



Too Weak to Breathe: The Necessity of Early Recognition of Acute Inflammatory Demyelinating Polyneuropathy

Andrew Balster, MD and Alan J. Hunter, MD, FACP

Department of Medicine, Oregon Health & Science University

Introduction

- Acute inflammatory demyelinating polyneuropathy (AIDP) is a life-threatening condition involving acute onset polyneuropathy that results in ascending areflexic weakness and paralysis.
- AIDP often has a recognizable triggering event, classically infectious. However, infections only account for two thirds of cases and has become increasingly recognized that there are also other triggering events.

Case Presentation

A 23-year-old man with Ewing sarcoma presented to the emergency department for subacute progressive weakness for one week prior to admission. Patient noted numbness in his hands and feet, which progressed to worsening bilateral lower extremity weakness complicated by several falls due to inability to support his weight.

PMH:

- Ewing sarcoma status post chemotherapy and radiation with recent recurrence of sarcoma status post recent salvage left hemipelvectomy complicated by a bladder injury requiring an intraoperative repair and bilateral nephrostomy tubes.
- Chemotherapy induced neuropathy

Social history:

Married. Domiciled. Two children. No alcohol, tobacco or illicit drug use.

Physical Exam:

Physical exam showed diffuse areflexia in upper and lower extremities in addition to asymmetric motor weakness of the left lower extremity compared to right.

Imaging:

MRI Spine Total: Questionable smooth increased enhancement of cauda equina nerve roots around the conus, could indicate inflammatory or demyelinating etiology.

EMG:

EMG showed severe diffuse demyelinating and axonal polyradiculoneuropathy supportive of the diagnosis of AIDP and chronic chemotherapy induced polyneuropathy

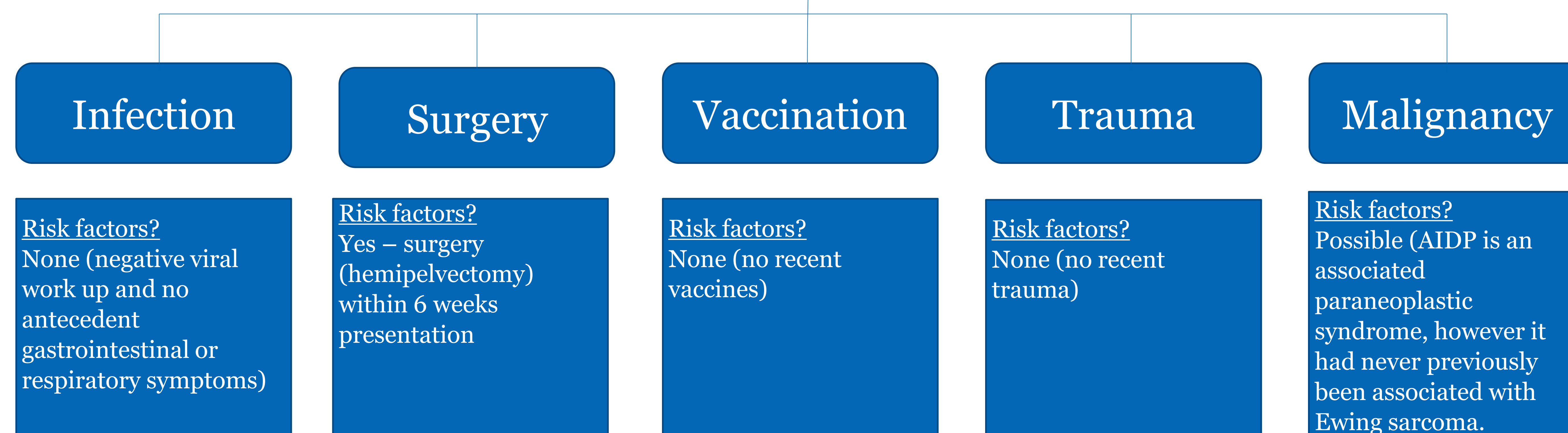
CSF Studies:

Component	Value
RBC	<1
WBC	1
Protein	113
Glucose	82
Appearance	Clear

Hospital Course

- Following diagnosis AIDP, a negative inspiratory force (NIF) was found to be 20 cm H₂O.
- Serial exams revealed a down trending NIF and within 48 hours the patient was intubated for respiratory decompensation.
- Broad infectious work up including lumbar puncture was sent, which was unrevealing.
- Treatment included IVIG without appreciable improvement.
- He subsequently underwent tracheostomy and gastrostomy tube placement.
- He was discharged to a medical rehabilitation hospital and eventually liberated from the ventilator

Possible Triggering Events



Discussion

- AIDP is classically thought to result in symmetrical ascending weakness. However, this case it is important to consider there is heterogeneity in clinical presentation. Notably, there are only two required features to diagnose AIDP, which are progressive weakness and areflexia. All other features such as symmetry are supportive of the diagnosis, though not required.
- Of importance, nearly 30% of cases of AIDP eventually involve muscles of respiration. Thus, AIDP is a time sensitive diagnosis. Additionally, once the diagnosis is made, institution of serial monitoring of parameters for respiratory muscle function is critical to identify patients at high risk for respiratory failure.
- Clinicians should be mindful that while infection is often the antecedent event, it is important to consider other triggers such as trauma, surgery, immunization and neoplasm.

Teaching Points

- AIDP is a time sensitive diagnosis.
- Following diagnosis, early and serial exams are critical to monitor for respiratory failure.
- It is important to consider other triggering events besides infection.

References

- Hocker S, Nagarajan E, Rubin M, Wijdiicks EFM. Clinical factors associated with Guillain-Barré syndrome following surgery. *Neurology: Clinical Practice*. 2018;8(3):201-206. doi:10.1212/cpj.0000000000000451
- MH O, LE D, CW R. Guillain-Barre Syndrome as a paraneoplastic syndrome from Ewing sarcoma. *Sarcoma Research - International*. 2021;6(1). doi:10.26420/sarcomaresint.2021.1046
- Newswanger DL, Warren CR. Guillain-Barre syndrome. *American Family*