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Anti-neurofascin demyelinating diseases (Anti-NF diseases)

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Abstract

Neurofascins are proteins crucial in nerve function; when the body's immune system attacks these proteins, disorders known as anti-neurofascin demyelinating diseases occur. Both the central and peripheral nervous systems are susceptible to injury from these assaults to the myelin sheath that surrounds nerve fibers. Prominent medical problems associated with anti-NF diseases include Multiple Sclerosis (MS), Neuromyelitis Optica (NMO), and Chronic Inflammatory Demyelinating Polyneuropathy (CIDP). The combination of clinical indicators, blood testing, and imaging studies is used to diagnose anti-NF disorders. The intensity of the symptoms and the particular disease determine the course of treatment. Medications that alter the immune system, medications that halt the progression of the illness, occupational and physical therapy, pain management, and supportive care are a few of the treatments. In the treatment of patients with anti-NF illnesses, nurses are essential. They offer patients all-encompassing treatment that attends to their requirements on a social, emotional, physical, and educational level. Nursing responsibilities include patient safety, communication and advocacy, long-term planning, symptom management, immunomodulatory therapies, rehabilitation and adaptive techniques, evaluation and monitoring, education and support, and documentation.

Keywords: Neurofascin, antibodies, central nervous system, peripheral nervous system, neurologist

Introduction

Auto-antibodies that target neurofascins cause disorders known as anti-neurofascin demyelinating illnesses. The protective covering that envelops nerve fibers, known as the myelin sheath, is formed and maintained in part by proteins called neurofascins. These auto-antibodies can disrupt the myelin sheath and impede nerve function by attacking neurofascins. Both the central nervous system (CNS) and the peripheral nervous system (PNS) are susceptible to anti-neurofascin demyelinating disorders. Some examples of these diseases are:

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

The condition known as CIDP results in gradual weakening and loss of sensation in the limbs. It is brought on by inflammation of the myelin sheath around peripheral nerves. A subset of CIDP instances has been associated with auto-antibodies directed against distinct neurofascins, including NF186, NF180, NF166, and NF155. These neurofascins may play a role in the pathophysiology of CIDP since they are expressed in different regions of the nervous system.

Neuromyelitis Optica (NMO)

NMO is an uncommon condition that results in demyelination and inflammation of the spinal cord and optic nerve. It can result in neurological symptoms like paralysis and vision loss. Auto-antibodies against the CNS water channel protein aquaporin-4 are typically linked to non-motor neuropathy (NMO). Nonetheless, auto-antibodies against neurofascins, particularly in the PNS, are also present in certain NMO instances.

Multiple Sclerosis (MS)

Chronic MS disease results in CNS inflammation and demyelination. Many symptoms, including exhaustion, tingling, numbness, vision issues, balance issues, cognitive decline,

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and mood swings, might result from it. Although the precise etiology of MS is uncertain, it is believed to be an autoimmune illness. It has been discovered that auto-antibodies against neurofascins, specifically NF-155, are present in certain MS cases. The integrity of the nodes of Ranvier, which are openings in the myelin sheath through which nerve impulses are carried, depends on NF-155, a neurofascin. Antibodies against NF-155 have the potential to obstruct this mechanism and harm nerves.

NF-186 is another neurofascin that might be connected to MS. The axon beginning segment (AIS), a part of the nerve cell where action potentials are produced, is where NF-186, a neurofascin, is expressed. Anti-NF-186 antibodies have the potential to disrupt this process and modify the excitability of neurons.

Anti-NF antibodies have been found to be present in approximately 10% of MS cases.

Imaging examinations, laboratory testing, and clinical evaluations are all necessary for the diagnosis of anti-NF disorders. Depending on the exact ailment and the intensity of symptoms, different treatment methods are available. Immunomodulatory drugs, disease-modifying therapies, occupational and physical therapy, pain management, and supportive care are a few of the frequently used treatments. Comprehensive care is provided to patients with these demyelinating illnesses, addressing their specific needs as part of the nursing management of anti-NF diseases. Assessment and monitoring, education and support, symptom management, immunomodulatory medicines, rehabilitation and adaptive techniques, psychological support, patient safety, communication and advocacy, long-term planning, and documentation are some of the essential components of nurse management.

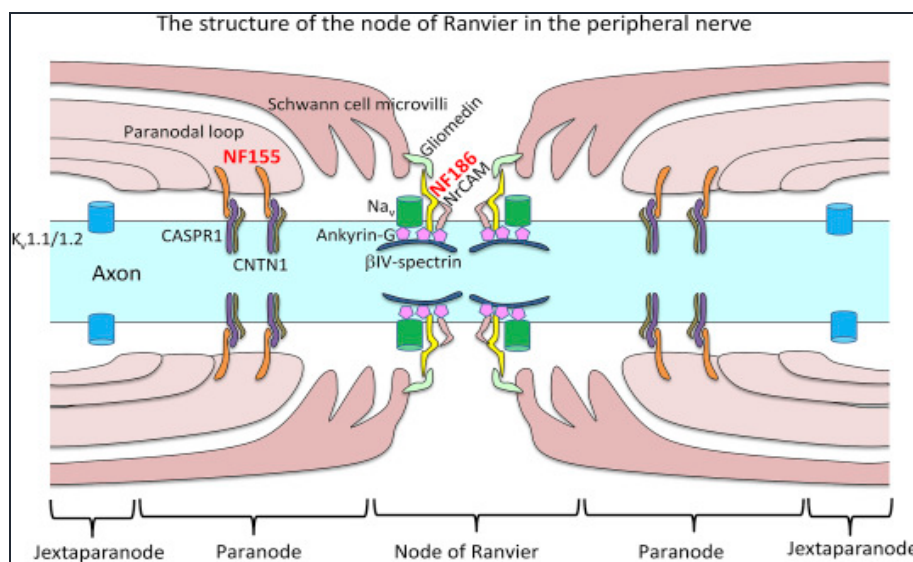


Fig 1: Anti Neurofascin Antibody

Signs and symptoms of anti-neurofascin demyelinating diseases

The symptoms of anti-neurofascin (anti-NF) diseases can vary depending on the specific condition and whether it affects the central nervous system (CNS) or the peripheral nervous system (PNS). Here are some common symptoms associated with these diseases:

Peripheral Nervous System (PNS) Involvement Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

- Progressive weakness and sensory loss in the limbs (arms and legs).
- Tingling or numbness.
- Reduced reflexes.
- Difficulty walking or maintaining balance.
- Pain, especially in the legs.

Neuromyelitis Optica (NMO)

- Severe optic neuritis (inflammation of the optic nerve), leading to vision loss or blindness.
- Transverse myelitis (inflammation of the spinal cord), causing weakness, sensory disturbances, and bladder dysfunction.
- Attacks may be relapsing-remitting.

- NMO is often associated with anti-aquaporin-4 antibodies, but anti-NF antibodies can also play a role.

Guillain-Barré Syndrome (GBS)

- Acute onset of weakness, often starting in the legs and spreading upward.
- Sensory symptoms (tingling, numbness).
- Reduced reflexes.
- Can progress rapidly and lead to paralysis.
- GBS is not exclusively associated with anti-NF antibodies but may overlap with CIDP.

Central Nervous System (CNS) Involvement Multiple Sclerosis (MS)

- Variable symptoms depending on the affected CNS areas.
- Visual disturbances (optic neuritis).
- Weakness, spasticity, and coordination difficulties.
- Fatigue.
- Cognitive changes.
- Anti-NF antibodies are found in some atypical MS cases.

Other CNS Disorders

- Some anti-NF antibodies may contribute to other

demyelinating CNS disorders, but research is ongoing.

Diagnostic Evaluation

Clinical Assessment

- A neurologist assesses the patient's neurological examination results, medical history, and specific symptoms (such as weakness, changes in sensation, or visual problems) to help lead the diagnosis procedure.
- It could also be significant if there is a family history of autoimmune diseases or other disorders.

Laboratory Tests

Antibody Testing

- Blood tests can detect anti-NF antibodies.
- Anti-NF antibodies are associated with various demyelinating conditions, including CIDP, NMO, and atypical MS.
- Testing for specific neurofascin subtypes (NF155, NF186, etc.) helps identify the underlying disease.

Cerebrospinal Fluid (CSF) Analysis

- Lumbar puncture (spinal tap) provides CSF samples.
- Elevated protein levels or abnormal cell counts may indicate demyelination.
- Oligoclonal bands in CSF are seen in some MS cases.

Imaging Studies

Magnetic Resonance Imaging (MRI)

- Brain and spinal cord MRI scans help visualize demyelination.
- Lesions (plaques) in specific areas suggest MS or NMO.

Nerve Conduction Studies (NCS) and Electromyography (EMG)

Assess nerve function and detect demyelination in peripheral nerves (CIDP).

Visual Evoked Potentials (VEP)

Measures visual pathway conduction (useful in MS and NMO).

Clinical Criteria and Exclusion

- Diagnosis is based on established criteria for each condition (e.g., McDonald criteria for MS).
- Other potential causes of demyelination (infections, metabolic disorders) must be ruled out.

Treatment

The treatment options for anti-neurofascin (anti-NF) diseases depend on the specific condition and the severity of symptoms. Here are some approaches commonly used:

Immunomodulatory Therapies

- **Corticosteroids:** These anti-inflammatory drugs can help reduce immune system activity and manage symptoms.
- **Intravenous Immunoglobulin (IVIG):** Infusions of immunoglobulins can modulate the immune response.
- **Plasma Exchange (Plasmapheresis):** Removes harmful antibodies from the blood.

Disease-Modifying Therapies

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)

1. Intravenous Immunoglobulin (IVIG) and corticosteroids are commonly used.
2. Immunosuppressants (such as azathioprine or mycophenolate) may be considered for refractory cases.

Neuromyelitis Optica (NMO)

- High-dose corticosteroids during acute attacks.
- Immunosuppressants (azathioprine, mycophenolate, rituximab) for long-term management.
- Aquaporin-4 antibody-targeted therapies (e.g., eculizumab).

Multiple Sclerosis (MS)

- **Disease-modifying therapies (DMTs):** These aim to reduce relapses and slow disease progression.
- Examples include interferons, glatiramer acetate, and newer oral or infusion-based DMTs.
- Monoclonal antibodies (such as ocrelizumab, natalizumab) may be used.
- Symptomatic management for specific symptoms (e.g., muscle spasticity, fatigue).

Physical and Occupational Therapy

1. Rehabilitation programs help maintain function, improve mobility, and manage symptoms.
2. Exercises, gait training, and adaptive techniques are essential.

Pain Management

Neuropathic pain in anti-NF diseases can be challenging. Medications like gabapentin or pregabalin may help.

Supportive Care

- Addressing specific symptoms (e.g., bladder dysfunction, visual impairment).
- Psychological support for coping with chronic conditions.

Individualized Approach

1. Treatment plans are tailored to each patient's needs.
2. Regular follow-up and monitoring are crucial.

Remember that early intervention and close collaboration with a neurologist are essential for optimal outcomes.

Nursing Management

The nursing management of anti-neurofascin (Anti-NF) diseases involves comprehensive care to address the unique needs of patients affected by these demyelinating conditions. Here are essential aspects of nursing management:

Assessment and Monitoring

1. Regular assessment of neurological symptoms, functional status, and disease progression.
2. Monitor for signs of relapse, worsening symptoms, or treatment-related adverse effects.
3. Assess pain levels and provide appropriate pain management.

Education and Support

1. Educate patients and their families about the disease, its course, and treatment options.
2. Provide emotional support, as these chronic conditions can significantly impact patients' lives.
3. Encourage adherence to treatment plans and follow-up appointments.

Symptom Management

Collaborate with the healthcare team to manage specific symptoms

- **Weakness:** Assist with mobility, transfers, and activities of daily living.
- **Visual Impairment:** Support patients with vision loss due to optic neuritis.
- **Bladder Dysfunction:** Address urinary symptoms and provide guidance.
- **Fatigue:** Help patients conserve energy and prioritize activities.
- **Pain:** Assess and manage neuropathic pain.

Immunomodulatory Therapies

1. Understand the medications used (e.g., corticosteroids, IVIG, plasma exchange).
2. Monitor for treatment-related side effects (e.g., infections, infusion reactions).

Rehabilitation and Adaptive Techniques

Collaborate with physical and occupational therapists

1. Design individualized exercise programs.
2. Teach adaptive techniques for daily living.
3. Promote independence and functional improvement.

Psychosocial Support

1. Address anxiety, depression, and coping strategies.
2. Encourage participation in support groups or counseling.

Patient Safety

1. Falls prevention strategies for patients with weakness or balance issues.
2. Educate patients about potential risks during relapses (e.g., falls, visual disturbances).

Communication and Advocacy

1. Advocate for patients' needs within the healthcare system.
2. Communicate effectively with other team members (Physicians, therapists, social workers).

Long-Term Planning

1. Discuss long-term goals, lifestyle adjustments, and community resources.
2. Involve patients in care planning and decision-making.

Documentation

1. Maintain accurate records of assessments, interventions, and patient responses.
2. Document patient education and communication.

Conclusion

Anti-neurofascin diseases are a group of disorders caused by auto-antibodies that target neurofascins, proteins that are involved in the formation and maintenance of the myelin

sheath that surrounds nerve fibers. These antibodies can damage both the central and peripheral nervous system, leading to various symptoms such as weakness, numbness, pain, vision loss, and cognitive impairment. Anti-neurofascin diseases include chronic inflammatory demyelinating polyneuropathy (CIDP), neuromyelitis optica (NMO), and some cases of multiple sclerosis (MS) and combined central and peripheral demyelination (CCPD). The diagnosis and treatment of these diseases depend on the specific type of neurofascin antibody, the clinical presentation, and the response to immunotherapy.

Conflict of Interest

Not available

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Not available

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