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Posterior Reversible Encephalopathy Associated with the Dysautonomia of Guillain-Barré Syndrome

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Guillain-Barré Syndrome (GBS) generally presents with paresthesias, ascending weakness, and areflexia, although dysautonomia is a well-known phenomenon. Posterior Reversible Encephalopathy Syndrome (PRES) classically presents with headache, altered alertness, confusion, seizures and abnormalities of vision; hemianopia, visual neglect, and cortical blindness may also occur. There are a few reports that describe PRES in association with GBS prior to treatment, and authors speculate that the dysautonomia of GBS leads to acute elevations in blood pressure (BP) and PRES in these unusual cases. This conclusion stems from the high improbability of both rare diseases occurring simultaneously purely by coincidence. A couple of studies have examined the use of urinary catecholamines to measure dysautonomia in GBS, and both conclude that elevated urinary catecholamines correlate with clinical dysautonomia in GBS. We describe a patient with concurrent PRES and GBS whose urine catecholamines were elevated when most symptomatic and then normalized with recovery, which helps to support the theory that the dysautonomia of GBS led to the

patient's acute elevation in blood pressure and ultimately PRES.

Case Report

A 73-year-old normotensive male presented to an outside hospital with a one-day history of difficulty opening envelopes, turning keys and "numbness" in his feet. He reported a sore throat a week before. During the next five days his weakness progressed to include both proximal upper and lower extremities, although deep tendon reflexes remained. An MRI of the cervical spine was unremarkable. Lumbar puncture revealed no leukocytes and a protein of 93. He was subsequently transferred to our hospital with suspected GBS.

Upon arrival, the patient complained of visual loss and headache. His BP was 210/110. He was visually inattentive to his left side and had a left visual hemianopia. Immediate head CT showed bilateral occipital hypodensities consistent with PRES, which subsequent MRI confirmed (Figure 1). He was immediately treated with intravenous labetalol, and visual symptoms resolved within

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Table 1: Urine Catecholamines

24 Hour Urine Metanephrines	2 nd day of admission µg/24 hours (NL)	*14 days later µg/24 hours (NL)
Metanephrine	484 (44-261)	200 (35 – 460)
Normetanephrine	1030 (468-560)	958 (110-1050)

NL = normal lab values

*outside laboratory

45 minutes. Other significant exam findings at that time included quadraparesis and absent reflexes except for trace brachioradialis reflexes. Electromyography (EMG) demonstrated an acute mixed motor and sensory demyelinating polyneuropathy. We diagnosed GBS and treated the patient with five sessions of plasmapheresis.

The patient required three days of intravenous labetalol and nicardipine and then was transitioned to oral amlodipine. He underwent investigations to work up hypertension: EKG and MRI of the renal arteries were normal. Urine catecholamines on hospital day two were elevated and repeat levels upon resolution of signs and symptoms at the rehabilitation hospital were normal (Table 1).

Discussion

Our patient initially presented with characteristic GBS symptoms, and spinal fluid and EMG supported the diagnosis. Prior to any treatment, including intravenous immunoglobulin (IVIg), the patient developed PRES. Known precipitants are acute elevations of BP, renal decompensation, immunosuppressants and IVIg. We surmise that the dysautonomia of GBS led to hypertension and ultimately PRES in this patient.

Review of the literature shows a number of case reports describing GBS in combination

with PRES, although most are related to treatment of GBS with IVIg. Many of the cases unrelated to IVIg are in the pediatric literature, but three case reports describe simultaneous GBS and PRES prior to treatment in adults.¹⁻³ In all three cases, patients improved with BP control and treatment with IVIg. These authors suspected GBS dysautonomia-induced hypertension as the etiology of PRES. However, they based this conclusion without additional data, on the unlikelihood of both diseases coincidentally occurring concurrently.

Two studies have examined the use of urinary catecholamines to measure dysautonomia in GBS.^{4,5} In one study,⁴ 7 of 25 patients with GBS had dysautonomia, and their urinary measurements revealed high values at the height of paralysis. With recovery, the catecholamines returned to within normal limits. In a second study,⁵ 10 of 12 patients with GBS dysautonomia had an increase of urinary methoxylated metabolites. Patients without clinical dysautonomia had normal values. The authors concluded that elevated urinary catecholamines correlate with clinical dysautonomia in GBS and can be used as a biological marker. Our patient had elevated urine catecholamines when most symptomatic, and these values normalized with recovery. These findings help support our theory that the dysautonomia of GBS led to PRES in our patient.

However, the literature reveals cases in which patients develop PRES after IVIg treatment for GBS,^{6,7} although no clear mechanism has been described. The three above-cited cases all treated these patients with IVIg with no reported complications. We treated our patient with plasmapheresis because of the concern of exacerbating PRES with IVIg.

Our case adds to the growing literature of patients with simultaneous GBS and PRES. We treated the patient with plasmapheresis, because IVIg also poses an independent risk of PRES. However, further research is necessary to determine if urine catecholamines are a useful marker of GBS dysautonomia and if there are any differences in outcomes for patients with concurrent GBS and PRES treated with IVIg versus plasmapheresis.

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Figure 1: MRI of brain, FLAIR sequence

