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“Severe GBS Variant in Newly Diagnosed SLE”

Background: Guillain-Barré syndrome (GBS) affects 1-2 people per 100,000 per year.¹ Variants include acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), and acute motor and sensory axonal neuropathy (AMSAN). AMSAN is a rare, severe variant with a prolonged recovery time.² While GBS is usually triggered by infections, there have been case reports of systemic lupus erythematosus (SLE)-associated GBS. It is estimated that the prevalence of SLE-associated GBS is 0.6-1.7%.³ This case presents AMSAN in the context of newly diagnosed SLE, emphasizing the importance of recognizing this rare overlap and considering SLE as a potential trigger and modifier of disease course.

Case Presentation: An 18-year-old female with no prior medical history presented with facial and bilateral lower extremity swelling as well as shortness of breath for one month. She was found to have a right-sided pleural effusion status post thoracentesis, which was consistent with a transudative process. She also had a new diagnosis of SLE after having positive Smith, RNP, and chromatin antibodies as well as hypocomplementemia. She was started on steroids, hydroxychloroquine, and mycophenolate mofetil. Five days into her hospitalization, she began to have ascending weakness and dysarthria, ultimately progressing to quadriplegia and respiratory failure requiring intubation. She underwent lumbar puncture. CSF studies revealed glucose 59, protein 69, and WBC 12 with lymphocytic predominance. She was initiated on IVIG for empiric treatment of GBS. Due to minimal improvement, she also received a course of PLEX. EMG was consistent with the diagnosis of GBS, and steroids were quickly tapered per neurology recommendations. She had a slow neurological recovery ultimately requiring tracheostomy. After completion of IVIG and PLEX, she was able to subtly move her fingers. She underwent sural nerve biopsy, with results currently pending.

Discussion: This case highlights the challenges of managing severe axonal GBS in newly diagnosed SLE. While most GBS cases are infection-triggered, systemic autoimmunity may exacerbate axonal injury, raising the question of whether SLE is causal or coincidental. This distinction affects management, as active SLE may warrant adding corticosteroids or other immunosuppressants to standard IVIG or plasma exchange.³ In this case, steroids were initially added due to concern that the GBS was related to her new SLE diagnosis; however, it was quickly tapered per neurology’s recommendations due to concern it may worsen her GBS, as it was unclear how much of a role SLE was truly playing. Early recognition enables a broader workup and timely involvement of specialists, potentially improving outcomes.