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Consensus statement on the diagnosis and treatment of autoimmune nodopathies from the Peripheral Neuropathy Association of the Chinese Society of Neurology

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Abstract

Autoimmune nodopathis are a group of immune-mediated peripheral neuropathy, with antibodies to Ranvier nodal regions that includes anti-NF155, contactin1 (CNTN1), and contactin-associatedprotein1 (Caspr1) antibodies. Clinical features, neurophysiology, cerebrospinal fluid test, peripheral nerve imaging, sural neuropathology and antibody detection, diagnostic criteria were summarized in this consensus, as well as suggested principles of treatment with rituximab, glucocorticoids, plasma exchange and other immunotherapy medicines.

Keywords: Autoimmune nodopathies, diagnosis, treatment, consensus

INTRODUCTION

Autoimmune nodopathy (AN) is a class of immune-mediated peripheral neuropathy in which anti-NF155, contactin1 (CNTN1), and contactinassociated protein1 (Caspr1) antibodies are detectable in serum, which are located at paranodal regions. Anti-NF186/NF140 antibodies, -gliomedin -neuronal cell adhesion molecules (NrCAM) antibodies are antibodies against nodal molecules., mainly IgG4 isoforms.[1] The onset of this group of diseases can be acute, subacute or chronic, and the clinical manifestations are motor and sensory peripheral neuropathy. Nerve electrophysiological testing can show demyelinating changes, such as decreased conduction velocity, prolonged distal motor latencies, motor conduction block, abnormal temporal dispersion, and prolongation of F-wave latency. There is no obvious inflammation or macrophage-mediated demyelination on sural nerve biopsy, while detachment between myelin loops and axons can be seen at the paranode, and axonal degeneration occurs in severe cases. AN generally responds poorly to Intravenous Immunoglobin (IVIG) therapy, and most patients respond to glucocorticoids, rituximab, and plasmapheresis. The specific clinical features vary among AN with different antibodies. There are few epidemiological data; it is estimated that, AN accounts for about 5~10% of patients with chronic inflammatory demyelinating polyneuropathy (CIDP).[2-4] In the past 10 years, the understanding of AN has gradually deepened. In order to standardize the diagnosis and treatment of AN, the Peripheral Neuropathy Association of Chinese Society of Neurology organized relevant experts to discuss the disease and formulated this consensus.

Definition of antibodies related to paranodal region and autoimmune nodopathy

Nodal region is the main structure of peripheral nerve myelinated fibers to achieve saltatory conduction, including three parts: nodal area, paranodal area, and juxtaparanodal area.[5,6] The molecular structure is different at different area, antibodies against different molecular structures

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can be produced, leading to peripheral neuropathy. From the current studies, AN refers to a group of diseases, with the clinical manifestations similar to CIDP, and poor response to IVIG. Anti-NF155 antibodies, anti-CNTN1 antibodies, anti-Caspr1 antibody or anti-NF186/140 antibody could be detected in serum, which are mainly IgG4 subtypes. With the deepening of understanding, it is possible that more antibodies may be identified in the future. This consensus may be updated as knowledge and research progresses.

Clinical features of autoimmune nodopathy

The clinical manifestations of AN usually manifests as multiple motor and sensory peripheral neuropathy, and the clinical manifestations vary according to the different pathogenic antibodies.

Anti-neurofascin 155 autoimmune nodopathy[7-13]

Anti-NF155 IgG4 related AN is the most common AN in clinical practice, accounting for an incidence of about 1-10% of CIDP. Young and middle-aged patients are more common, most of them have chronic onset, and a few can be subacute. Limb weakness in the distal muscles is more severe than the proximal muscles, sensory impairment in the distal extremities is very severe, which can lead to ataxia. High-amplitude low-frequency tremor can be seen in the upper limbs, as well as in the head and tongue; 30%~50% of patients can have cranial nerve involvement such as facial muscle weakness, ophthalmoplegia, and papilledema. Cerebral white matter lesions on MRI could be detected in a small number of patients with NF155 antibody.[14]

Anti-contactin 1 autoimmune nodopathy[15-18]

It accounts for 0.7-2.4% of CIDP, and it is more common in middle-aged and older men. Most of the patients have acute or subacute onset, which progresses rapidly, and can be similar to Guillain-Barré syndrome in the early stage. There are also patients with chronic onset. Weakness of the distal limbs and sensory ataxia are common, and postural tremor may be present, but not as common in AN with anti-NF155 antibodies. There are also cases with predominant sensory involvement. Cranial nerve involvement and neuralgia could occur in some patients. More than half of the patients can be accompanied by nephrotic syndrome.

Anti-Caspr1 antibody or Anti-Caspr1 / contactin 1 antibody autoimmune nodopathy[5,10,19]

It accounts for about 0.2~1.9% of CIDP. Acute or subacute onset with rapid progression being more common. About half of patients are misdiagnosed with Guillain-Barre syndrome at an early stage. The clinical manifestations include sensorimotor involvement, sensory ataxia, and tremor. Nearly half of patients have neuralgia, which can be a prominent symptom. Cranial nerves may be affected.

Anti-neurofascin 186/140 autoimmune nodopathy[11,20]

AN associated with anti-NF186/140 antibody is rare. Most of the patients have acute and subacute onset. There is usually symmetrical motor and sensory function involvement. The distal limbs is more severely affected than the proximal limbs. A few patients have asymmetric manifestations. There is usually no tremor or ataxia. There may be associated nephrotic syndrome.

Notes

1. AN can present with acute, subacute, or chronic onset, with a clinical manifestation of motor and sensory peripheral neuropathy. Limb weakness is more serve distally than proximally, often with ataxia.
2. Anti-NF155 antibody-related patients always have obvious tremor, ataxia, and occasional central white matter lesions on MRI.
3. Neuralgia may be more prominent in AN related to anti-Caspr1 antibody, with GBS-like onset, and occasionally sensory involvement is the mainstay.
4. Anti-CNTN1 antibody-related patients often have a similar onset of Guillain-Barre syndrome, and may have sensory involvement, nephrotic syndrome is more common.
5. Tremor and ataxia are rare in NF186 antibody-related patients, and there may be asymmetrical manifestations, and some patients with nephrotic syndrome.

Neurophysiological features of autoimmune nodopathy

Neurophysiological changes can be similar to demyelination lesions. There is no significant difference in electrophysiology between AN with different antibodies.[1-4,20] Motor nerve conduction studies always show significantly prolonged distal

motor latency, decreased conduction velocity, motor conduction block, abnormal temporal dispersion, decreased occurrence rate of F wave and prolonged F wave latency. For the cut-off of those parameters for diagnosing demyelination, can refer to the criteria of CIDP suggested by EAN/PNS[20], although markedly prolonged distal motor latencies and probable conduction block at Erb's point were more frequently present in AN than in CIDP.[21] Compound muscle action potential(CMAP) amplitude are frequently decreased significantly, especially in the lower limbs. In patients with anti-NF140/186 antibody, motor NCS showed conduction block but no temporal dispersion. A small number of patients with AN have been reported to present with the electrophysiological characteristics of axonal peripheral neuropathy, such as those related to NF186 antibodies. Sensory nerve conduction studies always showed decreased conduction velocity and sensory nerve action potential. Concentric needle electromyography showed abnormal spontaneous potentials and reduced recruitment of motor unit potentials on needle electromyography. Although AN presented acute or subacute onset, the needle electromyography always showed large MUP at the first visit because the underlying pathology might have occurred before the onset of symptoms.

Notes

In AN, motor conduction studies showed significantly prolonged distal latencies, decreased conduction velocity, conduction block, abnormal temporal dispersion and other characteristics. motor and sensory nerve conduction amplitude often decreased significantly in the early stage.

Cerebrospinal fluid changes

In AN, cerebrospinal fluid (CSF) showed albuminocytologic dissociation. In anti-NF155 antibody, anti-CNTN1 antibody, and anti-Caspr1 antibody-related AN, cerebrospinal fluid protein are often significantly elevated, with an average of 2~3g/L.[4] CSF protein may be normal or slightly elevated in patients with anti-NF140/NF186 antibodies.[20]

Notes

CSF protein is often markedly elevated in AN, and CSF albuminocytologic dissociation can be used as a basis to support the diagnosis of AN, but it is not specific.

Peripheral neuroimaging

Ultrasound of peripheral nerves shows increased cross-sectional area of the nerves and abnormal nerve fascicular signaling. Nerve thickening is more pronounced at proximal part, the lumbosacral and brachial plexus. Nerve thickening is common in patients with NF155, CNTN1, and Caspr1 antibodies, and is also seen in patients associated with NF186 antibodies, but not as common as the first three.[3,20,22]

Magnetic resonance imaging is mainly used for the detection of morphological changes of the lumbosacral and brachial plexus; the cauda equina nerve root, which showed thickened in most patients and contrast enhancement in some patients.[3,12,23]

Notes

Peripheral nerve thickening on ultrasound or magnetic resonance imaging can be detected in AN, more prominently at lumbosacral and brachial plexus. Nerve thickening on imaging is nonspecific and can be used as a supplement to electrophysiological testing as basis for diagnosis. Imaging is more valuable when the amplitude of sensorimotor nerve conduction is too low to accurately determine conduction velocity.

Sural nerve pathological changes

In IgG4-associated AN, sural nerve pathology shows endometrial edema, axonal degeneration, no demyelination and remyelination, and no inflammatory cell infiltration.[24] Immunohistochemistry shows positive IgG4 staining for antibodies in node (NF140/NF186) or paranodal region (NF155/CNTN1/Caspr1).[25] Electron-microscopy showed that the myelin loop of the paranodal area was separated from the axon.[26]

Notes

Sural nerve biopsy is not necessary to diagnose AN. When the diagnosis of AN is in doubt, pathology can be helpful in confirming the diagnosis.

Antibody test

Positive serum anti-node antibody is the key to the diagnosis of AN, including mainly anti-NF155, anti-CNTN1, anti-Caspr1 and anti-NF140/186 antibodies. Cerebrospinal fluid (CSF) has a low rate of antibody positivity. The IgG3 subtype is

seen in the acute phase in some patients with AN and can be converted to the IgG4 subtype during follow-up. Other IgG subtypes has also been reported at different stages after the onset of AN.[1-4,25] When clinical improvement occurs with AN treatment, anti-node and paranodal antibody titers decrease, or become negative. When performing antibody testing, the CBA method is recommended. It is better that ELISA methods be performed for confirmation after CBA methods, although ELISA method is associated with higher false negatives.

Notes

Positive test for anti-node and paranod antibody are mandatory for the diagnosis of AN. Antibody titers can vary depending on the disease. The antibody test results, especially positive results, should be interpreted in the context of the clinical information to avoid being misled by false positives due to quality control issues.

Diagnosis and differential diagnosis

Diagnosis of AN includes four key aspects:

Clinical manifestations: (1) Acute, subacute or chronic course of the disease, which continues to progress 8 weeks after onset. (2) Clinically consistent with multiple motor sensory peripheral neuropathy. (3) It may be accompanied by ataxia, tremor or neuralgia. (4) It can be accompanied by nephrotic syndrome,

Electrophysiological manifestations: (1) Motor nerve conduction measurement showed prolonged distal latencies, decreased conduction velocity, abnormal temporal dispersion, conduction block, and decreased F-wave conduction velocity, similar to the characteristics of demyelination. (2) Massive axonal degeneration: marked amplitude decrement in motor and sensory responses, abnormal spontaneous potentials and reduced recruitment of motor unit potentials on needle electromyography can be seen.

Antibody test: Serum anti-node/paranode antibodies are positive.

Cerebrospinal fluid: Albuminocytologic dissociation, and cerebrospinal fluid protein is often significantly elevated. AN needs to be distinguished from many other motor and sensory demyelinating polyneuropathy caused by a variety of other causes, such as CIDP, paraproteinemic neuropathies, familial amyloidosis peripheral neuropathy, peripheral neuropathy associated with paraneoplastic syndromes, and others. AN is often misdiagnosed as Guillain-Barré syndrome in the early stage of the disease, and also needs to be distinguished from poisoning (such as n-hexane poisoning), and positive anti-node/paranode antibodies are the key to diagnosis.

Notes

1. When patients present with polyneuropathy that progresses over 8 weeks, with demyelination-mimic changes in the motor NCS, AN should be considered. This is particularly so if accompanied by significant ataxia, tremor, or comorbidity of kidney disease.
2. Positive serum anti-node and paranode antibodies are the main basis for diagnosis of AN.
3. When nerve conduction studies confirmed demyelinating neuropathies, there are a variety of disorders that need to be differentiated from AN.

Treatment

Immunotherapy

Immunotherapies are of great importance to improve the prognosis by controlling the progression of the disease as soon as possible. There is a lack of robust randomized, double-blind, placebo-controlled trials to support the choice of treatment for AN. Clinical recommendations are based on multiple small observational studies. Rituximab, glucocorticoids, and plasma exchange can all be used in the treatment of AN. IVIG therapy is ineffective in the majority of patients with AN.[1-4] In previous studies, AN was diagnosed in patients with refractory CIDP, while with the universal testing of anti-node/paranode antibody, more clinical phenotypes and antibody types may be recognized, and more specific treatment may be initiated.

Rituximab: When AN is diagnosed, there is a preference to initiate rituximab therapy as early as possible to reduce disability. About 80% of patients are effective when treated with rituximab, and about 20% have recurrence. In the use of rituximab, patients typically begins to respond on an average of 3 months (weeks to months) after administration and should be maintained periodically thereafter.[27-30] CD19+ levels should

be monitored regularly during treatment. There is no uniform recommendation for the protocol of rituximab treatment in AN. Rituximab was given once a week at 375 mg/m^2 for four consecutive weeks in some reports, and about 10% of patients will have serious adverse reactions. A low-dose treatment regimen had been suggested, starting with 100mg intravenous infusion once a week for a total of 4 weeks, and then100mg once a month for a total of 2 months. Some centers suggested that rituximab be given 100mg on the first day, 500mg on the second day, and 500mg every 6 months thereafter. Differences in efficacy between different regimens are poorly documented, but the latter two regimens are associated with a low incidence of adverse effects.

Glucocorticoid therapy: Glucocorticoids are commonly used in AN, and nearly half of patients improved.[3,9,13,16,31] The most commonly used oral regimen is prednisone 1mg/kg per day or 60 mg daily for 4 weeks, then tapered 5~10 mg every 2~4 weeks depending on clinical improvement. It usually takes 1~3 months to determine the effect of treatment. If treatment is effective, the dose can be reduced to 20 mg or less per day for maintenance in about 6 months. For patients with severe disease, pulse methylprednisolone can be given, 500~1000 mg intravenous infusion per day, and then changed to oral maintenance after 3~5 days.

Plasmapheresis: About one-third to one-half of patients with AN respond to plasmapheresis. The amount of plasma exchange is 30 to 50 ml/kg each time, 3 to 5 times in 1~2 weeks. Depending on the clinical situation, it can be used regularly at intervals of 1~2 months.[3,9,13,16,31]

IVIG treatment: In NF186 antibody-positive AN, nearly half of patients respond to IVIG.[20] IVIG can be given at 400 mg/kg per day, intravenous infusion in 3 to 5 days. IVIG is not effective in patients with IgG4 subtypes. In other types of AN, IVIG response is less than 20% and often transient, with a decrease in response during reuse.[1-4]

Other immunosuppressants: when the above treatment is not satisfactory or cannot be used, azathioprine, mycophenolate mofetil, cyclosporine, cyclophosphamide and other immune preparations can be tried according to personal experience, and some patients may respond to these immunosuppressants.

Other treatments

Some patients have acute onset, rapidly progressive course of disease, quadriplegia, and even respiratory muscle involvement. It is thus necessary to closely monitor the progress of patients, enhance the nursing, and start ventilator-assisted support if necessary. Patients with neuralgia can be treated with gabapentin, pregabalin, and amitriptyline.

Rehabilitation exercises can help promote the recovery of nerve function. Patients with nephrotic syndrome may also be seen to improve with the improvement of neurological function after immunotherapy. Thus, renal function and urine protein should be monitored in all AN patients.

Notes

1. Rituximab, glucocorticoids, and plasma exchange can all be used for the treatment of AN. With the exception of some NF186 antibody-associated AN, IVIG is less likely to be effective in the treatment of AN.
2. Rituximab is effective in most patients with AN, and it is necessary to observe closely its side effect.
3. If glucocorticoid therapy is effective and is reduced to a small dose without clinical recurrence, glucocorticoids can be continued for maintenance.
4. If the effect of glucocorticoid therapy is poor, or if there is recurrence during dose reduction, a switch to rituximab can be considered.
5. Whether the combination treatment with glucocorticoids and rituximab can reduce disability remains to be studied.
6. Plasmapheresis can be tried in acute critically ill patients with AN.
7. If glucocorticoids, rituximab, or plasmapheresis are not effective, other immunological agents such as cyclophosphamide can be used based on personal experience.

Prognosis

Most patients with AN can achieve functional improvement through rational drug use, which usually requires a long period of maintenance therapy, and some patients can relapse after drug withdrawal. The prognosis of patients with anti-CNTN1 antibodies is worse than that of patients with anti-NF155 antibodies, and nearly half of patients require wheelchair assistance. A small percentage of patients have an acute, severe

illness, or comorbid renal disease, which can lead to significant disability or death.[9,12]

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DISCLOSURE

Conflict of Interest: All authors declare that there is no conflict of interest

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