01/24/25 Morning Report with @CPSolvers

"One life, so many dreams" **Case Presenter**: Hugh Larkin **Case Discussants**: Mark Heslin and Rabih Geha

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Chief Concern: 51yo female with progressive limb weakness, bulbar symptoms & *respiratory compromise.*

HPI:

Dx with Grave disease 1 month ago - started carbimazole & propranolol and then subsequently developed *bilateral leg paresthesias* that spontaneously improved, and patient discharged home after a normal MRI (brain and cord), nerve conduction studies and an LP.

Further Information

Since discharge - *profound weakness with inability to walk;* associated with **weight loss** despite having an appetite along with dysphagia. No bowel/bladder/sensory/vision issues.

Admitted to ICU due to low FVC

PMH:
Previously excellent baseline

Fam Hx:
Insignificant

Health-Related Behaviors:
No substances.
No travel

Allergies: None

ICU admission

Started on steroids and IVIG.
Ophthalmoplegia & FVC worsening requiring intubation.
No improvement after one week and started on rituximab

Vitals: O2 98% (RA), RR 18, BP 126/81, HR 110, apyrexial
Exam: CVS, Pulmonary Abdominal exams all normal
Neurologic: Diplopia but no ophthalmoplegia. Sarcopenic in arms and legs. No fasciculations. Normal tone, coordination and sensation. Upper and lower limbs: **4 out of 5 proximal leg and arm weakness** with normal distal strength
FVC: <1 liter

Notable Labs & Imaging:

Hematology:
WCC 9, Plat 320, Hb 17

Chemistry
Urea 5.6 (normal), **Creatinine** 57 (normal) **Calcium** 2.5 (Normal). **Na** 135 **K** 4.6, **Albumin** 39 **CK** 620, **Troponin** 137, **TSH** 0.12 **free T4** 19.3 (slightly elevated)

Immunologic: **CRP** 1 **Anti-TPO & anti-TSH receptor antibodies** both *strongly positive.*
ANA/ANCAs negative. Negative tumor markers.

Imaging
EMG: Clear myopathic changes.
NMJ studies: normal. **Autoimmune neurology panel**: Negative
Myositis panel: Anti-Ro 52 positive
Muscle biopsy: Inflammatory cells around necrotic fibers, muscle fiber phagocytosis, NMJ disruption and AcH-receptor architecture.
AcH receptor antibody: Markedly elevated **MuSK antibody** negative.
MRI Thymus: Cystic Hyperplasia

Dx: Mixed Myasthenia Gravis and Myositis (MMM)

Problem Representation: 51yo female recent Dx of antibody(+) Grave Dz, now admitted to ICU for severe progressive proximal muscle weakness with respiratory compromise, with antibodies returning positive against AcH-receptors & Ro 52, with the final diagnosis of Myathenia Gravis & Myositis

Teaching Points (Anmolpreet) :

I] Grave's disease: confirmatory test for final diagnosis. Side effects of carbimazole to be looked out for: agranulocytosis→ CBC with DLC

II] Acute neuropathy: heavy-metal toxicity, Acute Intermittent Porphyria, AIDP, Guillain Barre Syndrome

III] CNS vs PNS ds: bulbar weakness and respiratory compromise: PNS > CNS

IV] Lower limb weakness: *lumbar disc disease*. Ask for numbness, tingling, bowel/bladder symptoms (bladder scan), radiating pain– which localises it to the back; *tumor in an unusual location in spinal cord, degenerative disease*
Localisation: spinal cord and below→ nerve root →nerve→ neuromuscular junction→muscle; regions to be ruled out :-
Brain-absence of cortical symptoms -
Spinal cord– absence of bowel/bladder symptoms
LEMS- absence of profound autonomic symptoms
?Myasthenia gravis: beta-blockers/thyrotoxicosis triggering myasthenic crisis?
Next step in a patient with head-to-toe symptoms and intact sensorium: Lumbar puncture

V] Thyrotoxic Periodic Paralysis (TPP): intermittent sudden onset of muscle weakness, especially in lower extremities; in the setting of hypokalemia

VI] Diaphragmatic weakness: leads to hypercapnia and acidosis. There is increased hypercapnia required to make the patient hypoxemic. Forced vital capacity is done to test.

VII] Proximal muscle weakness: localisation in muscle (high chance), NMJ (diplopia), nerve (atrophy in arms); myopathies can be due to:

- Immune-mediated myopathy (inflammatory)
- Metabolic myopathy (endocrine)

VIII] Empiric Rx: ?IV IG, ?Pyridostigmine; **Next step: EMG**

IX] MDA5 is the rapidly progressive one; *next step*: MRI muscle + biopsy

X] Overlap between NMJ ds and myositis