Fulminant Guillain Barré syndrome as the presenting clinical manifestation of systemic lupus erythematous

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Introduction

- Neuropsychiatric manifestations as the early presentation of systemic lupus erythematous have been reported up to 17.6% in retrospective studies
- These can involve the CNS or PNS and can have a wide range of severity
- Fulminant Guillain Barré syndrome is a rare complication of SLE with devastating consequences

Case Background

- A 54 year old male without significant past medical history presented to the emergency department with a chief complaint of "fatigue" and "feeling inflamed"
- He was discharged home with supportive management
- One week later, he re-presented to the ED with rapid progression of muscular weakness, dysphagia, and respiratory decline

Diagnosis of GBS

- **MRI of spine:** multiple levels of nerve root enhancement with cauda equina thickening
- **Lumbar puncture:** Albuminocytologic dissociation
- **EMG/NCS:** absent motor potentials and normal sensory potentials without evidence of demyelination
- Diagnosis of GBS-Acute motor axonal neuropathy variant was made
- Despite pulse dosed steroids, five sessions of plasmapheresis, and two rounds of IVIG, progressed to ventilator dependence, flaccid tetraplegia, facial motor paralysis, and complete areflexia

Diagnosis of SLE

- One year later, patient presented to our SCI service for wound care
- Work up for anasarca and declining renal function:
- **Proteinuria:** 2.6 g/24h
- **Lymphopenia:** $0.3x10^3$ / uL
- **ANA titer:** 1:1280 in homogenous pattern
- **C3/C4:** consistent with hypo-complement
- **Renal Biopsy:** consistent with WHO Class II Lupus nephritis
- Diagnosis of systemic lupus erythematous was made

Treatment

- Started on NIH – high dose regimen cyclophosphamide ($0.5g/m^2$ monthly IV pulses for total goal of 6 months)
- Additionally, started on steroid taper and daily hydroxychloroquine
- Patient has since demonstrated decreased ventilator dependence as well as activation of proximal muscles including deltoids, pectoralis major, and triceps

Conclusions

- Due to the impact on treatment selection, identifying SLE as the possible underlying entity when GBS is diagnosed is critical
- Immunosuppression, while not indicated in the treatment of GBS, has seen success in SLE-GBS and may promote neurological recovery.

References