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Bipin Ghimire

Samiksha Pandey

Ashbina Pokharel

Emma Herrmann

Mohammad Muhsin Chisti

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Program: Beaumont Health Royal Oak
Program Director: Sandor Shoichet, MD, FACP
Presenter: Bipin Ghimire
Additional Authors: Dr. Samiksha Pandey, Dr. Ashbina Pokharel, Dr. Emma Herrman, Dr. Mohammad Mushin Chisti

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With a worldwide prevalence of 3-4 per million, Lambert-Eaton syndrome is a rare entity. It often occurs as a paraneoplastic syndrome, most commonly associated with small cell lung cancer. Less frequently, it occurs as an autoimmune disease in the absence of cancer. Both entities demonstrate circulating immunoglobulins against presynaptic voltage-gated calcium channels and Electromyogram (EMG) with an increase in compound muscle action potential on repetitive stimulation.

A 35-year-old female with past medical history of hypothyroidism presented with bilateral lower extremity weakness, generalized fatigue, blurry vision, and slurred speech in 2010. Upon further workup, Lambert-Eaton syndrome was diagnosed after she tested positive for anti-voltage-gated calcium channels antibodies, with suggestive EMG. Workups including CT chest/abdomen/pelvis, Positron Emission Tomography (PET) scan, and colonoscopy were negative for evidence of malignancy. A treatment regimen was initiated which included Intravenous Immunoglobulin (IVIG) every 3-months, Mycophenolate, and intermittent Intravenous Methylprednisolone, but this provided no symptomatic relief. Next, she was started on 3-4, Diaminopyridine (an experimental drug during that time), but her symptoms persisted. She was later hospitalized with quadriparesis, which resulted in her being wheelchair-bound. At that time, Rituximab was started, which dramatically improved her symptoms and normalized her antibodies level. She is now able to walk with minimal assistance and continues to receive Rituximab therapy.

In present times, 3-4, Diaminopyridine is the first-line treatment of Non-tumor Lambert-Eaton syndrome, which has proven to be adequate therapy in most patients. Refractory cases are treated with IVIG. Further management includes immunosuppressive therapy, such as steroids, Azathioprine, Mycophenolate, or Cyclosporine. There has been scattered evidence for the use of Rituximab in patients who have failed immunosuppressive therapy. In our patient, significant improvement in symptoms with Rituximab favors the limited studies that have been completed and further adds to the body of evidence.