04/6/24 Neuro Morning Report with @CPSolvers

***"One life, so many dreams" Case Presenter**: Kirtan Patolia(@KirtanPatolia) **Case Discussants**: Aaron Berkowitz, Gerardo(@gerarlunap) and Rahul*

"When stars align: A Soul-Stirring diagnosis"

CC: Numbness and tingling in lower extremities and difficulty walking for 3 months

HPI: 23 M, endorses progressively worsening tingling in all toes last 3 months, onset of symptoms are insidious, he doesn't recall any antecedent flu like symptoms, nausea, vomiting, diarrhea, trauma. Tingling accompanied by numbness extending to the level of thighs. He also has unsteady gait, recurrent falls, difficulty in standing while eye closed, now he has paresthesia in fingers.
No weakness, burning pain, diplopia, hearing changes, vertigo, headaches, fevers, chills, SOB, rashes, photophobia, bowel or bladder disturbances.

PMH: none

Meds: none

Fam Hx:
No family history

Soc Hx: no illicit drug use, no OTC supplements

Health-Related Behaviors:

Allergies:

Vitals: unremarkable
Exam: Gen: He is in wheelchair.
Mental Status: AOx3
Cranial Nerves: intact. No nystagmus.
Motor: 5/5 throughout.
Sensory: decreased light touch pinprick temperature till level of ankles and elbows. Severely reduced vibration and proprioception till ankles. Romberg+.
Reflexes: plantar downgoing; 1+ in UE; absent in LE
Coordination: dysmetria F-N and H-S. Terminal tremor.
Other: gait: wide based gait in all position

Notable Labs & Imaging:
Basic lab: CBC BMP LFT normal.
HIV, lyme, hepatitis: neg
ANCA ANA, RF, ESR, CRP neg.
Mercury arsenic and heavy metal screening: negative

Imaging:
MRI brain: normal, no spine MRI.
CT chest abdomen pelvis: no sign of malignancy.
EMG/NCS: symmetric demyelinating sensory and motor in extremities. No conduction blocks.
CSF: OP nl, WBC 1, Glucose nl, Protein: 400.
SPEP SIFE: normalAnti MAG: negative

Initially dx with CIDP. Unresponsive to steroids, IVIG. Started rituximab but did not continue tx. Plex added w/no response. Worsened w/ motor symptoms, contractures, OM in fingers.
Neurofascin 155 antibody positive, IgG4 positive.

Dx: Anti-Neurofascin Antibody Related Neuropathy

Problem Representation: 23yM w/out PMHx p/w subacute sensory > motor demyelinating polyneuropathy w very high levels of protein in CSF, unresponsive to steroids and IVIG.

Localization: Numbness and tingling: somatosensory pathway.
Bilateral LE: Below T2 → spinal cord, roots, peripheral nerves. LMN signs: periphery.
Sensory: Excludes NM junction or Muscle. All sensory modalities affected: large and small fibers.
Ataxia: Sensory vs Cerebellar. Atypical CIDP: consider paranodal instead of myelin affection.

Teaching Points (Anmolpreet): **#EndNeurophobia**
I] Numbness and tingling in LE (subacute): length dependent neuropathy? - means symptoms begin in toes because of the longest nerve fibres from spinal cord :

- **Axonal** is length dependent – toxic, metabolic (Alcohol? DM? *Metformin*? Vit B12?)
- **Demyelinating** is non length dependent (both proximal & distal n. fibers affected) – immune mediated (GBS, CIDP)

Somatosensory pathway: sensory receptors→ brain
Different sensations from different pathways. 2 ascending pathways:

1. Dorsal column (heavily myelinated) : pressure, vibration, fine touch, propriocep
2. Anterolateral column-Spinothalamic tract (lightly myelinated) : pain, temp

II] Difficulty walking: what is required? strength*(weak)* , sensation*(numb)* and coordination(*ataxic)*, vision, vestibular function
III] Paresthesia in hands: expanding cord lesion? Most neuropathies we see are Sensory, symmetric, length dependent, chronic.--> axonal dependent.- labs! - Vit B12, HbA1C
Paraprotein associated neuropathy:- subacute neuropathy→ SPEP, light chains
The patient is found to have pure sensory loss!
IV] Ganglionopathy: affecting dorsal root ganglion. Both large and small fibers seem to be affected. Polyradiculoneuropathy? Seems to be sensory ataxia.
V] Ataxia: Cerebellar/Sensory? seemed cerebellar ataxia because of terminal tremor. In CIDP, there can be a tremor with ataxia→Subacute demyelinating neuropathy. CIDP → median age = 50 years
VI] NEW ds! → Nodo or Paranodopathies looks like CIDP! But has associated **Paranodal or nodal**(Node of Ranvier)**antibodies** (most common antibody- **Neurofascin-155**→ IgG4 class; Other antibodies: NF186(Nephrotic), CNTN1, CASPR1) and don't respond to IVIG like CIDP; responds to Rituximab; *therefore empiric treatment with IVIG can help!*
VII] Albuminocytologic dissociation (high protein and low WBCs) : non specific pattern of **inflammation** ; could be GBD, CIDP, Amyloidosis, Paraproteinemia, POEMS, DM, Refsum, CSF Block(Froin's Syndrome)