A Guillain-Barré Mimic with Rapid Deterioration: Pan-Neurofascin Autoimmune Nodopathy in a patient with New Onset Systemic Lupus Erythematosus

Laiba Iqbal

Background:
Autoimmune nodopathies are immune-mediated neuropathies that can resemble GBS, making diagnosis challenging. They are associated with IgG4 antibodies targeting key nodal and paranodal proteins such as NF186, NF155, CNTN1, and Caspr1. Unlike other immune neuropathies, they typically show little to no response to IVIG, whereas treatments like rituximab have demonstrated greater effectiveness. In 2021, the EAN/PNS classified autoimmune nodo-paranodopathy as a distinct form of polyneuropathy, separate from CIDP. Clinically, it is characterized by a more aggressive course, frequent cranial nerve involvement, and poor or absent response to IVIG.

We present a case of acute-onset, severe pan-nodopathy that initially mimicked GBS, ultimately associated with new-onset SLE. Although the mechanisms underlying this form of polyautoimmunity remain poorly understood, increased awareness is critical to enable early recognition and appropriate management, helping to prevent a potentially life-threatening disease course.

Case Presentation:
An 18-year-old woman with new-onset SLE, nephrotic syndrome and pneumonia presented with rapidly progressive ascending numbness and weakness, including bulbar involvement. Initial clinical findings suggested GBS, and IVIG was initiated. Despite the treatment, she deteriorated over three days to quadriplegia with respiratory failure requiring intubation. Electrodiagnostic studies demonstrated a progressive sensorimotor axonal polyneuropathy with severe active denervation. Nerve and muscle biopsies showed mild axonal loss . Given her rapid decline and lack of response to IVIG, additional immunotherapy was initiated, including PLEX, high-dose steroids, CellCept, Plaquenil, and Cytotoxan. Serologic testing ultimately identified pan-neurofascin (NF155/NF186) IgG4 antibodies, confirming autoimmune nodopathy. Rituximab was not available, so ofatumumab was initiated showed drastic improvement over the subsequent weeks.

Conclusion:
Autoimmune nodopathies can present with acute demyelinating neuropathy mimicking GBS but often demonstrate continued progression or a poor response to intravenous immunoglobulin. Recognition of this entity is important, as it carries distinct treatment implications and may require escalation to targeted immunosuppressive therapy. The

temporal association between recent infection and new-onset systemic lupus erythematosus in our patient raises the possibility of compounded immune activation contributing to her severe and rapidly progressive neurologic presentation. This case highlights the importance of reassessing the diagnosis in patients with presumed GBS who fail to improve or who demonstrate an atypical clinical course.