PERIPHERAL NERVE HYPEREXITABILITY

DEFINITION

- Group of disorders characterized by muscle stiffness, cramps, and muscle twitches with involuntary abnormal electrical activity on needle EMG.
- It can be subclassified as:

PRIMARY SECONDARY

The main primary syndromes are neuromyotonic ,cramp fasciculation syndrome and Morven's syndrome ,voltage gated potassium channel autoimmunity

Secondary PNH syndromes are generally seen in patients with focal or diffuse diseases affecting the peripheral nerves.

They can also be classified as IMMUNE MEDIATED & GENETIC.

The 3 causes of NMT are acquired ,paraneoplastic ,hereditary

Acquired form is most common ,which is causes by antibodies against neuromuscular junction

Autoimmune neuromyotonia is typically caused by antibodies that bind to potassium channels on the motor nerve resulting in continuous/hyper-excitability

ISAACS SYNDROME

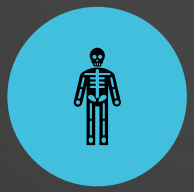
SIGNS AND SYMPTOMS

MUSCULAR HYPERACTIVITY

MUSCULAR CRAMPS STIFFNESS

WALKING DIFFICULTIES

HYPERHYDROSIS



MYOKYMIA-MUSCLE TWITCHING WITH A RIPPLING APPEARANCE

FASCICULATIONS

3 MAIN TYPES ARE

- Chronic
- Monophasic
- Relapsing remitting

TREATMENT

- Anticonvulsants – phenytoin, carbamazepine
- Plasma exchange , IVIG
- Botox injection can provide short term relief

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DIAGNOSIS

Continuous muscle contraction –myokymia in face and hands, rhythmic tics or twitches and muscle cramps.

Dx is confirmed by EMG

Associated with painful muscle cramps and muscle twitches ,muscle in the legs thighs and calves are most commonly affected

Burning prickling sensation ,muscle stiffness, hyperreflexia.

CRAMP FASCICULATION SYNDROME

CAUSES

Genetic autoimmune dysfunction

Peripheral neuropathy

Metabolic abnormalities

Tumors most commonly thymoma

DIAGNOSIS

- Depends on characteristic symptoms such as muscle cramps, twitching and pain often worsened with exercise without muscle weakness or wasting .
- EMG

TREATMENT

Carbamazepine, gabapentin, lamotrigine or pregabalin.

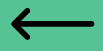
PREDNISONE used to treat cases of CSF that do not respond to other treatments.

MOORVANS SYNDROME

Muscle weakness, fatigue ,muscle twitching excessive sweating salivation ,small joint pain

Weight loss

Visual ,auditory hallucination ,complex behavior during sleep and progressive nocturnal insomnia, severe constipation, enacted dreams



Mechanism –antibodies to voltage gated potassium channel

TREATMENT

- Plasmapheresis
- Prednisone
- Haloperidol

EPISODIC ATAXIA TYPE 1

Characterized by attacks of generalized ataxia induced by emotion and stress, with myokymia both during and between attacks.

Mutation of kcna1 which encodes the voltage gated potassium channel are responsible for this subtype of ataxia.

Treatment options include carbonic anhydrase inhibitors and phenytoin.