Introduction
Guillain–Barré syndrome (GBS) is an acute immune-mediated polyneuropathies, which has various clinical presentations. The original description of “ascending paralysis” includes the most common variants. However, there are a number of uncommon variants of GBS. We present one unusual case of inflammatory demyelinating polyneuropathy with ascending upper extremity and descending lower extremity paralysis without bulbar or sensory involvement.

Summary
A 55-year-old man presented to the emergency department (ED) with a 3-week history of progressive descending weakness originated from bilateral upper limb with progression of weakness to his lower limbs. One week prior to his presentation, he was evaluated by his primary care physician and received 1 week of oral steroid. However, he did not notice any improvement in his symptoms.

Examination revealed symmetrical upper greater than lower limb weakness with absent deep tendon reflexes. Extensive work-up for evaluation of underlying structural, infectious, inflammatory, and paraneoplastic etiologies were negative.

Electrodiagnostic and cerebral spinal fluid (CSF) results were consistent with acute inflammatory demyelinating polyneuropathy (AIDP) (Figure 1(a) and (b)). The patient was treated with a 4-day course of intravenous immunoglobulin.

AIDP is the most common form of GBS accounting for about 85%–90% of those cases in North America, Europe, and most of the developed world. Presentation is typical when there is a progression of symmetric muscle weakness that is ascending from the lower extremities making its way more proximal and accompanied by absent or depressed tendon reflexes. This is typically due to an immune response to a previous infection that has cross-reactivity with peripheral nerve components such as the myelin or axon which subsequently results in demyelination.

Case report
A 55-year-old male presented to the ED complaining 3-week period of increasing upper extremity weakness that had progressed to his proximal lower extremities, with no significant past medical history. The patient described his weakness as spontaneous, numbing and aggravated by movement and initially beginning only in fingers and hands, with it progressing to his forearms bilaterally. The patient later expressed similar symptoms starting in his buttocks and progressing to his thighs bilaterally. The patient denied numbness, paresthesia, headache, fever, neck stiffness, visual changes, shortness of breath.

Keywords
Neurology, Guillain–Barré syndrome, paralysis, acute inflammatory demyelinating polyneuropathy

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breath, or recent infection. The patient was seen by his primary care physician 1 week prior to admission and received oral methylprednisolone tapering dose for 7 days.

He presented to the ED due to progression of his weakness. Past medical history was significant for Stage-III rectal cancer status post-colostomy, chemotherapy, and radiation, spherocytosis with splenomegaly. He denied history of tobacco, alcohol, or illicit drug use.

He was married and living with his wife and children.

Bilateral 2/5 weakness in the upper extremities and 3/5 weakness in the lower extremities. There was no cranial nerves or bulbar muscles involvement.

He had no dermatomal sensory loss with depressed deep tendon reflexes in upper and lower extremities.

Laboratory findings including complete blood count, electrolytes, thyroid-stimulating hormone—were within normal limits. Testing for HIV, ANA, West Nile, Syphilis, and Influenza A/B was also negative.

Magnetic resonance imaging (MRI) of entire neuro axis with and without contrast was unremarkable for any acute or chronic pathologies that may correlate with his symptoms.

Computerized tomography (CT) of abdomen and pelvis with contrast was negative for reoccurrence of the patient’s past cancer history.

CSF analysis, which was performed on the second day of presentation, showed protein level of 101 mg/dL and white blood cell (WBC) count of 7/mcL. Given the patient’s elevated WBC,7 possible differentials could be infections such as syphilis, toxoplasmosis, herpes simplex virus 1 (HSV-1), or even HIV.8 These were all tested and seen to be negative while patient was inpatient. Electromyography (EMG) showed evidence of acute polyneuropathy including fibrillation potentials and decreased recruitment in four extremities (Figure 1(a) and (b)). Nerve conduction studies (NCS) showed prominent conduction block with prolonged distal latencies and slowed conduction velocities in upper and lower extremities (Table 1). Findings were consistent with severe, acquired demyelinating polyneuropathy. This was further confirmed with the presence of antiganglioside antibodies, anti-GM1 IgG, which were also positive.

The patient received four doses of Intravenous immunoglobulin (IVIg) with gradual improvement over the course of the following weeks. The patient initially had minimal response. Following the administration of four doses of intravenous immunoglobulin, he did have marked improvement a week following the last dose. The patient was discharged and was admitted for acute rehabilitation with physiotherapy and occupational therapy.

**Discussion**

GBS is the most common cause of acute muscular paralysis, affecting 0.4–2.4/100,000 people annually. AIDP is the most common form of GBS. It commonly presents with ascending weakness and typical cerebrospinal fluid albumin cytologic dissociation. There are several case reports of

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**Figure 1.** (a) Left Ulnar motor NCS. (b) Left personal motor NCS. These figures show the presence of conduction block and temporal dispersion indicative of an acquired demyelinating polyneuropathy.

i: wrist, ii: Below Elbow, iii: Above Elbow, iv: Axilla, v: Ankle, vi: Below Knee, vii: Above Knee.
Miller–Fisher Syndrome (MFS) with Botulism Toxin Ingestion presenting as “Descending Paralysis.” However, those case report presentations involved bulbar muscles.

Unusual features of GBS include papilledema, facial myokymia, hearing loss, meningeal signs, vocal cord paralysis, and mental status changes. MFS variant of GBS, characterized by ophthalmoplegia, ataxia, and areflexia, occurs in approximately 10% of cases in the United States and Europe and 20% of cases in Asia.

The patient’s past medical history of GBS, paraneoplastic disease, chronic anemia, myeloproliferative or myelodysplastic process, and possible multiple myeloma were all possible differentials—with GBS being the most likely diagnosis. GBS was not immediately diagnosed due to the patient's presentation of ascending upper extremity and descending lower extremity and the need for additional clinical investigation. After extensive review of the literature, the authors believe that this to be the first case of GBS in this form.

The patient had an extensive imaging workup with MRI cervical and thoracic spine with and without contrast. Imaging showed diffusely abnormal marrow signaling throughout the cervical and thoracic spine concerning for severe anemia, myeloproliferative, or myelodysplastic process. The patient’s past medical history and laboratory findings of severe anemia with a hemoglobin of 10.6 g/dL would explain the changes seen.

Clinically, the patient was not experiencing any bone pain, fatigue, renal disease, hypercalcemia, or weight loss, which was in support against a possible paraneoplastic disease or multiple myeloma. Patient was also screened for JAK2 mutations that were negative. These screenings helped rule out myeloproliferative or myelodysplastic process. Due to the patient’s history of stage-III rectal cancer, repeat CT abdomen and pelvis was performed that was subsequently negative. MRI imaging of the C-spine and T-spine showed changes consistent with chronic anemia with evidence of stable diffusely abnormal marrow signal, hemoglobin was stable at 10–11 gm/dL. However, chronic anemia did not explain his ascending upper extremity and descending lower extremity because weakness with chronic anemia is generalized and is often accompanied by muscle pain, which our patient did not experience. NCS of the left upper and lower extremities showed prominent conduction block with temporal dispersion in all motor nerve tested. EMG of the left lower extremity showed markedly reduced recruitment in muscles test. CSF findings along with the improvement after administration of Intravenous immunoglobulin (IVIg) further supported the diagnosis of GBS. Typical findings on MRI of the spine often show thickening and enhancement of the intrathecal spinal roots and cauda equine. On MRI of the brain, in GBS, findings include enhancement of the oculomotor, abducens, and facial nerves may be seen. No acute findings were described on our patients MRI Brain.

**Conclusion**

Atypical presentations of AIDP, as seen in this case report, may delay diagnosis and treatment.

In order to prevent delay in treatment of patients with AIDP, it is important to be aware of the presence of atypical presentations and its variants. Given the multitude of variation in the possible presentation of GBS, our case did not meet the criteria of those variants.

**Declaration of conflicting interests**

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

**Ethical approval**

Our institution does not require ethical approval for reporting individual cases or case series.

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**Table 1. Motor nerve condition.**

| Nerve     | Side | Site            | Latency (ms) | Norm (less than) | Amplitude (µV) | Norm (greater than) | Distance (mm) | NCV (Nerve Conduction Velocity) (m/s) |
|-----------|------|-----------------|--------------|------------------|----------------|---------------------|---------------|----------------------------------------|
| Median    | Left | Palm            | 2.0          | 2.4              | 5200           | 3500                | *Palm*        | 80                                     |
|           |      | Wrist           | 6.2          | 4.2              | 4600           | 3500                | Palm-wrist    | 19                                     |
|           |      | Elbow           | 14.6         | 8.8              | 270            | 3500                | Wrist-Elbow   | 250                                    |
|           |      | Axilla          | 16.4         | 11.6             | 130            | 3500                | Elbow-Axilla  | 100                                    |
| Ulnar     | Left | Wrist           | 3.2          | 3.4              | 2900           | 3500                | **Wrist**     | 80                                     |
|           |      | Below elbow     | 7.8          | 7.5              | 280            | 3500                | Wrist-Below   | 245                                    |
|           |      | Above elbow     | 9.6          | 9.6              | 260            | 3500                | Below elbow   | 100                                    |
|           |      | Axilla          | 11.2         | 11.2             | 270            | 3500                | Above Axilla  | 105                                    |
| Peroneal  | Left | Ankle           | 6.2          | 5.5              | 3000           | 3500                | **Ankle**     | 85                                     |
|           |      | Below knee      | 18.3         | 12.0             | 280            | 3500                | Ankle-Below   | 375                                    |
|           |      | Above knee      | 20.7         | 14.9             | 240            | 3500                | Below knee    | 85                                     |

| Median Left Palm | NCV | 2.0 | 2.4 | 5200 | 3500 | *Palm* |
| Median Wrist     |     | 6.2 | 4.2 | 4600 | 3500 | Palm-wrist |
| Median Elbow     |     | 14.6| 8.8 | 270  | 3500 | Wrist-Elbow |
| Median Axilla    |     | 16.4| 11.6| 130  | 3500 | Elbow-Axilla |
| Ulnar Left Wrist |     | 3.2 | 3.4 | 2900 | 3500 | **Wrist** |
| Ulnar Below elbow|     | 7.8 | 7.5 | 280  | 3500 | Wrist-Below |
| Ulnar Above elbow|     | 9.6 | 9.6 | 260  | 3500 | Below elbow |
| Ulnar Axilla     |     | 11.2| 11.2| 270  | 3500 | Above Axilla |
| Peroneal Left Ankle| | 6.2 | 5.5 | 3000 | 3500 | **Ankle** |
| Peroneal Below knee| | 18.3| 12.0| 280  | 3500 | Ankle-Below |
| Peroneal Above knee| | 20.7| 14.9| 240  | 3500 | Below knee |

*Palm: 80, Wrist: 19, Palm-wrist: 80, Wrist-Below: 245, Below elbow: 100, Above Axilla: 105.
Informed consent
Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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