Acute idiopathic polyneuritis classified under the umbrella term of Guillain Barre Syndrome (GBS) is a variegated condition with several forms. GBS classically presents as an acute paralyzing illness initiated by a preceding infection or vaccination. The dominant features are progressive, bilateral muscle weakness accompanied by absent or very diminished deep tendon reflexes. However one variant, rapidly progressive sensory ataxic neuropathy, is a rare subset of GBS that presents with predominant sensory neuropathy, ataxia, and antibodies to GD1b.

Our discussed patient did not show any motor weakness, but had severe progressive ataxia and paresthesias, two falls, inconclusive serologic studies, and no reported preceding infection. Because these features somewhat mimic those of an ischemic stroke, TIA, or central demyelinating disease, clinicians must be aware of the variants of GBS to ensure rapid diagnosis and treatment to prevent long term complications of the disease and shorten rehabilitation length.

Introduction

GBS is a heterogenous disorder with a worldwide prevalence of 1-2/100,000 people/year with an incidence that increases by 20% after every decade of life after age 10. GBS has many variants with its own specific pathophysiological, pathological and clinical findings such as Acute inflammatory demyelinating polyradiculoneuropathy (AIDP) representing 85-90% of cases in the US and Europe, and Fish Miller Syndrome characterized by ophthalmoplegia, ataxia, and areflexia, representing only 5% of cases. However, rapidly progressing sensory ataxic neuropathy (RPSAN) is an extremely rare variant characterised by acute, monophasic, sensory neuropathy accompanied by reduced muscle stretch reflexes, CSF albuminocytological dissociation and nerve conduction features of demyelination; and sensory ataxia.

GBS is most often caused by a preceding infection from Campylobacter jejuni, and studies show that infection with this organism results in a 100 fold increased risk for developing GBS, and an overall worsened prognosis with slower recovery and greater neurologic disability. However GBS has also been associated with preceding influenza infection, HIV infection, cytomegalovirus, Epstein-Barr virus, varicella-zoster virus, herpes simplex virus, and hepatitis A, B, C, and E viruses. Also the bacteria Haemophilus influenzae, Escherichia coli, and Mycoplasma pneumoniae have been implicated but the epidemiology is not well established.

The pathophysiology of GBS RPSAN is thought to be due to a preceding infection that causes an immune response that cross reacts to the peripheral nerve or the axons of the peripheral nerve. Responses can be directed to the myelin or the axons of the peripheral nerve.

A 56 year old male presented to the ER with a 1 weeks history of increasing LE muscle weakness and two reported falls. He also reported severe low back pain and headache. The patient had been to the ER 3 days prior where a CT and MRI of the brain were performed with no abnormalities noted and patient was discharged. The patient continued to have weakness and went to his PCP where the patient was then sent to the ER again. The patient reported a trip to Mexico 4 weeks ago and denied any infection, nausea, vomiting, or diarrhea while on that trip or after. However the patient noticed “tightness in his legs”, and progressive numbness on the bottom of his feet and in his genitals. Two days before admission the patient noticed numbness and tingling in his hands and tongue and much greater loss of balance requiring someone to hold him up to walk.

Physical Exam:

Neurological: Mild to moderate headache and back pain. Fluent speech and appropriate to questions. No facial droop, EOMI, no nystagmus or diplopia. Bilateral sensory loss and paresthesias in the lower extremities from the toes to the ankle. Diminished sensation and proprioception in the lower extremities. Cerebellar examination reveals dysmetric finger to nose testing. Reflex examination demonstrates areflexia in all extremities.

Muskuloskeletal: 5/5 muscle strength bilaterally in upper and lower extremity. Normal plantar and dorsiflexion. Unable to bear weight.

OMM: Significant muscle hypotonicity and tissue texture changes of the hamstrings with hip flexion. Somatic dysfunction of L4-S1 with tissue texture changes and hypotonicity of the paraspinal muscles.

Intervention: IVIG and Plasmapheresis were started upon ruling acute cerebellar or spinal demyelinating etiologies.

References


Further Workup

Patient had an acute UTI with sepcticemia that required rehospitalization 7 days after discharge since the patient was unable to feel normal sensations in his bladder or with urination due to the sensory neuropathy. The patient was readmitted a third time one week later for increasing sensory abnormalities and paresthesias requiring retreatment with IVIG and plasmapheresis. In the outpatient neurology clinic EMG studies showed significant demyelination suggesting severe demyelination and a very prolonged recovery period. The patient to date is still experiencing persistent ataxia and sensory loss and also new onset vertigo and is undergoing further testing with his neurologist.

Conclusions

GBS should be in the differential for any patients with progressive bilateral neurologic deficits with recent travel to outside countries and a history of upper respiratory (URI) or gastrointestinal (GI) symptoms in the past 30 days. However a lack of a GI or URI prodrome should not exclude GBS from the differential diagnosis. Furthermore a lack of muscle paralysis should not exclude GBS from the differential due to the spectrum of the disease and its many variations. Patients may also have relapsing events and can continue to develop chronic inflammatory demyelinating neuropathy that must be closely monitored by an outpatient neurologist.