. The MuSK patients often have severe bulbar symptoms and the reasons for this are not yet clear. In the Lambert Eaton myasthenic syndrome there are antibodies to the voltage-gated calcium channel (VGCC); and in acquired neuromyotonia antibodies to voltage-gated potassium (VGKC) channels. Each of these conditions, which are usually chronic and unremitting, is associated with good clinical responses to immunotherapies (in conjunction with symptomatic therapies), and the roles of the antibodies have been established by a variety of in vitro and in vivo approaches. However, VGKC antibodies are also found in Morvan's which includes peripheral nerve hyperexcitability, autonomic dysfunction and central involvement. This condition is often associated with thymoma. VGKC antibodies are also increasingly found in patients with a predominantly non-paraneoplastic limbic encephalitis and in patients with adult-onset epileptic disorders. Most of these patients do well with immunotherapies. A more complex encephalopathy is associated with antibodies to NMDA receptors. These patients are often younger and can be children. Females seem to be more common than males, and may have ovarian teratomas. The disease involves seizures, cognitive and neuropsychiatric disorders, movement disorders, autonomic disturbance, reduced consciousness and other brainstem features. The patients respond slowly to immunotherapies and may relapse if not adequately treated. Finally, high levels of antibodies to glutamic acid decarboxylase (GAD) appear to be markers not only for the stiff person syndrome (frequently) but also for other immune-mediated CNS diseases such as limbic encephalitis, and antibodies to glycine receptors are beginning to be found in patients with progressive rigidity and startle syndromes.
17. Nonparaneoplastic limbic encephalitis and familial myotonia: A common physiopathogenetic background?

**Author(s):** Licchetta L., Bisulli F., Naldi I., Pittau F., Mostacci B., Broli M., Di Vito L., Stipa C., Tinuper P.

**Citation:** Epilepsia, June 2010, vol./is. 51/(139), 0013-9580 (June 2010)

**Publication Date:** June 2010

**Abstract:** Purpose: To describe a 25-year-old woman with autosomal dominant myotonia, nonparaneoplastic limbic encephalitis (LE) and celiac disease (CD). Method: The proband was referred to us for daily temporal pharmacoresistant seizures and drastic weight loss at age 15, followed 5 years later by a severe memory impairment. Later the proband’s father and brother were referred to us for a myotonic disorder since adolescence. Genealogic tree reconstruction did not disclose other family members reporting symptoms suggestive of myotonia. The proband and the other affected relatives underwent a full clinical, neurophysiological and immunological evaluation. Blood samples were collected for direct sequencing of CLCN1 gene. Results: The proband underwent repeated neuropsychological evaluations, disclosing a severe memory impairment in immediate and delayed recall. Interictal EEG showed frequent epileptiform abnormalities over temporal regions. Brain MRI disclosed bilateral hippocampal hyperintensity in T2-weighted sequences. Laboratory screening revealed autoimmune hypothyroidism, CD (confirmed by biopsy), oligoclonal bands and anti-GAD antibodies in CSF. No seizures were reported by the other affected relatives; their laboratory tests, including autoimmune screening, were negative, whereas neurological evaluation disclosed muscle hypertrophy and myotonia. The proband presented only a subclinical myotonia (elicited by the tongue percussion). Electromyography showed myotonic discharges without myopathic signs in all affected members. Genetic analysis disclosed a missense mutation (I290M) in heterozygosis, confirming the clinical hypothesis of Thomsen disease. Conclusion: An ion channelopathy leading to central and muscle hyperexcitability as the pathogenesis of the association between autoimmune LE and autosomal dominant myotonia is intriguing, but a casual association cannot be excluded.

Source: EMBASE

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Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

18. Autoimmune epilepsies [French] Epilepsies d'origine auto-immune

**Author(s):** Seeck M., Zacharia A., Rossetti A.O.

**Citation:** Revue Medicale Suisse, May 2010, vol./is. 6/247(925-929), 1660-9379 (05 May 2010)

**Publication Date:** May 2010

**Abstract:** There is increasing recognition of an autoimmune origin of pharmacoresistant epileptic disorders. Besides the paraneoplastic limbic encephalopathies (LE), reports of syndromes of non-paraneoplastic LE are increasingly reported in the last 5-10 years. Three antibodies are now relatively well described: Voltagegated potassium channels (VGKC), Glutamic acid decarboxylase (GAD) and N-methyl-D-aspartate receptor-(NMDA) antibodies. We review clinical syndromes, associated imaging and laboratory findings. While most reports arise from adult populations, children and adolescents are also concerned as
19. Anti-VGKC antibody-associated limbic encephalitis/morvan syndrome

**Author(s):** Misawa T., Mizusawa H.

**Citation:** Brain and Nerve, April 2010, vol./is. 62/4(339-345), 1881-6096 (April 2010)

**Publication Date:** April 2010

**Abstract:** Anti-voltage-gated potassium channel antibodies (anti-VGKC-Ab) cause hyperexcitability of the peripheral nerve and central nervous system. Peripheral nerve hyperexcitability is the chief manifestation of Issacs syndrome and cramp-fasciculation syndrome. Morvan syndrome is characterized by neuromyotonia with autonomic and CNS involvement. Manifestations involving the CNS without peripheral involvement are characteristic of limbic encephalitis and epilepsy. The clinical features of anti-VGKC-Ab-associated limbic encephalitis are subacute onset of episodic memory impairment, disorientation and agitation. Hyponatremia is also noted in most patients. Cortico-steroid therapy, plasma exchange and intravenous immunoglobulin are effective in treating not only the clinical symptoms but also hyponatremia. Unlike other anti-VGKC-Ab-associated neurological disorders, paraneoplastic cases are rare. Thus, anti-VGKC-Ab-associated limbic encephalopathy is considered to be an autoimmune, non-paraneoplastic, potentially treatable encephalitis. Morvan syndrome is characterized by widespread neurological symptoms involving the peripheral nervous system (neuromyotonia), autonomic system (hyperhidrosis, severe constipation, urinary incontinence, and cardiac arrhythmia) and the CNS (severe insomnia, hallucinations, impairment of short-term memory and epilepsy). Many patients have an underlying tumor, for example thymoma, lung cancer, testicular cancer and lymphoma; this indicates the paraneoplastic nature of the disease. Needle electro-myography reveals myokimic discharge. In nerve conduction study, stimulus-induced repetitive discharges are frequently demonstrated in involved muscles. Plasma exchange is an effective treatment approach, and tumor resection also improves symptoms. Both VGKC-Ab-associated limbic encephalitis and Morvan syndrome can be successfully treated. Therefore, when these diseases are suspected, it's important to measure the anti-VGKC-Ab level.

**Source:** EMBASE

**Full Text:**

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20. Improvement of non-paraneoplastic voltage-gated potassium channel antibody-associated limbic encephalitis without immunosuppressive therapy.

**Author(s):** Gast H, Schindler K, Z'graggen WJ, Hess CW

**Citation:** Epilepsy & Behavior, April 2010, vol./is. 17/4(555-7), 1525-5050;1525-5069 (2010 Apr)

**Publication Date:** April 2010

**Abstract:** We describe a 61-year-old patient with clinical evidence of limbic encephalitis who improved with anticonvulsant treatment only, that is, without the use of immunosuppressive agents. Three years following occurrence of anosmia, increasing memory deficits, and emotional disturbances, he presented with new-onset temporal lobe epilepsy, with antibodies binding to neuronal voltage-gated potassium channels and bitemporal hypometabolism on FDG-PET scan; the MRI scan was normal. This is most likely a case of spontaneous remission, illustrating that immunosuppressive therapy might be suspended in milder courses of limbic encephalitis. It remains open whether treatment...
with anticonvulsant drugs played an additional beneficiary role through the direct suppression of seizures or, additionally, through indirect immunomodulatory side effects.

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21. Voltage-gated potassium channel antibody-related limbic encephalitis in an HIV-positive female with dialysis-dependent renal failure

Author(s): Apea V., Nortley R., Kirwan C., Sen A., Aboud M.
Citation: HIV Medicine, 2010, vol./is. 11/(64), 1464-2662 (2010)
Publication Date: 2010

Abstract: Background: Limbic encephalitis (LE) is an increasingly recognised autoimmune, paraneoplastic encephalitis. In recent years, a nonparaneoplastic form associated with voltage-gated potassium channel (VGKC) antibodies has been identified. VGKCs are transmembrane channels crucial in nerve action potentials. VGKC antibody-associated LE (VGKC-LE) is a potentially treatable encephalitis. We present the first reported case of VGKC-LE in a HIV positive patient. Case: A 34 year old Ghanaian lady with a five year history of HIV 1 infection and dialysis-dependent renal failure presented to A+E with a two day history of a left-sided headache and increasing confusion, characterised by altered behaviour, incoherent speech, disorientation and marked agitation. She was on abacavir, lamivudine and boosted saquinavir (viral load <40 copies/ml and CD4 count 257). Her last dialysis was two days earlier. Physical examination was unremarkable. Management: CT brain revealed chronic small vessel disease (basal ganglia). MRI brain (incomplete) revealed old infarcts. An electroencephalogram showed mild cortical dysfunction. Cerebrospinal fluid (CSF) analysis revealed a protein of 0.6g/l with a white cell count of <1x10^6/l. The patient was commenced on IV aciclovir. Over the following week, she became less agitated, but cognition was still poor. Her mental test score (MTS) was 2/10, with a receptive dysphasia. CSF viral PCR, STS, TB PCR and JC virus were negative. A repeat MRI reported sub-cortical white matter change in the right temporal lobe ?LE. After three weeks of aciclovir, repeat CSF was unchanged and she was slow to improve with fluctuating cognition and aggressiveness. At this point, results confirmed the presence of serum VGKC antibodies at a level of 479pmol/l (normal level <100 pmol/l). A five day course of plasma exchange (PEx) was commenced. After the first there was a significant improvement in cognition, memory and affect. Her MTS increased to 6/10. PEx was followed by a tapering course of oral prednisolone. A full body CT and PET scan both suggested no underlying malignancy. Her improvement was sustained and her personality returned to normal. Conclusion: This B cell-mediated dysregulation of VGKC function adds to the spectrum of autoimmune phenomena associated with HIV. Encephalopathy is a common diagnostic problem facing HIV physicians. This case highlights a potentially reversible and under-recognised cause which should be actively considered by clinicians.

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22. NMDA receptor antibody encephalitis: A two-stage predominantly non-paraneoplastic disorder in males and females

Author(s): Irani S.R., Bera K., Waters P., Lang B., Bien C.G., Vincent A.
Citation: Annals of Neurology, 2010, vol./is. 68/(53), 0364-5134 (2010)
Publication Date: 2010
Abstract: Antibodies to the NMDA subtype of glutamate receptors (NMDAR-Abs) have been associated with a newly-described encephalopathy (Dalmau et al 2008). Using a modified cell-based assay (CBA) for detection of NMDAR-Abs, and sending out questionnaires to the clinicians, we obtained clinical data on 44 NMDAR-antibody positive patients. NMDAR-Abs were mainly IgG1 and were able to deposit complement on cultured hippocampal neurons. Only 20% overall (9/44) had a detected tumour (follow-up 3.6-121 months, median 16 months), including eight young adult females and one older male, whereas the more frequent non-paraneoplastic cases had a wide range of ages and included ten children. In these non-paraneoplastic cases, good clinical outcomes, broadly correlating with antibody levels measured with a quantitative fluorescent NMDA receptor antibody assay (FIPA), were associated with early (<40 days) administration of steroids plus other immunotherapies (p<0.0001). Although a few patients had epilepsy or limbic encephalitis only, temporal analysis of the clinical and paraclinical features suggested that the disease usually progresses through two main stages with likely cortical (neuropsychiatric, seizures) and subcortical (movement disorders, dysautonomia, reduction in consciousness) origins respectively.

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23. Status epilepticus due to paraneoplastic and nonparaneoplastic encephalitides.

Author(s): Dalmau J

Citation: Epilepsia, December 2009, vol./is. 50 Suppl 12/(58-60), 0013-9580;1528-1167 (2009 Dec)

Publication Date: December 2009

Source: MEDLINE

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24. A treatable dementia

Author(s): Albertyn C.H., Austin N., Connolly S., McCabe D.J.H., Murphy R.P.

Citation: Journal of Neurology, Neurosurgery and Psychiatry, November 2009, vol./is. 80/11, 0022-3050 (November 2009)

Publication Date: November 2009

Abstract: Background Limbic encephalitis is an inflammatory process involving the temporal lobes presenting with behavioural change, short-term memory deficits, seizures and sleep disturbance. Cases of reversible limbic encephalitis associated with neuronal surface antibodies have been described. Voltage-gated-potassium-channel-Ab-associated encephalopathy is now recognised as an autoimmune, largely non-paraneoplastic, potentially treatable encephalitis. The impact of seizures in this disorder has not been widely described. Case Description A 67-year-old woman presented with generalised seizures, rapid onset dementia, sleep disturbance and a movement disorder. Repetitive, stereotypical, unilateral arm posturing associated with hemifacial spasm is described. A global cognitive deficit in temporal, frontal and parietal function is found. A dramatic response to immunomodulatory therapy is observed. Results of Investigations Hyponatremia was found. CSF was negative for Creutzfeld-Jacob disease. There was high signal intensity in both temporal lobes on MRI brain. EEG captured unilateral limb posturing without a definite ictal focus. Furthermore, an EEG video will be shown demonstrating limb posturing followed by secondary generalisation. Voltage-gated potassium channel antibodies were positive at 708 pM. A search for underlying malignancy was negative. Discussion This case highlights frequent atypical partial seizures in the presentation of
voltage-gated potassium channel antibody limbic encephalitis. This is a novel case where these movements were captured on EEG videotelemetry.

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25. Successive affection of bilateral temporomesial structures in a case of non-paraneoplastic limbic encephalitis demonstrated by serial MRI and FDG-PET.
Author(s): Chatzikonstantinou A, Szabo K, Ottomeyer C, Kern R, Hennerici MG
Citation: Journal of Neurology, October 2009, vol./is. 256/10(1753-5), 0340-5354;1432-1459 (2009 Oct)
Publication Date: October 2009
Source: MEDLINE
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Author(s): Verhelst H
Citation: Developmental Medicine & Child Neurology, July 2009, vol./is. 51/7(499-500), 0012-1622;1469-8749 (2009 Jul)
Publication Date: July 2009
Source: MEDLINE
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27. Antibodies against ion channels as potential pathogenic targets in immunemediated epilepsies
Author(s): Kirschstein T.
Citation: Epilepsia, June 2009, vol./is. 50/(12-13), 0013-9580 (June 2009)
Publication Date: June 2009
Abstract: In the 1960s, a paraneoplastic syndrome with temporal lobe seizures, memory loss, and affective symptoms was described and referred to as limbic encephalitis. Nowadays, immune-mediated brain disorders are increasingly recognised as common causes of temporal lobe epilepsy. In the past decade, also patients with non-paraneoplastic limbic encephalitis have been discovered. A number of these patients harbour autoantibodies against voltage-gated potassium channels (VGKC). Dysfunction of the target proteins, the potassium channel subunits Kv1.1, Kv1.2, and Kv1.6, seems to underlie the pathogenesis of this disorder. Assuming that potassium channel function would be impaired by the binding to the autoanti-body this might lead to reduced hyperpolarisation and thus facilitate epileptogenic discharges. Recently, another syndrome was described in a series of young women with ovarian teratomata presenting a clinical picture of epileptic seizures, memory loss, psychiatric symptoms, and autonomous instability. In a majority of these patients, autoantibodies against N-methyl D-aspartate
receptors (NMDA) have been detected. The epitope of these autoantibodies was localised to the extracellular loop of the NMDA receptor subunit NR1. Whereas NR1-containing NMDA receptors are ubiquitously expressed in the brain, the patients’ autoantibodies particularly label NMDA receptors in the hippocampus and in the fore-brain, rather than in the cerebellum. This intriguing region-specificity is believed to be due to the posttranslational modifications such as glycosylations in the antigen epitope that do not occur in many brain regions such as the cerebellum. Thus, together with the findings in limbic encephalitis associated with VGKC autoantibodies, antibodies against neuronal ion Table 1. Patients’ characteristics No. Sex PRS Epilepsy MRI Semiology OP OP Type Histology AED Engel (yrs) (yrs) Localization Side Aura Seizure 2degreeGTC (yrs) (n) 1 f 52 23 temp.-occ. left x simple-partial x 43 Biopsy Angiopathy (Vasculitis ?) 6 n.a. 2 m 25 25 temp.-mes. right x complex-partial x 45 TLR Angiopathy (Vasculitis ?) 9 i B 3 m 5 24 temp.-occ. left x complex-partial x 36 TLR, Lx Vasculitis 4 i B 4 m 29 27 front-temp. left simple-partial x 33 TLR, FLR Encephalitis 4 III A 5 f 10 10 hemiatrophy right x tonic-clonic x n.d. n.a. 9 n.a. 6 m 14 14 frontal left x complex-partial 17, 20 FLR Encephalitis 6 IV B, I A 7 m 10 22 temp.-occ. right x complex-partial x 23 Lx Vasculitis 3 III A channels are not solely diagnostic tools, but also seem to be pathogenically relevant in immune-mediated epilepsies.

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28. Histopathology of chronic encephalitis

Author(s): Bauer J., Bien C.G.

Citation: Epilepsia, June 2009, vol./is. 50/(11), 0013-9580 (June 2009)

Publication Date: June 2009

Abstract: Various inflammatory CNS diseases are accompanied by recurrent epileptic seizures. Well known encephalitic conditions in this regard are Rasmussen encephalitis (RE), paraneoplastic limbic encephalitis (PLE), non-paraneoplastic limbic encephalitis (NPLE), anti-N-methyl D-aspartate receptor (NMDAR) encephalitis, and herpes simplex virus encephalitis. We analysed the pathology and immune response in RE, PLE (with anti-Hu, and anti-Ma2 antibodies), NPLE (with antibodies against voltage gated potassium channels, or glutamic acid decarboxylase 65kDa, GAD65) and NMDAR encephalitis. All these encephalitides cases have in common an acute or chronic loss of neurons and the presence of astro-gliosis and microglial nodules. The inflammatory lesions comprise of lymphocytes and low numbers of macrophages. In RE and PLE more than 70% of the T lymphocytes are CD8 positive. In NPLE and NMDAR encephalitis, only 48% of the T cells were of the CD8 subtype. Another difference was the presence of cytotoxic granules (Granzyme-B, GrB) positive lymphocytes. Whereas in RE and PLE these cells reached numbers as high as 14 (PLE) to 20% (RE), in NPLE only 5% of all T cells were GrB positive. Moreover, in RE and PLE these cytotoxic T cells were clustered around neurons and releasing GrB, suggesting active killing of neurons. In addition to the role of cytotoxic T lymphocytes we also analysed the presence of immunoglobulin (Ig) and complement in brain lesions. Although the presence of anti-neuronal antibodies are the hallmarks of PLE and NPLE we found very little specific deposition of Ig and C9neo in the brains of RE, PLE and NPLE patients, suggesting that antibody-mediated destruction of neurons, at least at the time points analysed, does not play a significant role in the pathology of these diseases.

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29. Potassium channel antibodies in one Patient with reversible limbic encephalitis

Author(s): Dequatre N.

Citation: Journal of Neurology, June 2009, vol./is. 256/(S188), 0340-5354 (June 2009)

Publication Date: June 2009

Abstract: Objectives: Limbic encephalitis is more often considered as a paraneoplastic syndrome. However cases of autoimmune limbic encephalitis are reported in the literature. We report a case of limbic encephalitis with antibodies directed against voltage-gated potassium channels (VGKC antibodies). Patient and method: A 68 year old woman with multinodular goitre and type II diabetes mellitus was admitted for acute confusional state in a context of hyponatraemia and dysthyroidia. Correction of these metabolic disorders did not permit to reduce the symptoms. Additionally, the patient developed severe anterograde amnesia, non fluent dysphasia, insomnia and partial motor seizures. Results: Resonance magnetic imaging of the brain revealed hyperintense signals on T2 weighted images confined to the left hippocampus which were in favour of a limbic encephalitis. Investigations to find a paraneoplastic origin or a cancer were negative. Differential diagnosis like Herpes meningoencephalitis, Creutzfeld-Jakob disease and Hashimoto encephalitis has been excluded. Thus a VGKC antibodies dosage was performed and was positive at a level of 1250 pm. Intravenous corticosteroids 500 mg per day were given during 3 days, then oral prednisone 1 mg/kg per day. The patient also received two polyvalent immunoglobulins treatments (0,4 g/kg/day during 5 days). This treatment improved significantly amnestic, phasic impairments and behavioural symptoms. Conclusion: Clinical characteristics of limbic encephalitis are the same in paraneoplastic or non paraneoplastic causes but prognosis is very different. Indeed, antineuronal antibodies are directed against intracellular antigens and cannot be reached by immunotherapy, whereas VGKC antibodies are directed against membrane antigens and can be reached by treatments. Immunotherapy is efficient in case of non paraneoplastic limbic encephalitis and these forms have usually a good prognosis. VGKC antibodies dosage has to be made in all limbic encephalitis.

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30. Stiff person syndrome in association with nonparaneoplastic limbic encephalitis

Author(s): Franke K., Schulz R., Hoppe M., Korn-Merker E., Specht U., Ebner A.

Citation: Epilepsia, April 2009, vol./is. 50/(157), 0013-9580 (April 2009)

Publication Date: April 2009

Abstract: Purpose: Stiff person syndrome (SPS) is a rare neurological condition characterized by muscle stiffness, rigidity and painful spasms that occurs as a paraneoplastic, autoimmune or idiopathic variant. The autoimmune variant is known to occur in conjunction with other autoimmune disorders especially diabetes mellitus type I and autoimmune thyroiditis. Method: We describe the cases of two young women (21 and 25 years old) with refractory epilepsy due to limbic encephalitis (LE). Both patients initially presented with cognitive/psychiatric symptoms, memory loss and refractory temporal lobe seizures. MR imaging showed typical abnormalities in the medial temporal lobes for LE. Within the course of a few years (3-5 years) both of them developed additional motor symptoms with stiffness, increased muscle tonus and gait disturbance in the sense of a SPS, markedly alleviated by benzodiazepines. Results: In both patients autoimmune status showed increased levels of antibodies against glutamic acid decarboxylase (GAD) but no classical onconeural antibodies. Screening for tumors did not reveal underlying malignancies. Interestingly both patients suffer from other autoimmune diseases: one of them developed diabetes mellitus type I, the other Hashimoto thyroiditis. Conclusion: A SPS can occur in conjunction with LE. These cases support the assumption of common
autoimmune pathogenetic mechanisms and a relevance of GAD antibodies. Autoimmune LE patients should be screened for possibly associated other autoimmune disturbances e.g. diabetes, thyroid disease and stiff person syndrome. Clinicians should be aware of accompanying immune-mediated diseases. To our knowledge this is the first report on SPS in conjunction with nonparaneoplastic LE.

Source: EMBASE

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31. Immunological markers
Author(s): Vincent D.
Citation: Epilepsia, April 2009, vol./is. 50/(11), 0013-9580 (April 2009)
Publication Date: April 2009
Abstract: Immune mediated encephalopathies are beginning to be more widely recognized in adults, and to a certain extent in children. In adults, the best identified syndromes are the paraneoplastic conditions, associated with small cell lung cancers, ovarian, breast or other tumors. Most of these syndromes occur in middle-age and older individuals, as would be expected. They can involve the limbic system, brainstem, cerebellum or multiple levels of the CNS. Among younger patients, Ma2 antibodies are found in a form of limbic and hypothalamic encephalopathy with testicular tumors. And very recently, antibodies to NMDA receptors have been identified in young women with psychiatric or limbic presentations, dyskinesias, stupor often leading to coma. These patients usually have an ovarian teratoma. Both these conditions improve after removal of the tumor and immunotherapies to reduce the antibodies. A nonparaneoplastic form of limbic encephalitis is associated with antibodies to voltage-gated potassium channels. These antibodies are also present in some patients with idiopathic epilepsy, with or without cognitive or other limbic defects. They tend to do very well with immunotherapies. They are almost always adults, and children with similar presentations have only rarely been found positive unfortunately. Therefore, there are few antibodies identified so far in the majority of children with acute or subacute onset of encephalopathy. A few cases of NMDA receptor-antibody associated encephalopathies occur in young adults or children, and GluR3 antibodies may be present in some children with Rasmussen's encephalitis, although this is disputed. But the techniques available for measuring antibodies to different targets and identifying new antibodies are improving rapidly. There is an urgent need for the systematic study of childhood forms of encephalopathies.

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32. Non-paraneoplastic limbic encephalitis associated with antibodies to potassium channels leading to bilateral hippocampal sclerosis in a pre-pubertal girl.
Author(s): Kroll-Seger J, Bien CG, Huppertz HJ
Citation: Epileptic Disorders, March 2009, vol./is. 11/1(54-9), 1294-9361;1294-9361 (2009 Mar)
Publication Date: March 2009
Abstract: Limbic encephalitis (LE) is increasingly recognized as a precipitating factor of adult onset temporal lobe epilepsy frequently associated with bilateral hippocampal damage. So far, clinical data in children are rare and only comprise paraneoplastic forms of LE. We describe a 13-year-old pre-pubertal girl in whom non-paraneoplastic LE was
diagnosed according to diagnostic criteria proposed by Bien and Elger (2007). The girl presented with a subacute syndrome comprising memory impairment, affective disturbances, and refractory temporal lobe seizures. Serial MRI scans demonstrated an initial temporo-median swelling with T2/FLAIR signal increase progressing to bilateral hippocampal atrophy within seven months. Two years after onset of symptoms, antibodies to potassium channels were found to be slightly elevated. Immunosuppressive therapy with steroid-pulses was followed by a transient reduction of seizure frequency, even though this was started more than two years after onset of first symptoms. However, extended immunotherapy was refused by the patient's parents, so no full assessment of the treatment response was possible. In conclusion, this case shows that non-paraneoplastic LE leading to mesial temporal lobe epilepsy is not restricted to adult patients. The proposed diagnostic criteria therefore should be adapted for paediatric patients. Patients may profit from immunosuppressive therapy even when it is started at a late stage with already overt hippocampal sclerosis.

Source: MEDLINE

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33. Autoimmune limbic encephalitis

Author(s): Vedeler C.A., Storstein A.

Citation: Acta neurologica Scandinavica. Supplementum, 2009, vol./is. /189(63-67), 1600-5449 (2009)

Publication Date: 2009

Abstract: Autoimmune limbic encephalitis (LE) can arise both by paraneoplastic and non-paraneoplastic mechanisms. Patients with LE usually have a subacute onset of memory impairment, disorientation and agitation, but can also develop seizures, hallucinations and sleep disturbance. The following investigations may aid the diagnosis: analysis of cerebrospinal fluid (CSF), electroencephalography, magnetic resonance imaging, fluorodeoxyglucose positron emission tomography and neuronal antibodies in the serum and CSF. Neuronal antibodies are sometimes, but not always, pathogenic. Autoimmune LE may respond to corticosteroids, intravenous IgG (IVIG) or plasma exchange. The cornerstone of paraneoplastic LE therapy is resection of the tumour and/or oncological treatment. Several differential diagnoses must be excluded, among them herpes simplex encephalitis.

Source: EMBASE

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34. Immunotherapy responsive startle with antibodies to voltage gated potassium channels.


Citation: BMJ Case Reports, 2009, vol./is. 2009/, 1757-790X (2009)

Publication Date: 2009

Abstract: Antibodies to potassium channels (VGKC-Ab) were first associated with acquired neuromyotonia and its variant with CNS involvement, Morvan's syndrome. Recently, VGKC-Ab were found in patients with non-paraneoplastic limbic encephalitis (LE), characterised by personality changes, seizures and memory impairment. These patients may respond to immunotherapies. Thus the association of VGKC-Ab and non-paraneoplastic LE established the concept of a potentially reversible autoimmune encephalopathy. We describe a patient with startle syndrome and VGKC-Ab, without neuromyotonia or LE, who responded dramatically to plasma exchange (PE) and
immunosuppression, adding to the spectrum of disorders associated with VGKC-Ab.

Source: MEDLINE

35. **Paraneoplastic limbic encephalitis associated with chronic lymphocyte leukaemia**

**Author(s):** Jura R., Stourac P., Bednarova J., Mitasova A.

**Citation:** European Journal of Neurology, 2009, vol./is. 16/S3(583), 1351-5101 (2009)

**Publication Date:** 2009

**Abstract:** Background and aims: Limbic encephalitis (LE) is mostly of infectious or autoimmune origin. Autoimmune LE has subgroups: paraneoplastic (PLE) and non-paraneoplastic (NPLE). PLE harbours paraneoplastic antibodies associated with cancer of lung, thymus, breast, testis and teratoma. NPLE has antibodies against voltage-gated potassium channels (VGKC). LE presents with depression, seizures, hallucinations and short-term memory loss to frank dementia. A case of PLE and chronic lymphocyte leukaemia (CLL) is reported for the first time. Methods: Immunoblot, radioimmunoassay, MR, flowcytometry. Case report: Male, 64 years with medical history of chronic bronchopulmonal disease was admitted to hospital because of progressive mental decline predominantly of short-term memory. Mild cerebellar and brainstem signs were present. MRI scan revealed hyperintesities in amygdallohippocampal areas in consent with limbic encephalitis. Cerebrospinal fluid examination discovered pleocytosis and proteinorhachia without oligoclonal IgG bands and infectious antibodies. Immunofluorescence and immunoblot for paraneoplastic antibodies (anti-Hu, anti-Yo, anti-Ri, anti-CV2, anti-Ma, anti-amphiphysin) and RIA for VGKC antibodies were negative. Concomitantly, the patient was diagnosed for chronic lymphatic leukaemia by flowcytometry with mediastinal lymphadenopathy. Immunosuppressive therapy with prednisolone, mabthera and dexamethasone stabilized the patient oncologically, however, with persisting neurological deficit. The association of PLE with CLL was not yet described. We suppose paraneoplastic etiology in spite of negative known paraneoplastic antibodies and by excluding other current known aetiologies of LE. Conclusion: We report a case of PLE associated with CLL without presence of known paraneoplastic antibodies for the first time. Further search for potential new antibodies in expanding LE entity is warranted.

Source: EMBASE

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36. **Plasma exchange for anti GAD associated non paraneoplastic limbic encephalitis.**

**Author(s):** Mazzi G, Roia DD, Crucciati B, Mata S, Catapano R

**Citation:** Transfusion & Apheresis Science, December 2008, vol./is. 39/3(229-33), 1473-0502;1473-0502 (2008 Dec)

**Publication Date:** December 2008

**Abstract:** Limbic encephalitis (LE) is a neurological syndrome usually presenting in a paraneoplastic form. Recently many cases were reported with no concurring neoplasia, presenting with specific antibodies for voltage-gated potassium channel or for neuronal membrane antigens. Anti-glutamic acid decarboxylase (GAD) antibodies act against GABAergic receptors of the central nervous system. These antibodies were found in coeliac disease serum and in neurologic patients. We are reporting a case of a 21-year-old coeliac woman manifesting complex multiple-daily partial drug-resistant seizures for 7 years. The diagnosis was of a non paraneoplastic limbic encephalitis, unresponsive to high dose cortisone and IGIV infusion. The use of therapeutic plasma exchange (TPE) has resulted in an improvement in symptoms with quite a long disease-free period of time. When the frequency of the procedures was decreased, seizures appeared again and, after suspension of TPE, the clinical status worsened. The role of TPE in the treatment of LE still
37. Acute limbic encephalitis and NMDA type-glutamate receptor

**Author(s):** Takahashi Y., Yamazaki E., Nishimura S., Tsunogae H., Niwa K., Dalmau J., Imai K., Fujiwara T.

**Citation:** Clinical Neurology, November 2008, vol./is. 48/11(926-929), 0009-918X (November 2008)

**Publication Date:** November 2008

**Abstract:** We compared clinical characteristics and autoantibodies against GluR2 between 95 patients with non-paraneoplastic non-herpetic acute limbic encephalitis (NPNHALE) and 19 patients with non-herpetic acute encephalitis accompanying ovarian teratoma (NHALE-OT). Onset age (mean +/- SD) was 27.7 +/- 18.6 years old in NPNHALE, 27.5 +/- 6.5 in NHALE-OT. Preceding factors were found in 63.8% of patients with NPNHALE and 89.5% of patients with NHALE-OT (Fisher's exact test, p = 0.025), and major preceding factors were upper respiratory infections or fever in both groups. Symptoms at the onset were disorder of behavior and talk > seizures > impairment of consciousness in NPNHALE, and disorder of behavior and talk > seizures > disorientation in NHALE-OT. Symptoms at the acute stage were similar between NPNHALE and NHALE-OT, but duration of hospital stay was longer in NHALE-OT (209.0 days) than NPNHALE (87.5 days) (Mann Whitney test, p < 0.0001). At the onset, cell counts in CSF were 51.6 +/- 66.4/mm³ and protein levels were 35.4 +/- 14.7 mg/dl, and IgG levels were 6.5 +/- 4.2 mg/dl in NHALE-OT, and these data were not significantly different between NPNHALE and NHALE-OT. In acute stage, autoantibodies against whole molecule of GluR2 in CSF were detected in 51.8% (29/56) of adult NPNHALE and NHALE-OT, and 40% (6/15) of NHALE-OT patients by immunoblot. These autoantibodies in both groups included epitopes to n-terminal of GluR2. Antibodies against NMDAR complex (Dalmau's method) in CSF were detected in 90.9% (10/11) of NHALE-OT patients.

**Source:** EMBASE

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38. Non-paraneoplastic limbic encephalitis associated with anti-glutamic acid decarboxylase antibodies.

**Author(s):** Mata S, Muscas GC, Naldi I, Rosati E, Paladini S, Cruciatii B, Bisulli F, Paganini M, Mazz G, Sorbi S, Tinuper P

**Citation:** Journal of Neuroimmunology, August 2008, vol./is. 199/1-2(155-9), 0165-5728;0165-5728 (2008 Aug 13)

**Publication Date:** August 2008

**Abstract:** Limbic encephalitis (LE) is a neurological syndrome that may present in association with cancer, infection, or as an isolate clinical condition often accompanying autoimmune disorders. Here we have characterized the clinical and laboratory features of two patients presenting with subacute onset, and chronic evolution, of anterograde amnesia and drug-resistant epilepsy associated with thyroid autoimmunity and in absence of tumoral pathology despite long follow-up. Antibodies against onconeural antigens, voltage gated potassium channel and glutamate receptors, which may accompany paraneoplastic as well as non-paraneoplastic LE, were negative. However, biochemical studies showed high titeres, and sustained intrathecal synthesis, of antibodies directed against neuronal glutamic acid decarboxylase (GAD). In one patient, plasma exchange determined a dramatic improvement of the neurological deficits along with the decrease of
autoantibodies.

Source: MEDLINE

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39. Synaptophysin is an autoantigen in paraneoplastic neuropathy

Author(s): Tschernatsch M., Klotz M., Probst C., Hosch J., Valtorta F., Diener M., Gerriets T., Kaps M., Schafer K.H., Blaes F.

Citation: Journal of Neuroimmunology, June 2008, vol./is. 197/1(81-86), 0165-5728 (15 Jun 2008)

Publication Date: June 2008

Abstract: Paraneoplastic neurological syndromes (PNS) are often associated with antineuronal autoantibodies and many of them could be identified in the recent years. However, there are still new antineuronal binding patterns with yet unidentified autoantigens. We here describe a new autoantibody associated with paraneoplastic sensorimotor and autonomic neuropathy in a patient with small cell lung cancer. In indirect immunofluorescence test, the patient's serum colocalised with the synaptic protein synaptophysin in the cerebellum and myenteric plexus of the gut. Immunoblotting showed a 38 kDa reactivity, which is also the molecular weight of synaptophysin. Therefore a Western Blot with recombinant synaptophysin has been used and revealed reactivity of the serum against synaptophysin. In patients with non-paraneoplastic neuropathies or healthy controls, anti-synaptophysin autoantibodies were not detectable. In 20 SCLC patients without neurological syndromes, two patients had low-titer anti-synaptophysin autoantibodies. The patient's serum and IgG fraction showed cytotoxicity to primary cultured myenteric plexus neurons. We conclude that synaptophysin is an autoantigen in paraneoplastic neurological syndromes. 2008 Elsevier B.V. All rights reserved.

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40. Non-paraneoplastic limbic encephalitis revealed by anterograde amnesia

[French] Une encephalite limbique non paraneoplasique revelee par une amnesie anterograde

Author(s): Lang P.O., Sellal F.

Citation: Presse Medicale, May 2008, vol./is. 37/5 PART 1(775-782), 0755-4982 (May 2008)

Publication Date: May 2008

Abstract: Limbic encephalitis is a syndrome, most commonly paraneoplastic, related to an often undiagnosed cancer of unpredictable prognosis. Neurological symptoms are progressive for a few weeks before stabilizing. Case: We report a case of limbic encephalitis in a 56-year-old man, revealed by anterograde amnesia. The diagnosis was suggested after magnetic resonance imaging (MRI) showed bilateral hippocampal lesions, with signals that were hypointense in IR sequences and hyperintense in FLAIR. The non-neoplastic causation was suggested by an array of clinical, laboratory, imaging, and therapeutic arguments. After five years of follow-up, no neoplasia has been found. Discussion: This case provides the opportunity for a comparison of the radiologic, imaging, and neurologic findings related to bilateral lesions of the hippocampus. 2008 Elsevier Masson SAS. All rights reserved.

Source: EMBASE

Full Text:
41. Excellent outcome after prolonged status epilepticus due to non-paraneoplastic limbic encephalitis.

Author(s): Kondziella D, Andersen O, Asztely F, Holmberg B, Hedstrom A, Szentgyorgyi E

Citation: Acta Neurologica Belgica, March 2008, vol./is. 108/1(21-3), 0300-9009;0300-9009 (2008 Mar)

Publication Date: March 2008

Abstract: Limbic encephalitis (LE) is frequently associated with malignancy. Non-paraneoplastic LE is less common and in this form, voltage-gated potassium channel (VGKC) antibodies are usually found. However in 2007 the spectrum was further extended by a report on four patients with presumed non-paraneoplastic LE in whom neither VGKC-antibodies nor other antibodies could be found (Samarasekera et al. 2007). Despite immunomodulatory treatment all these patients had severe neurological residual symptoms. Here we describe a further patient in whom extensive diagnostic procedures suggested non-paraneoplastic antibody-negative limbic encephalitis. Although this woman had prolonged status epilepticus during seven weeks, her outcome was excellent.

Source: MEDLINE

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Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

42. Clinical application of functional MRI for memory using emotional enhancement: deficit and recovery with limbic encephalitis.

Author(s): Baxter L, Spencer B, Kerrigan JF

Citation: Epilepsy & Behavior, November 2007, vol./is. 11/3(454-9), 1525-5050;1525-5050 (2007 Nov)

Publication Date: November 2007

Abstract: Although some functional MRI memory studies show reliable neural activity in the hippocampus and mesial temporal lobe (MTL), most typically report results from group studies. However, fMRI memory probes need to be robust enough to show MTL activity in individual patients to be helpful in diagnosis and treatment planning. We present the case of a patient with non-paraneoplastic limbic encephalitis who had severe anterograde amnesia with subsequent recovery to illustrate a fMRI probe of MTL activity that is easily administered to neurological patients. The task uses emotionally positive and affiliative stimuli to elicit responsivity in the amygdala-hippocampus region. In this patient, weak bilateral hippocampal activation was observed in the acute stage that increased after recovery, paralleling findings on structural MRI and neuropsychological memory assessment. This case study demonstrates that using emotional stimuli to enhance memory responsivity may be an effective way to visualize clinical changes in individual patients.

Source: MEDLINE

Full Text:
Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

43. Limbic encephalitis: a cause of temporal lobe epilepsy with onset in adult life.

Author(s): Bien CG, Elger CE

Citation: Epilepsy & Behavior, June 2007, vol./is. 10/4(529-38), 1525-5050;1525-5050 (2007 Jun)
Publication Date: June 2007

Abstract: Limbic encephalitis (LE) was described in the 1960s as a clinical-pathological syndrome in adults. Initially, the paraneoplastic form was the center of interest. An increasing number of diagnostically valuable autoantibodies in patients' sera (and cerebrospinal fluid) have been identified. Lately, the impact of non-paraneoplastic LE cases has been acknowledged. In the serum of some of these patients, antibodies against voltage-dependent potassium channels (VGKC antibodies) have been detected. The characteristic MRI course of LE patients has recently been described in detail: hippocampal swelling and T2/FLAIR signal increase are early findings. After a few months, the swelling regresses, followed by hippocampal atrophy with continuous signal increase. A general consensus on formal diagnostic criteria for all LE subsyndromes has not yet been reached. This article proposes such diagnostic criteria and formulates suggestions for treatment.

Source: MEDLINE

Full Text: Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

44. Course and outcome of acute limbic encephalitis with negative voltage-gated potassium channel antibodies.

Author(s): Samarasekera SR, Vincent A, Welch JL, Jackson M, Nichols P, Griffiths TD

Citation: Journal of Neurology, Neurosurgery & Psychiatry, April 2007, vol./is. 78/4(391-4), 0022-3050;1468-330X (2007 Apr)

Publication Date: April 2007

Abstract: BACKGROUND: Limbic encephalitis is a potentially treatable immunological condition. The presence of voltage-gated potassium channel antibodies (VGKC-Ab) in the cerebrospinal fluid (CSF) and serum of patients with the condition is a marker of the disease associated with a non-paraneoplastic form and good response to treatment. Recent work has highlighted absent serum VGKC-Ab and distinct immunology in patients with the paraneoplastic form of limbic encephalitis.METHODS: The cases of four patients with the typical clinical presentation, neuropsychological features and brain imaging of acute limbic encephalitis, in the absence of any evidence for associated cancer during a follow-up of at least 18 months are described here.RESULTS: All patients had negative testing for VGKC-Ab measured during their acute presentation. All patients made some recovery, although they were left with marked cognitive deficits and persistent seizures.CONCLUSION: These cases demonstrate that the absence of VGKC-Ab in limbic encephalitis does not necessarily imply a paraneoplastic form. Further work is required to establish the immunological basis for the disorder in these patients, and the optimal treatment regimen.

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45. Erratum: Dementia in a patient with non-paraneoplastic limbic encephalitis associated with relapsing polychondritis (Pathology (2006) 38, (596-599))

Author(s): Yan M., Cooper W., Harper C., Schwartz R.

Citation: Pathology, April 2007, vol./is. 39/2(292), 0031-3025;1465-3931 (2007 Apr)

Publication Date: April 2007

Source: EMBASE
46. **Protein kinase C gamma autoimmunity in paraneoplastic cerebellar degeneration and non-small-cell lung cancer**

**Author(s):** Sabater L., Bataller L., Carpentier A.F., Aguirre-Cruz M.L., Saiz A., Benyahia B., Dalmay J., Graus F.

**Citation:** Journal of neurology, neurosurgery, and psychiatry, December 2006, vol./is. 77/12(1359-1362), 1468-330X (Dec 2006)

**Publication Date:** December 2006

**Abstract:** BACKGROUND: The clinical and immunological profiles of patients with paraneoplastic cerebellar degeneration (PCD) and non-small-cell lung cancer (NSCLC) are not well known. OBJECTIVE: To review the clinical and immunological features of patients with PCD, NSCLC and without well-characterised onconeural antibodies. METHODS: The clinical features of nine patients with the diagnosis of classical PCD and NSCLC, included in our archives, were retrospectively reviewed. The presence of antibodies to cerebellar components was determined by immunohistochemistry and immunoblot of rat cerebellum. A cDNA library of human cerebellum was screened with the positive sera to identify the antigen. RESULTS: Nine patients with PCD and NSCLC were identified. Six patients were men, and the median age at diagnosis of PCD was 63 (range 47-73) years. PCD was completely reversed in two patients, and partially in one, after treatment of the tumour. The serum of one of the patients with PCD showed a unique reactivity with Purkinje cells. The screening of a cerebellar-expression library resulted in the isolation of protein kinase Cgamma (PKCgamma). PKCgamma immunoreactivity was not observed in the serum of 170 patients with non-paraneoplastic neurological syndromes, 27 patients with PCD, no onconeural antibodies and small-cell lung cancer, and 52 patients with NSCLC without paraneoplastic neurological syndromes. The NSCLC from 11 patients without PCD did not express PKCgamma at either the RNA or protein level. However, many cells of the NSCLC of the patient with PKCgamma antibodies expressed PKCgamma. CONCLUSION: PCD occurs in patients with NSCLC without typical onconeural antibodies and is associated with immune reactions against key proteins of the Purkinje cells.

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47. **Dementia in a patient with nonparaneoplastic limbic encephalitis associated with relapsing polychondritis [10]**

**Author(s):** Yan M., Harper C., Schwartz R.

**Citation:** Pathology, December 2006, vol./is. 38/6(596-599), 0031-3025;1465-3931 (December 2006)

**Publication Date:** December 2006

**Source:** EMBASE

**Full Text:**
Available in fulltext at EBSCOhost
Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.
48. Chronic limbic encephalitides [German] Chronische limbische enzephalitiden

Author(s): Bien C.G., Elger C.E.

Citation: Aktuelle Neurologie, December 2006, vol./is. 33/10(553-559), 0302-4350 (December 2006)

Publication Date: December 2006

Abstract: Limbic encephalitis (LE) was described in the seventh decade of the 20th century as a clinical-pathological syndrome in adults. Initially, the paraneoplastic form was in the center of interest. The demonstration of autoantibodies in patients' sera, which react with brain cells (and tumor cells), represented an important diagnostic progress. On the one hand, antibodies against intracellular antigens, on the other hand against cell membrane antigens ("neuropil antibodies") have been found in cases with paraneoplastic LE. Lately, the impact of the non-paraneoplastic cases has been acknowledged. In a part of these patients, the serum antibodies against voltage-dependent potassium channels (VGKC antibodies) have been detected. The characteristic MRI course of LE patients has recently been described in detail: Hippocampal swelling and T2-/FLAIR-signal increase are early findings. After some months, swelling regresses and hippocampal atrophy comes about with continuous signal increase. There has not yet been an agreement on formal diagnostic criteria for all LE-subsyndromes. In this article, such diagnostic criteria are proposed. The therapy with immunosuppressive or immunomodulating substances is usually without effect in paraneoplastic LE cases with antibodies against intracellular antigens. Successes in the sense of clear improvements are achieved in the LE forms associated with VGKC and neuropil antibodies. Georg Thieme Verlag KG Stuttgart.

Source: EMBASE

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49. Voltage-gated potassium channel antibodies associated limbic encephalitis in a patient with invasive thymoma

Author(s): Ohshita T., Kawakami H., Maruyama H., Kohriyama T., Arimura K., Matsumoto M.

Citation: Journal of the Neurological Sciences, December 2006, vol./is. 250/1-2(167-169), 0022-510X (01 Dec 2006)

Publication Date: December 2006

Abstract: Recently, limbic encephalitis (LE) associated with Voltage-gated potassium channel antibody (VGKC-Ab) has been postulated as a new autoimmune disorder. Most previously reported cases of VGKC-Ab-associated LE were non-paraneoplastic, and reports of a paraneoplastic type are rare. Here we describe a 59-year-old woman with paraneoplastic VGKC-Ab-associated LE preceding the recurrence of invasive thymoma. There was a close temporal relationship between the clinical course and the changes of the VGKC-Ab titer. Unlike many of the non-paraneoplastic VGKC-Ab-associated LE cases, our cases showed the more extensive high intensity lesions on MRI and the absence of seizure and hyponatremia. 2006 Elsevier B.V. All rights reserved.

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50. Antibodies to voltage-gated potassium and calcium channels in epilepsy

Author(s): Majoie H.J.M., de Baets M., Renier W., Lang B., Vincent A.

Citation: Epilepsy Research, October 2006, vol./is. 71/2-3(135-141), 0920-1211 (October 2006)

Publication Date: October 2006

Abstract: Objective: To determine the prevalence of antibodies to ion channels in patients with long standing epilepsy. Background: Although the CNS is thought to be protected from
circulating antibodies by the blood brain barrier, glutamate receptor antibodies have been reported in Rasmussen's encephalitis, glutamic acid decarboxylase (GAD) antibodies have been found in a few patients with epilepsy, and antibodies to voltage-gated potassium channels (VGKC) have been found in a non-paraneoplastic form of limbic encephalitis (with amnesia and seizures) that responds to immunosuppressive therapy. Methods: We retrospectively screened sera from female epilepsy patients (n = 106) for autoantibodies to VGKC (Kv 1.1, 1.2 or 1.6), voltage-gated calcium channels (VGCC) (P/Q-type), and GAD. All positive results, based on the values of control data [McKnight, K., Jiang, Y., et al. (2005). Serum antibodies in epilepsy and seizure-associated disorders. Neurology 65, 1730-1735], were retested at lower serum concentrations, and results compared with previously published control data. Demographics, medical history, and epilepsy related information was gathered. Results: The studied group consisted predominantly of patients with long standing drug resistant epilepsy. VGKC antibodies were raised (>100 pM) in six patients. VGCC antibodies (>45 pM) were slightly raised in only one patient. GAD antibodies were <3 U/ml in all patients. The clinical features of the patients with VGKC antibodies differed from previously described patients with limbic encephalitis-like syndrome, and were not different with respect to seizure type, age at first seizure, duration of epilepsy, or use of anti-epileptic drugs from the VGKC antibody negative patients. Conclusion: The results demonstrate that antibodies to VGKC are present in 6% of patients with typical long-standing epilepsy, but whether these antibodies are pathogenic or secondary to the primary disease process needs to be determined. 2006 Elsevier B.V. All rights reserved.

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51. Stiff person syndrome and motor mononeuropathy with conduction block: A singular association

Author(s): La Spada S., Negro C., Nozzioli C., De Marco V., Passarella B.
Citation: Clinica Terapeutica, May 2006, vol./is. 157/3(237-239), 0009-9074 (May/June 2006)
Publication Date: May 2006
Abstract: The "Stiff person syndrome" (SPS) is a rare dysimmune chronic neurological disorder, sometimes paraneoplastic, characterized by progressive stiffness, painful persistent or spasmody muscle contractions, mostly involving spine and lower extremities. In 60 to 90 percent of cases, non-paraneoplastic forms are associated to the presence of anti-glutamic acid decarboxylase (anti-GAD) antibodies in the cerebrospinal fluid and in the serum, while anti-amphiphysin antibodies are frequently associated to paraneoplastic types. The relevant treatment consists of three basic approaches: increase in the inhibitory processes in charge of muscle activity control, re-modulation of the immune response, removal of any associated neoplasia. Indications regarding the efficacy of high-dose intravenous immunoglobulin (IVIG) also in this dysimmune pathology are on the increase. We described an unusual case of autoimmune SPS associated with an exclusively motor left peroneal nerve neuropathy, with conduction block, treated with high-dose intravenous immunoglobulin (IVIG), oral cyclosporine, sodium valproate, baclofen and diazepam.

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52. Rapid eye movement sleep behavior disorder and potassium channel antibody-associated limbic encephalitis.

Citation: Annals of Neurology, January 2006, vol./is. 59/1(178-81), 0364-5134;0364-5134
Of six patients registered in our center with nonparaneoplastic limbic encephalitis associated with antibodies to voltage-gated potassium channels, the five men had rapid eye movement sleep behavior disorder (RBD) coincident with voltage-gated potassium channel antibody-associated limbic encephalitis onset. In three patients, immunosuppression resulted in resolution of RBD in parallel with remission of the limbic syndrome. RBD persisted in two patients with partial resolution of the limbic syndrome. Our findings suggest that RBD is frequent in the setting of voltage-gated potassium channel antibody-associated limbic encephalitis and can be related to autoimmune-mediated mechanisms. In addition, these observations suggest that impairment of the limbic system may play a role in the pathogenesis of RBD.

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53. Neuromuscular junction channelopathies: A brief overview

Author(s): Newsom-Davis J.

Citation: Acta Neurologica Belgica, December 2005, vol./is. 105/4(181-186), 0300-9009 (December 2005)

Publication Date: December 2005

Abstract: The neuromuscular junction lacks the protection of the blood-nerve barrier and is vulnerable to antibody-mediated disorders. In myasthenia gravis (MG), 85% of patients have IgG antibodies to acetylcholine receptors (AChRs). About half the remaining patients have IgG antibodies to Muscle Specific Kinase (MuSK), an AChR-associated transmembrane post-synaptic protein concerned in AChR aggregation. Bulbar weakness is typically predominant in this form of MG, and females are more often affected. The Lambert-Eaton Myasthenic Syndrome (LEMS) can occur in a paraneoplastic form (P-LEMS) usually with small cell lung cancer, or in a non-paraneoplastic form (NP-LEMS). In both, IgG antibodies to nerve terminal voltage-gated calcium channels (VGCCs), detectable in over 90% of patients, lead to VGCC loss and impaired quantal release of transmitter and may be implicated in the occasionally associated cerebellar ataxia. Neuromyotonia (NMT) and Cramp-Fasciculation syndrome (C-FS) are manifestations of peripheral nerve hyperexcitability and share some clinical and electromyographic features. Antibodies to voltage-gated potassium channels (VGKCs) are present in about 40% of NMT patients, but less frequently in C-FS, and appear to cause loss of functional VGKCs. They may also be implicated in the Maladie de Morvan and limbic encephalitis that can associate with NMT. The antibodies described here provide valuable aids to diagnosis and management. The Congenital Myasthenic Syndromes are a group of genetically determined heterogeneous disorders, usually recessively inherited. The commonest mutation sites appear to be the acetylcholine receptor-subunit and rapsyn.

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54. BR serine/threonine kinase 2: A new autoantigen in paraneoplastic limbic encephalitis

Author(s): Sabater L., Gomez-Choco M., Saiz A., Graus F.

Citation: Journal of Neuroimmunology, December 2005, vol./is. 170/1-2(186-190), 0165-5728 (30 Dec 2005)

Publication Date: December 2005
Abstract: We describe a new antigen, BR serine/threonine kinase 2 (BRSK2), identified by an antibody present in the serum of a patient with limbic encephalitis and small-cell lung cancer (SCLC). Patient's serum immunolabeled the neuronal cytoplasm and, less intense, the neuropil of rat brain but did not immunoreact with other rat tissues with the exception of testis. Immunoblots of rat brain homogenate identified several immunoreactive bands in the range of 88-82 kDa and a weaker broad band of 47-43 kDa. Probing a rat hippocampus expression library with the patient's serum resulted in the isolation of BR serine/threonine kinase 2 (BRSK2), a protein (also know as SAD1B kinase) preferentially expressed in the brain and testis and implicated in neuronal polarization as well as synaptic development. Eluted IgG from the BRSK2 clone gave a similar immunolabeling than the patient's serum by immunohistochemistry and immunoblot of rat brain and testis. BRSK2 antibodies reacted with two SCLC from patients without paraneoplastic neurological syndromes. No anti-BRSK2 antibodies were found in the serum of 50 patients with SCLC without PNS, 19 with limbic encephalitis without onconeural antibodies, 50 with anti-Hu antibodies and several paraneoplastic neurological syndromes, including 14 with limbic encephalitis, and 160 with a variety of non-paraneoplastic neurological syndromes. Our study suggests BRSK2 may be an autoantigen involved in the pathogenesis of SCLC-associated limbic encephalitis. 2005 Elsevier B.V. All rights reserved.

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55. Pilomotor seizures and status in non-paraneoplastic limbic encephalitis.


Citation: Epileptic Disorders, September 2005, vol./is. 7/3(205-11), 1294-9361;1294-9361 (2005 Sep)

Publication Date: September 2005

Abstract: BACKGROUND AND AIMS: To describe an unusual clinical presentation of a patient with voltage-gated potassium channel Ab- positive, non-paraneoplastic limbic encephalitis.METHODS: We performed video-EEG monitoring, structural MRI, (18)F-FDG-PET, (1)H-MRS, neuropsychological testing and antibody serology.RESULTS: A 42-year-old male patient presented in an acute phase of non-paraneoplastic limbic encephalitis confirmed by MRI, with antibodies to voltage-gated potassium channels. His pilomotor status was pharmacoresistant to antiepileptic drugs, but responded to corticosteroid and azathioprine treatment. The MRI findings improved. The pilomotor seizures recurred when the immunosuppressive therapy was discontinued after 18 months. MRI at that time was consistent with hippocampal sclerosis. Complete seizure control was achieved after reintroduction of steroids.CONCLUSION: Pilomotor seizures were the predominant seizure type in this case of non-paraneoplastic limbic encephalitis. Immunosuppressive therapy may provide recovery including seizure control. However, long-term immunosuppression may be necessary to prevent relapse. Hippocampal sclerosis and chronic epilepsy might evolve as sequelae of limbic encephalitis.

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56. Temporal lobe seizures, amnesia and autoantibodies - identifying a potentially reversible form of non-paraneoplastic limbic encephalitis.

Author(s): Vincent A, Bien CG

Citation: Epileptic Disorders, September 2005, vol./is. 7/3(177-9), 1294-9361;1294-9361 (2005 Sep)
57. FDG-PET and MRI in potassium channel antibody-associated non-paraneoplastic limbic encephalitis: correlation with clinical course and neuropsychology.

Author(s): Fauser S, Talazko J, Wagner K, Ziyeh S, Jarius S, Vincent A, Schulze-Bonhage A

Citation: Acta Neurologica Scandinavica, May 2005, vol./is. 111/5(338-43), 0001-6314;0001-6314 (2005 May)

Abstract: OBJECTIVES: We report a patient with potassium channel antibody-associated non-paraneoplastic limbic encephalitis (NPLE) in whom repeated fluorodeoxyglucose positron emission tomography (FDG-PET) and magnetic resonance imaging (MRI) are correlated with epileptic activity and memory performance during the course of disease. CASE SUMMARY: A 32-year-old woman suffered from prolonged global amnesia after two generalized tonic-clonic seizures due to NPLE. Initially, MRI showed swelling of the left hippocampus. In FDG-PET, however, bitemporomesial hypermetabolism was seen corresponding to frequent bitemporal independent seizure patterns. Also neuropsychological impairments pointed to a bitemporal involvement at this early stage. In parallel with improved control of electrographic seizure patterns, improvement was seen in FDG-PET and in memory performance. During the whole course, MRI showed only left-sided abnormalities, which correlated with a permanent verbal memory impairment. CONCLUSION: FDG-PET was more sensitive in showing the initial bitemporal involvement and correlated well with EEG findings and neuropsychological impairment in the acute phase of disease. In contrast, structural MRI better reflected persistent neuropsychological deficits.

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58. Nonparaneoplastic, nonherpetic limbic encephalitis with severe episodic hypothermia: a case report.

Author(s): Fukushima K, Yasaki M, Kaneko K, Fushimi T, Yamamoto K, Hashimoto T, Oguchi K, Ikeda S

Citation: European Neurology, 2005, vol./is. 54/3(170-4), 0014-3022;0014-3022 (2005)

Publication Date: 2005

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Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

59. Nonparaneoplastic limbic encephalitis with relapsing polychondritis.

Author(s): Ohta Y, Nagano I, Niiya D, Fujioka H, Kishimoto T, Shoji M, Abe K

Citation: Journal of the Neurological Sciences, May 2004, vol./is. 220/1-2(85-8), 0022-
Relapsing polychondritis (RP), which shows pain, swelling and destruction of the affected parts, is a rare autoimmune disorder affecting cartilage. We report a patient with RP that affected skull cartilage, who subsequently developed multifocal meningoencephalitis. The patient presented with severe recent memory disturbance, anxiety and moderate depression. MRI study showed bilateral median temporal lobe lesions including hippocampi and amygdaloidal bodies, abnormal findings that disappeared after treatment with high-dose steroids. This is thought to be the first case of RP presenting amnesic syndrome and mental disorder associated with nonparaneoplastic limbic encephalitis involving bilateral hippocampi and amygdaloidal bodies detected by MRI.

Source: MEDLINE

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60. Potassium channel antibody-associated encephalopathy: a potentially immunotherapy-responsive form of limbic encephalitis.


Citation: Brain, March 2004, vol./is. 127/Pt 3(701-12), 0006-8950;0006-8950 (2004 Mar)

Abstract: Patients presenting with subacute amnesia are frequently seen in acute neurological practice. Amongst the differential diagnoses, herpes simplex encephalitis, Korsakoff’s syndrome and limbic encephalitis should be considered. Limbic encephalitis is typically a paraneoplastic syndrome with a poor prognosis; thus, identifying those patients with potentially reversible symptoms is important. Voltage-gated potassium channel antibodies (VGKC-Ab) have recently been reported in three cases of reversible limbic encephalitis. Here we review the clinical, immunological and neuropsychological features of 10 patients (nine male, one female; age range 44-79 years), eight of whom were identified in two centres over a period of 15 months. The patients presented with 1-52 week histories of memory loss, confusion and seizures. Low plasma sodium concentrations, initially resistant to treatment, were present in eight out of 10. Brain MRI at onset showed signal change in the medial temporal lobes in eight out of 10 cases. Paraneoplastic antibodies were negative, but VGKC-Ab ranged from 450 to 5128 pM (neurological and healthy controls <100 pM). CSF oligoclonal bands were found in only one, but bands matched with those in the serum were found in six other patients. VGKC-Ab in the CSF, tested in five individuals, varied between <1 and 10% of serum values. Only one patient had neuromyotonia, which was excluded by electromyography in seven of the others. Formal neuropsychology testing showed severe and global impairment of memory, with sparing of general intellect in all but two patients, and of nominal functions in all but one. Variable regimes of steroids, plasma exchange and intravenous immunoglobulin were associated with variable falls in serum VGKC-Abs, to values between 2 and 88% of the initial values, together with marked improvement of neuropsychological functioning in six patients, slight improvement in three and none in one. The improvement in neuropsychological functioning in seven patients correlated broadly with the fall in antibodies. However, varying degrees of cerebral atrophy and residual cognitive impairment were common. Over the same period, only one paraneoplastic case of limbic encephalitis was identified between the two main centres. Thus, VGKC-Ab-associated encephalopathy is a relatively common form of autoimmune, non-paraneoplastic, potentially treatable encephalitis that can be diagnosed by a serological test. Establishing the frequency of this new syndrome, the full range of clinical presentations and means of early recognition, and optimal immunotherapy, should now be the aim.

Source: MEDLINE

Full Text:
61. Anti-Ma2 antibody related paraneoplastic limbic/brain stem encephalitis associated with breast cancer expressing Ma1, Ma2, and Ma3 mRNAs

Author(s): Sahashi K., Sakai K., Mano K., Hirose G.

Citation: Journal of Neurology, Neurosurgery and Psychiatry, September 2003, vol./is. 74/9(1332-1335), 0022-3050 (01 Sep 2003)

Publication Date: September 2003

Abstract: A 69 year old woman presented with cognitive impairment and supranuclear gaze palsy caused by paraneoplastic limbic/brain stem encephalitis associated with atypical medullary breast carcinoma. The cerebrospinal fluid from the patient harboured an anti-neuronal cell antibody against Ma2 antigen, but not against Ma1 or Ma3 antigen. Despite the antibody being restricted to the Ma2 antigen, the patient's cancer tissue expressed Ma1, Ma2, and Ma3 mRNAs. These results, and the expression of Ma2 mRNA in an atypical medullar breast carcinoma in another patient without paraneoplastic encephalitis, indicate that the induction of anti-Ma2 antibody depends on host immunoreponsiveness and not on the presence of the antigen itself in the cancer.

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Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.
Available in print at Pilgrim Hospital Staff Library

62. Successful immune treatment for non-paraneoplastic limbic encephalitis.

Author(s): Mori M, Kuwabara S, Yoshiyama M, Kanesaka T, Ogata T, Hattori T

Citation: Journal of the Neurological Sciences, September 2002, vol./is. 201/1-2(85-88), 0022-510X;0022-510X (2002 Sep 15)

Publication Date: September 2002

Abstract: A 21-year-old woman subacutely developed memory loss subsequent to gastroenteritis. Brain MRI with gadolinium enhancement showed symmetric involvement of the amygdala. The CSF was acellular with increased protein level. There was no evidence suggestive of neoplasm or viral infection. Combined treatment with plasmapheresis and immunoglobulin improved her clinical symptoms and lessened abnormalities manifested in the MRI. This case suggests the presence of immune-mediated limbic encephalitis without association with neoplasms or infections.

Source: MEDLINE

Full Text:
Available in fulltext at ULHT journal article requests. Complete the online form to obtain articles.

63. MRI and diffusion MRI in nonparaneoplastic limbic encephalitis.

Author(s): Sener RN

Citation: Computerized Medical Imaging & Graphics, September 2002, vol./is. 26/5(339-42), 0895-6111;0895-6111 (2002 Sep-Oct)
Abstract: This paper reports an 65-year-old woman with nonparaneoplastic limbic encephalitis. On MRI and diffusion MRI, diffuse and symmetrical, bilateral high-signal lesions were evident in the medial temporal lobes. The affected areas were hyperintense on b= 1000 s/mm(2) (heavily diffusion-weighted or true diffusion) images, a pattern similar to cytotoxic edema. However, apparent diffusion coefficient (ADC) values read directly from corresponding ADC maps at each temporal lobe were within normal limits: 0.96 and 0.97 x 10 x (-3) mm(2)/s, respectively, excluding cytotoxic edema. The changes were shown to be resolved on a 9-month follow-up MRI.

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64. Limbic encephalitis not associated with neoplasm as a cause of temporal lobe epilepsy.

Author(s): Bien CG, Schulze-Bonhage A, Deckert M, Urbach H, Helmstaedter C, Grunwald T, Schaller C, Elger CE

Citation: Neurology, December 2000, vol./is. 55/12(1823-8), 0028-3878;0028-3878 (2000 Dec 26)

Abstract: OBJECTIVE: To describe four patients with temporal lobe epilepsy with limbic encephalitis unrelated to neoplasm.METHODS: The authors performed a retrospective evaluation of patient data obtained during presurgical evaluation, with additional CSF analyses, serum analyses, and histopathologic investigations.RESULTS: The patients shared the following clinical features: onset of the disease in young adulthood with subacute onset or exacerbation of frequent intractable temporal lobe seizures, verbal and visual memory deficits, and affective abnormalities. MRI showed variably extended areas of increased T2 signal in limbic structures and adjacent areas. In the histopathologic investigation, chronic inflammation was observed without evidence of a viral origin. There was no evidence of an underlying malignancy.CONCLUSIONS: Nonparaneoplastic limbic encephalitis should be included in the differential diagnosis of adult patients with temporal lobe epilepsy.

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Author Information.
... Limbic Encephalitis: From Pathologic to Clinical-Immunologic Criteria.

Serial MRI of limbic encephalitis
H Urbach, BM Soeder, M Jeub, T Klockgether... - Neuroradiology, 2006 - Springer
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CI AKMAN, MC PATTERSON... - Medicine & Child ..., 2009 - Wiley Online Library
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