

Anti-Amphiphysin Antibody-Induced Paraneoplastic-Autoimmune Encephalitis: Case Report and Literature Review

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Summary. Anti-amphiphysin antibody-induced encephalitis is an immune-mediated inflammatory disease of the brain caused by onconeural antibodies. Amphiphysin antibodies are associated with various paraneoplastic neurological syndromes and tumours. The absence of a particular clinical pattern and pathognomonic features makes diagnosing paraneoplastic-autoimmune encephalitis complicated, especially when a malignancy is not initially apparent. Detection of specific antibodies in serum and cerebrospinal fluid remains the main diagnostic tool. Up to this date, no cases with epileptic seizures as one of the clinical manifestations of anti-amphiphysin encephalitis have been reported. In this article, we present a rare clinical case of a 54-year-old man diagnosed with autoimmune-paraneoplastic encephalitis caused by antibodies against amphiphysin which manifested with atypical clinical symptoms such as epileptic seizures, visual and perception impairment, paraesthesia, and elevated CSF protein levels with no detectable tumour.

Keywords: amphiphysin, autoimmune encephalitis, epileptic seizures, paraneoplastic syndromes.

INTRODUCTION

Encephalitis is an inflammatory brain disease associated with high morbidity and mortality, the etiology of which often remains unknown [1]. The incidence of various etiology encephalitis ranges from 1.5 to 12.3 cases per 100 000 inhabitants per year [2, 3]. Although encephalitis of infectious origin is the most common in clinical practice, the cause remains unidentified in about 50% to 80% of cases [4]. In recent decades, there has been an increasing focus on immune-mediated encephalitis, and recent studies show that at least 20% of encephalitis cases are caused by various autoantibodies [5]. Encephalitis associated with antibody synthesis can be classified according to whether the antibodies are directed against neuronal surface and synapse proteins, or against intracellular proteins also

called onconeural proteins. Autoimmune encephalitis develops due to antibodies to synaptic proteins or proteins on the neuron surface. The most commonly detected autoimmune antibodies are NMDAR, LGI1, CASPR2, AMPAR, GABA_AR, GABA_BR, VGCC [6]. These disorders are characterized by a wide range of clinical manifestations, and the most common symptoms are memory and cognitive impairment, involuntary movements, and epileptic seizures [7]. The conditions in which onconeural antibodies are detected are called classical paraneoplastic neurological syndromes (PNS). PNS are relatively rare, developing in less than 1% of patients diagnosed with oncological disease. The most frequently diagnosed oncological diseases associated with PNS are small cell lung carcinoma (SCLC), thymoma, and breast cancer [6]. Approximately 60% of patients with PNS have antibodies to cellular antigens in their serum and (or) cerebrospinal fluid (CSF) directed against intranuclear antigens such as -Hu, -Ma, -Yo, or amphiphysin [8]. These autoantibodies are specific for the tumour but not for the neurological symptoms, as the neurological syndrome may be associated with multiple antibodies [9]. PNS can affect any region of the nervous system and can present with multiple

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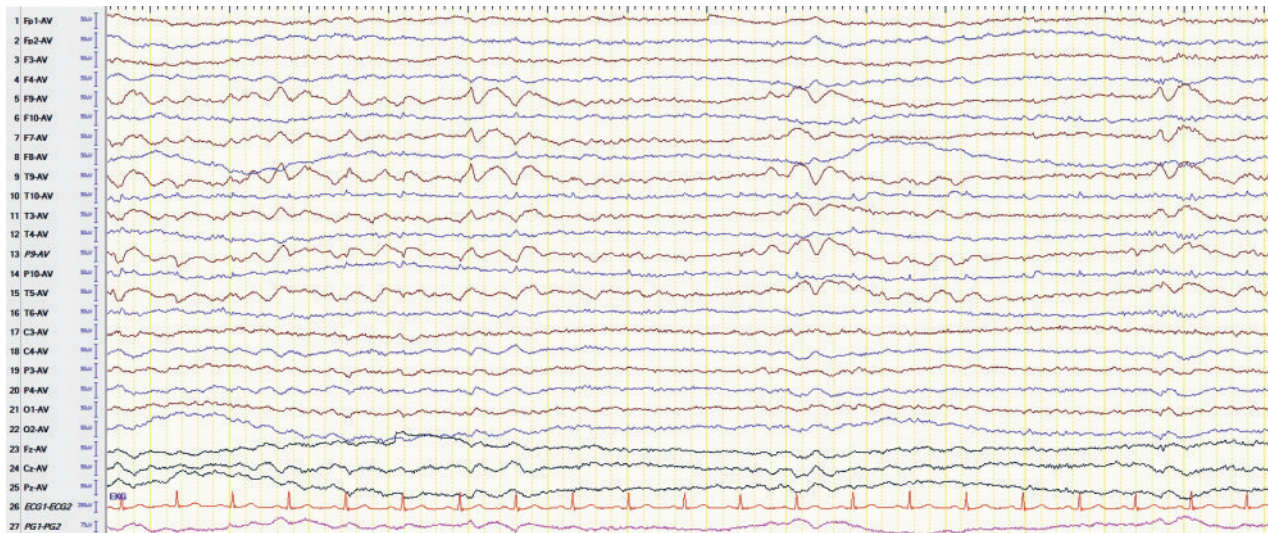


Fig. 1. **Electroencephalography**

Slow pathological waves and epileptiform potentials in the left temporal region.

clinical manifestations such as encephalitis, autonomic failure, peripheral neuropathy, cerebellar ataxia, visual complaints, and many others [10]. Due to the variety of symptoms and lack of pathognomonic features, the diagnosis is often challenging. While autoimmune NMDA encephalitis has diagnostic criteria, the diagnosis of other immune-mediated encephalitis is based on clinical symptoms, cerebrospinal fluid analysis, and specific autoantibody detection [11]. Some studies have tried to provide more accurate diagnostic guidelines for PNS to help clinicians, but to date there are still no internationally accepted diagnostic criteria [12]. In this article, we present a rare clinical case of autoimmune-paraneoplastic encephalitis caused by antibodies against amphiphysin which manifested with non-specific clinical symptoms and no detectable malignancy.

CASE REPORT

A Russian-speaking 54-year-old man presented with recurrent bilateral tonic-clonic seizures (BTCs), visual impairment in the right visual field, impaired perception of the right side of the body, feeling of selflessness in the right limbs, and paraesthesia in various areas. The first episode of BTCs occurred 7 months ago: at the beginning, the patient started to feel “parasitic movements” in his head, then spun around for a few seconds, and lost consciousness with bilateral tonic-clonic seizure. The episode was terminated with diazepam in the emergency department. The patient was diagnosed with epilepsy and started treatment with carbamazepine. The second identical episode occurred two months later. During later epileptic event, the patient remained intact (symptoms not evolved to bilateral tonic clonic), but frequency of episodes gradually increased from 1 time per month to 1 time per week. From anamnesis it was known that about 1.5 years ago the patient was admitted to the hospital with a suspicion of ischemia in the

left *arteria carotis interna* M2 region because of partial sensorimotor aphasia, right hemisyndrome, and right hemianopsia. As there were no indications for reperfusion therapy, symptomatic treatment was applied. Speech and vision problems persisted. The patient was consulted by an epileptologist and treatment was supplemented with valproic acid. Neurological examination showed that the patient had difficulties in formulating complaints, could not find the right word or even understand the questions. To some questions he replied in English. There was right hypoesthesia and visual field defects on the right side, but no pathological changes were found in the confrontational examination. Examination of the cranial nerves showed discordant movements of the eyeballs, restricted to the left and bottom, and a flattened nasolabial fold on the right side. Laboratory tests showed minor lymphocytic leucocytosis and dyslipidaemia, while thyroid, liver and renal function tests were normal. Low-amplitude beta frequency activity with slow pathological waves and epileptiform potentials in the left temporal region were recorded during electroencephalography (EEG) (Fig. 1).

Due to suspected autoimmune encephalitis, a panel of autoimmune encephalitis and onco-neural antibodies in serum was performed and the results showed high titres of anti-amphiphysin. Brain MRI showed encephalomalacia in the left temporoparietal area with a small surrounding gliosis, decreased left hippocampus, thinned grey matter layer, and dilated perivascular gaps in the subcortical and basal nucleus areas with no other infectious or neoplastic lesions (Fig. 2). Analysis of CSF showed increased level of protein. Since this is a paraneoplastic antibody, computer tomography (CT) of chest and ultrasound of abdomen was performed. A solid small peripheral focus of clear boundaries in the right lung S5 segment was found in chest CT (Fig. 3), however, biopsy results showed a reactive lymphadenopathy in the intrapulmonary lymph node. Based on clinical, laboratory, and imaging data, a clinical diagnosis of paraneoplastic-autoimmune encephalitis

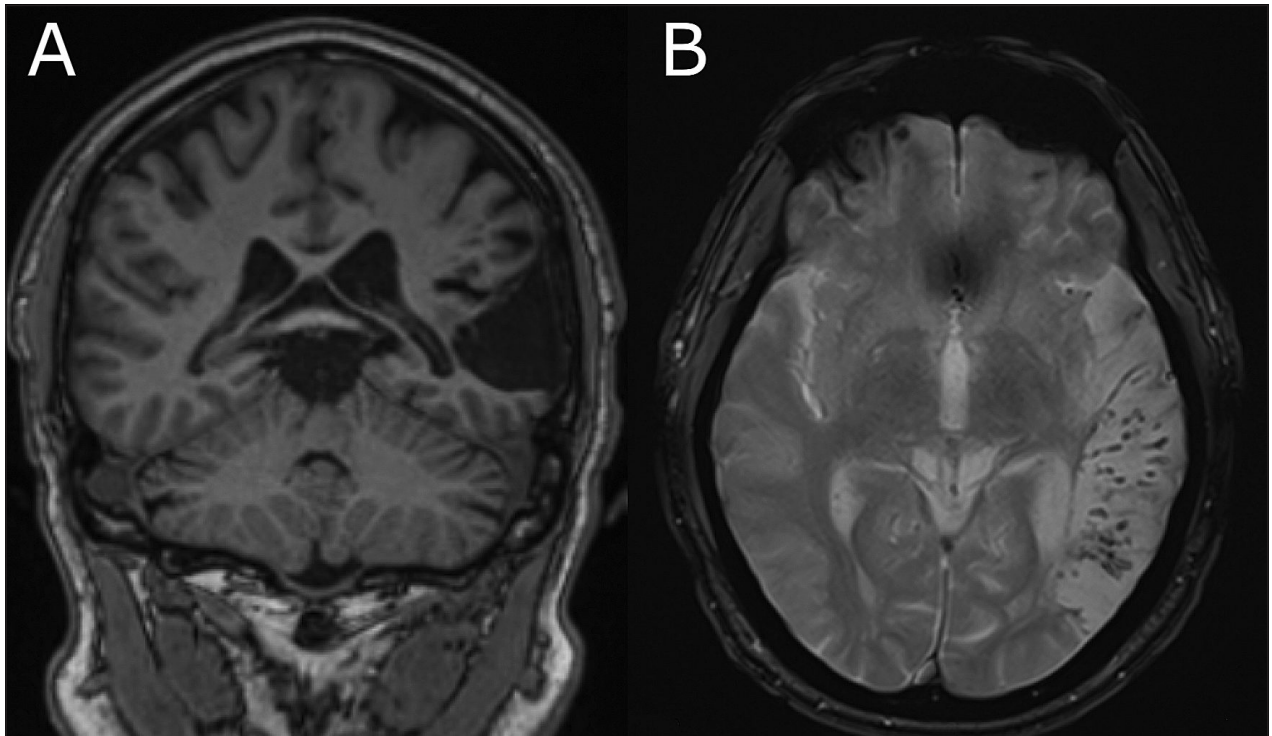


Fig. 2. Brain MRI scan

Hypointense signal in the left temporoparietal area, decreased left hippocampus, thinned grey matter layer, dilated perivascular gaps. A – coronal view, T1 sequence. B – axial view, T2 sequence.

caused by anti-amphiphysin antibodies was made. Immunosuppressive therapy was initiated with methylprednisolone pulse therapy 1 gram IV per day for 5 days followed by oral prednisolone of 60 mg per day for 1 month. Levetiracetam in addition to valproic acid was administered for seizure control. Despite the treatment, the seizures continued, the psychical status worsened, and the patient stopped taking prednisolone. Due to the high probability of oncologic disease, further follow-up with whole-body positron emission tomography (PET) scans every 3–6 months was needed, but the patient refused any further consultations and treatment and has not appeared to date.

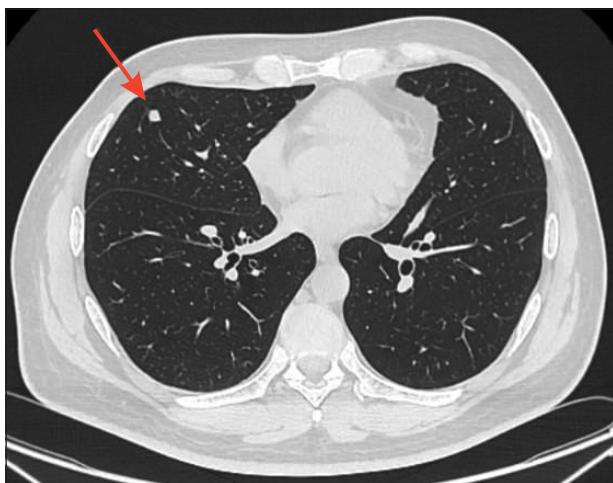


Fig. 3. Chest CT scan

Intrapulmonary lymph node in the right lung S5 segment (arrow).

DISCUSSION AND LITERATURE REVIEW

We presented a case of autoimmune-paraneoplastic encephalitis induced by anti-amphiphysin antibodies, which presented with epileptic seizures, visual and perception impairment. Encephalitis was suspected after EEG test, which showed an abnormal background activity with slow pathological waves and epileptiform potentials in the left temporal region. Brain MRI showed encephalomalacia in the left temporoparietal area with a small surrounding gliosis and atrophy of the left hippocampus. Most patients with encephalitis undergo brain MRI at early stages of the disease. Findings can be normal or non-specific, but sometimes they may suggest an autoimmune cause. Depending on the syndrome, MRI of the brain can reveal T2/FLAIR hyperintensity in limbic structures, cerebellar atrophy, or other changes [13]. However, the MRI findings in this case were incidental, showed no signs of autoimmune brain inflammation, and could not explain the pathological background activity. Epileptic seizures are a common feature of autoimmune encephalitis. Autoimmune NMDA receptor encephalitis often manifests with various types of seizures, most frequently with GTCS, as they develop in about 70% cases. However, other clinical symptoms such as orofacial dyskinesias, dystonia, psychiatric symptoms, and autonomic dysfunction are also present, and IgG NMDA GluN1 antibodies in serum or CSF are usually detected [14]. Another immune-mediated encephalitis, during which epileptic seizures occur, is associated with LGI1 autoantibodies. Antibodies that bind to LGI1 proteins in-

crease neuronal activity leading to electrical discharges in the brain manifested by seizures. This disease also presents with memory impairment, confusion, specific facio-brachial dystonic seizures, detection of LGI1 antibodies, hyponatremia, and typical changes in MRI (medial and temporal hyperintensity [15]). Other autoimmune diseases associated with seizures are anti-AMPA receptor encephalitis, which is more frequently diagnosed for middle-aged women, GABA_AR encephalitis, which is more common among children and in most cases presents with treatment-resistant seizures, *status epilepticus*, and *epilepsia partialis continua*, and GABA_BR encephalitis, which usually occurs later in life [16, 17]. Paraneoplastic encephalitis has a wide range of clinical manifestations including epileptic seizures; however, occurrence of seizures is quite rare while other symptoms are more common. Limbic encephalitis is one of the classical paraneoplastic neurological syndromes, in most cases caused by anti-Hu or CV-2/CRMP-5 antibodies and is often associated with SCLC, however, it presents with not only epileptic seizures but, especially limbic system damage symptoms, with behaviour and mood changes, short-term memory impairment, and additional features that vary according to the immune response [18, 19]. Other PNS usually do not present with seizures as it is not a typical clinical sign. EEG is an essential diagnostic test to determine the type of seizures and the location of pathological discharges in the brain, but alterations in EEG are rarely specific to autoimmune neurological syndromes, except some distinctive forms of encephalitis such as “extreme delta brush” seen in anti-NMDA receptor encephalitis [20]. Although immune-mediated encephalitis was suspected after recording epileptiform activity in EEG, the detection of antibodies directed against amphiphysin in serum led to the diagnosis of autoimmune-paraneoplastic encephalitis. Many patients with paraneoplastic syndromes have antibodies in their serum and cerebrospinal fluid that react with both the nervous system and the underlying cancer. The identification of these antibodies and their target neural antigens has substantially advanced the possibilities of early diagnosis and led to the concept that paraneoplastic neurologic disorders are immune-mediated. Autoantibodies are highly specific for identifying a neurologically disabled patient with paraneoplastic syndrome. These antibodies also indicate the site of the underlying cancer and react with the part of the nervous system that is responsible for clinical symptoms. However, not all patients have well-characterised paraneoplastic antibodies and other clinical symptoms, including patients with atypical syndromes or with undetectable tumours [21]. In more than half of cases of paraneoplastic encephalitis, clinical symptoms appear before oncological disease because the tumour is often too small to be detected, but the immune response is already expressed [10]. Autoimmune-paraneoplastic encephalitis caused by anti-amphiphysin antibodies is a rare disorder with various clinical manifestations, but to date, no cases with epileptic seizures presented. Amphiphysin I is a nerve terminal protein that plays a role in endocytosis. Anti-

amphiphysin antibodies react with a 128-kd protein found in synaptic vesicles and they were first described in patients with paraneoplastic stiff-man syndrome and breast cancer [17]. Some studies suggest that they are associated with various paraneoplastic syndromes (sensory neuronopathy, encephalomyelitis, limbic encephalitis) and tumours (SCLC, ovarian carcinoma, breast cancer, thymoma) [21]. A direct relationship between autoimmune brain tissue inflammation and seizures remains unclear and is difficult to study [22]. While the typical presentation of antibody-associated encephalitis consists of a subacute progressive decrease in the level of consciousness, often with fluctuations, altered cognition, and other neural tissue damage signs, the manifestation of the immune-mediated process in the described case was atypical. Moreover, a previous ischemic stroke in the patient’s medical history should be considered as a possible cause of epileptic seizures, since epilepsy is not a single disorder but rather a group of syndromes with variety of underlying diseases. The role of cerebrovascular disorders is observed in the development of 11% of all epilepsy cases. Arterial ischemic stroke causes many primary and secondary changes that can initiate epileptogenesis, therefore, epilepsy seizures are one of the most common poststroke outcomes [23]. In our case, there is possibility that seizures could be caused by dual pathology related to both ischemic and immune-mediated reasons, however, the signs of acute stroke were not evidently expressed, findings in MRI were incidental, and possible encephalitis of paraneoplastic or autoimmune origin was suspected only after performing EEG. Furthermore, our patient not only presented with epileptic seizures, but also complained of visual impairment which is most common in paraneoplastic brain stem encephalitis. However, this patient had no other brain stem damage symptoms as during this syndrome visual disturbances manifest as supranuclear or intranuclear ophthalmoplegia, opsoclonus, nystagmus, and other symptoms that are associated with brain stem damage (dysarthria, dysphagia, vertigo, central hypoventilation) [24]. As paraneoplastic neurologic disorders can affect any part of the nervous system, the variety of clinical symptoms and signs of the syndromes is very large. The absence of a particular clinical pattern makes diagnosing paraneoplastic encephalitis difficult as the analysis of CSF and detection of specific autoantibodies in serum and (or) CSF remain the main diagnostic tool [11]. Most common findings are moderate lymphocytic pleocytosis, increased protein concentration, high IgG index, and oligoclonal bands [12]. Analysis of our patient’s CSF showed only elevated protein concentration, however, the absence of pleocytosis does not rule out immune-mediated encephalitis [25]. Early diagnosis and treatment of these diseases is important because irreversible damage to the nervous system can occur if they are not treated in time. Since paraneoplastic syndromes are considered to be immune-mediated, two treatment approaches are used: removal of the source of the antigen by treatment of the underlying cancer and suppression of the immune response. For many paraneoplastic

syndromes, the first approach is the only effective treatment [26]. There are no established protocols for the treatment of most paraneoplastic syndromes, but a combination of both plasma exchange or intravenous immune globulin and immunosuppressive agents is used. The effect of treatment is highly dependent on the type of antibodies detected because for most paraneoplastic syndromes, immunotherapy is not effective, especially if encephalitis is caused by anti-Hu or anti-Yo antibodies [27]. Recent studies suggest that the combination of intravenous immunoglobulin or plasma exchange with cyclophosphamide may be effective in some cases [28]. No clinical studies have been performed to evaluate the efficacy of immunotherapy in the treatment of anti-amphiphysin-induced encephalitis. In our case, as tumour was not detected, immunosuppressive treatment was initiated with methylprednisolone pulse therapy and continued with oral prednisolone. Symptomatic treatment included treatment of seizures and psychiatric symptoms, as well as stabilization of vital signs. Most tumours are identified by imaging of the chest, abdomen, or pelvis using CT, fluorodeoxyglucose PET or both, but in many cases, initial search for cancer is unrewarding and the tumour is detected months or even a few years after the appearance of the neurologic syndrome. Whole-body PET may be the best screening method for locating the occult cancer [29]. If primary screening is negative, re-screening after 3–6 months and every 6 months for up till 4 years is required [30].

CONCLUSIONS

Amphiphysin antibodies are associated with various paraneoplastic neurological syndromes and tumours. Autoimmune-paraneoplastic encephalitis caused by anti-amphiphysin antibodies is a rare and usually severe disorder with various clinical manifestations, but no cases with epileptic seizures have been reported to this date. Diagnosing immune-mediated encephalitis is often difficult due to the absence of a particular clinical pattern and abnormalities in imaging or laboratory tests, especially when the malignancy is not initially apparent. In our case, the patient presented with atypical clinical symptoms such as epileptic seizures, visual and perception impairment, elevated protein levels in CSF, non-specific findings in MRI, and positive amphiphysin autoantibodies, which led to a diagnosis of autoimmune-paraneoplastic amphiphysin-associated encephalitis. The main diagnostic tool in this case remained the detection of specific onconeural antibodies in the serum. Amphiphysin autoantibodies in most cases are associated with SCLC or thymoma, so a primary screening for cancer was initiated. As symptoms often appear before the oncological disease is detected, subsequent follow-up repeating whole-body PET was required. Further research on PNS and adoption of international diagnostic recommendations could improve diagnostic accuracy, management and lead to better outcomes.

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**ANTIKŪNŲ PRIEŠ AMFIFIZINĄ SUKELTAS
PARANEOPLASTINIS-AUTOIMUNINIS
ENCEFALITAS: ATVEJO PRISTATYMAS
IR LITERATŪROS APŽVALGA**

Santrauka

Antikūnų prieš amfifizinę sukeltas encefalitas – imuninės kilmės galvos smegenų uždegimas, kurio metu susidarę onkoneuroniniai antikūnai prieš vėžinių ląstelių antigenus kryžmiškai pažeidžia nervinį audinį. Amfifizino antikūnai siejami su įvairiais paraneoplastiniais neurologiniais sindromais ir navikais. Dėl simptomų įvairovės ir tikslų diagnostinių kriterijų trūkumo, šios ligos diagnostika dažnai būna apsunkinta, todėl specifinių antikūnų nustatymas paciento serume ir cerebrospinaliniame skystyje išlieka pagrindine diagnostikos priemone. Iki šiol nėra aprašyta klinikinių atvejų, kur epilepsijos priepuoliai būtų minimi kaip viena iš encefalito, sukulto antikūnų prieš amfifizinę, klinikinių išraiškų. Straipsnyje pateikiame retą klinikinį antikūnų prieš amfifizinę sukulto atipinio paraneoplastinio-autoimuninio encefalito atvejį, kuris pasireiškė epilepsijos priepuoliais, parestezijomis bei regos ir suvokimo sutrikimais.

Raktažodžiai: amfifizinas, autoimuninis encefalitas, paraneoplastinis sindromas, epilepsija, traukuliaai.

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