Flight to a Hospital in Paradise: A Case of Acute Onset Ataxia in a Healthy Child

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Background

- With the successful eradication of polio in the USA, due to a consistent and through vaccination program, Guillain-Barre Syndrome (GBS) is now the most common cause of acute flaccid paralysis in healthy children.
- Once thought to be a single acute immune-mediated polyneuropathy, it is now known to have a multitude of variants, the most common being Acute Inflammatory demyelinating polyneuropathy, Acute Motor Axonal Neuropathy, Acute Motor and Sensory Axonal Neuropathy and Miller-Fisher Syndrome (MFS).
- MFS occurs in 5% of cases in the USA, and is characterized by a trifecta of areflexia, ophthalmoplegia and ataxia.

Case Presentation

- Patient is a 15-year-old white female, with no significant past medical history, who presented to the emergency department for evaluation of generalized weakness, abdominal pain, slurred speech, and ataxia.
- Two days prior to presentation, patient had acute onset nausea, vomiting, diarrhea and mild abdominal pain.
- Originally from Iowa, patient and her family were travelling to Myrtle Beach, South Carolina (SC) for spring break.
- Patient was mildly lethargic prior to boarding the plane; however, on landing in SC, patient's symptoms evolved to ataxia, and was unable to walk, needing a wheelchair to be transported to Grand Strand Medical Center.
- Labs were largely within normal limits, including CRP, ESR, CBC, infectious mononucleosis, Covid-19 and TSH.
- CT and MRI of the brain, and the cervical, thoracic, and lumbar spines were all unremarkable with no noted acute abnormalities.
- A lumbar puncture was performed, which yielded normal CSF analysis, including no albuminocytologic dissociation.
- Physical exam, however, was significant for upper and lower extremity ataxia, dysarthria, ptosis, and diminished reflexes bilaterally in both the upper and lower extremities.
- Initially she treated empirically with a one-week course of twice a day steroid, with mild improvement in ataxia.
- Through shared decision making with the family, it was decided to have patient follow up in Iowa City Hospital for further evaluation and treatment with intravenous immune globulin (IVIG) therapy with possible plasmapheresis.
- On initiation of IVIG therapy, our patient made progressive recovery, and with rehabilitation, patient was able to re-start school in 3 weeks.

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Labs

Vital Signs: Temp 96.5°F HR 60 BP 126/75 O^2 Sat 98%

140	106	8.0
4.1	23	0.6

5.9	14.8	263

Routine Labs
C-Reactive Protein: < 0.5
TSH: 3.09 T4: 1.09
Cortisol: 17.4. Parathyroid: 31.9
Urine Drug Screen: Negative
Pregnancy: Negative

Serum Protein Electrophoresis:
- **No M-spike observed**
- **Albumin 1.2 Alpha 1 0.3 Alpha 2 1.1 Beta 1.0 Gamma 0.9**

Select Autoimmune Labs:
- **Anti-Muscle Specific Kinase: < 1**
- **Anti-Nuclear Antibody: Negative**
- **Acetylcholine Receptor Antibody: < 0.03**

Cerebral Spinal Fluid:
- **Clear, Colorless**
- **Glu 60 Total Prot. 32 WBC 3 RBC 22**

Heavy Metals

	Levels
Ceruloplasmin	36.5
Copper	157 (H)
Arsenic	1
Lead	< 1

Vitamins

	Levels
B1	111.9
B6	6.1
B12	759
Methylmalonic Acid	138
Folate	8.4

Images

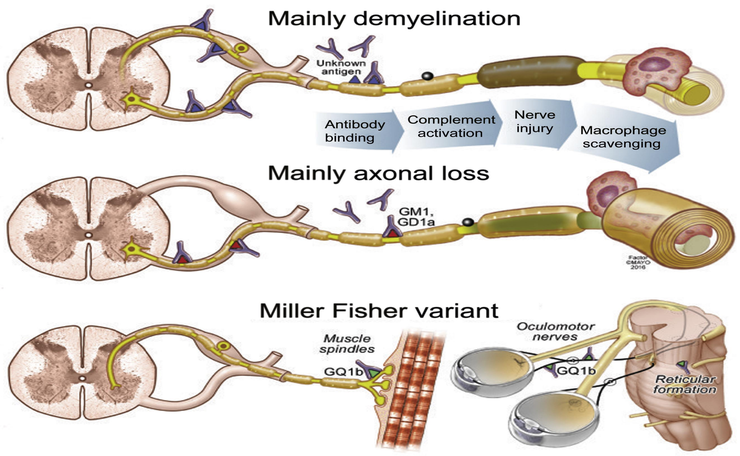

Figure 1: Immunoglobulin Targets in GBS and its Variants.

Discussion

- In MFS, immunoglobulins target gangliosides GQ1b on the neuromuscular junction (Figure 1).
- A minority of patients (10-30%) are Gq1b seronegative.
- Pathophysiology is thought to be related to molecular mimicry between peripheral nerves and microbial/viral antigens
- The association to infection is not limited to Campylobacter *jeujuni*, but also cytomegalovirus, influenza A, Mycoplasma *pneumoniae*, H. *influenza*, hepatitis A,B,E, to name a few.
  - MFS usually follows nonspecific respiratory or gastrointestinal infections.
- While pediatric forms of MFS usually have mild presentation with mainly ocular involvement, distal paresthesia have also been reported, such as in our case.
  - Additionally, there have been reported cases involving respiratory failure and transient comas.
- Supportive care is the mainstay therapy.
- Plasma exchange and IVIG improves recovery time.
- In our case, the strong prevalence of dysarthria, ptosis and ataxia on physical exam and the recent gastrointestinal symptoms on history strongly support the diagnosis of MFS
- As in this case, due diligence in ruling out other possible diagnoses was performed through imaging and lab studies.

Conclusion

- In cases suspicious of GBS with MF variant, presenting with normal imaging and labs, physicians should engage logical clinical judgement within the appropriate clinical settings, to employ accurate management to avoid significant morbidity and mortality.

References

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