Case Report

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Acute Ascending Paralysis: Time Limited Diagnostic Perplexity

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Abstract

Ascending paralysis is a rare neurological manifestation characterized by progressive weakness that starts in the lower extremities and ascends towards the upper body. Guillain-Barré Syndrome (GBS) is a common cause of ascending paralysis and is typically preceded by an infection. We present two case reports, who presented with ascending paralysis, which was initially suspected to be GBS. However, further evaluation revealed an underlying neuropathy contributing to the clinical presentation with a vasculitic etiology.

This case highlights the importance of considering alternative etiologies when evaluating patients with ascending paralysis, as over lapping clinical features between GBS and neuropathies can present diagnostic challenges. Early recognition and prompt initiation of appropriate treatment are crucial for favorable patient outcomes.

Keywords: Ascending paralysis; EGPA; Guillan Barré Syndrome; Peripheral Neuropathy; Eosinophilia; Asthma; Neuropathy.

INTRODUCTION

Ascending paralysis can also be associated with certain types of vasculitis, characterized by inflammation of blood vessels. It can affect the blood vessels supplying the nerves, leading to nerve damage and subsequent muscle weakness or paralysis.

In vasculitic neuropathy, with the involvement of the peripheral nerves in vasculitis, ascending paralysis may occur as a result of nerve damage caused by inflammation and impaired blood flow. This can be seen in various forms of vasculitis, including but not limited to:

1. *Polyarteritis nodosa:* A systemic necrotizing vasculitis that primarily affects small and medium sized arteries.
2. *Churg Strauss syndrome (eosinophilic granulomatosis with polyangiitis):* This rare form of vasculitis affects small to medium sized blood vessels, including those supplying the peripheral nerves.

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3. *Microscopic polyangiitis:* A systemic necrotizing vasculitis that primarily affects small blood vessels.

In vasculitis associated ascending paralysis, the specific mechanisms underlying nerve damage are complex and can involve immune mediated inflammation, ischemia (lack of blood supply), or a combination of both. The symptoms typically start in the lower extremities and progress upward, affecting the muscles and nerves at higher levels of the body.

Diagnosis of vasculitis associated ascending paralysis involves a thorough medical evaluation, including a detailed clinical history, physical examination, and various diagnostic tests such as blood tests, nerve conduction studies, electromyography, and imaging studies. Treatment options may include immune suppressive medications, such as corticosteroids or other immunosuppressants, to control the inflammation and halt the progression of nerve damage.

It is important to note that vasculitis associated ascending paralysis is relatively rare and may present differently in each individual. Prompt recognition and early treatment are crucial for managing the underlying vasculitis and minimizing potential long-term complications. While both AIDP and vasculitic neuropathy can present with ascending paralysis, the underlying mechanisms and treatments differ. AIP is an autoimmune condition triggered by infections, whereas vasculitic neuropathy involves inflammation of blood vessels supplying the nerves.

Here we see two unique presentations of ascending paralysis which posed a diagnostic dilemma.

CASE 1

A 63-year-old married male, who is a retired agriculturalist, has a medical history of adult onset asthma. He has been experiencing bilateral lower limb weakness for the past 30 days, which has been progressively worsening. The weakness starts from the distal muscles and spreads proximally, predominantly affecting the left side. The patient also reports associated paraesthesias. Additionally, he has been experiencing bilateral upper limb pain and weakness for the past 15 days, which has rapidly progressed in a distal to proximal pattern, again more pronounced on the left side. Paraesthesias are also present in the upper limbs. There are no girdle like sensations, bladder involvement, haematuria, or rashes. The patient also complains of a cough with breathlessness for the past 7 days, with mucoid expectoration. Exertional breathlessness and palpitations are present, accompanied by audible wheezes. In the past, the patient has had frequent admissions for recurrent asthma exacerbations with motor weakness over the past 2 years. However, there was improvement in respiratory and motor symptoms after receiving IV steroids at the local hospital during his admissions for exacerbations of breathlessness. The patient received the Covishield vaccination 3 months prior to the current admission.

On general examination, the patient's pulse was found to be elevated at 120 bpm. Blood pressure measurements in the supine position showed readings of 150/90 mmHg, while standing measurements displayed 130/90 mmHg. The jugular venous pressure (JVP) was elevated, and bilateral pitting pedal edema was observed. Moving on to the central nervous system (CNS) findings, wasting was noted in both the upper and lower limbs. Hypotonia was detected in all four limbs. In terms of muscle power, the patient exhibited grade 3/5 strength in the proximal muscles of the upper limbs and grade 1/5 strength in the distal muscles of the upper limbs. For the lower limbs, there was grade 3/5 strength in the proximal muscles and complete loss of strength (grade 0/5) in the distal muscles. Deep tendon reflexes (DTRs) were absent, but the abdominal reflex was present. The bilateral plantar reflex was mute. Sensory examination revealed pain and temperature sensation present in the upper limbs, with an early decay of vibration sense (left side greater than right) up to the shoulder joint. In the lower limbs, pain and temperature sensation were present, but there was a loss of vibration sense up to the knee joint and an early decay of vibration sense up to the hip joint. No cerebellar signs were noted. Moving to the respiratory system, bilateral lintensity of breath sounds were found to be increased in intensity. Extensive rhonchi and extensive fine inspiratory crepitations were heard in bilateral lung fields.

CASE 2

A 32-year-old married female, who is a home maker, presents with several chief complaints. She has been experiencing bilateral lower limb weakness for the past 30 days, with a progressive pattern from distal to proximal and a left-to-right distribution. The weakness is accompanied by paraesthesias, Additionally, the patient reports a painful skin lesion on her left foot for the past 20 days. The

lesion is tender, accompanied with blistering. She also complains of bilateral upper limb pain and weakness, which has rapidly progressed in a distal to proximal pattern, again more pronounced on the left side. Paraesthesias are present in the upper limbs as well. There are no girdle like sensations, bladder involvement, haematuria, or rashes. In terms of past medical history, the patient was diagnosed with possible asthma two years ago and has been using a metered-dose inhaler (MDI) and antihistamines for management. She underwent functional endoscopic sinus surgery (FESS) one year ago for nasal polypoidosis. The patient is in a postpartum status since nine months ago. Furthermore, she received the Covishield vaccination three months prior to the present presentation.

On general examination, the patient's pulse was found to be 112 beats per minute. No orthostatic hypotension. A skin lesion was observed on the left lower limb, measuring approximately 1x1.5 cm. It appeared as maculopapular lesions with healing blisters, suggesting a healing process. Central nervous system (CNS) findings, there was no wasting noted in the limbs. However, hypotonia, was present in all four limbs. Assessing muscle power, the patient exhibited grade 4/5 strength in the proximal muscles of the upper limbs and grade 3/5 strength in the distal muscles of the upper limbs, indicating some weakness. In the lower limbs, there was grade 4/5 strength in the proximal muscles and grade 3/5 strength in the distal muscles, Deep tendon reflexes (DTRs) were absent, while the abdominal reflex was present, suggesting intact lower spinal cord function. The bilateral plantar reflex showed flexor responses. Sensory examination revealed pain and temperature sensation present in the upper limbs, suggesting intact sensory pathways, but an early decay of vibration sense (greater on the left side) up to the shoulder joint. In the lower limbs, pain and temperature sensation were present, but there was a loss of vibration sense up to the knee joint and an early decay of vibration sense up to the hip joint, No cerebellar signs, such as coordination or balance issues, were noted. Shifting to the respiratory system, bilateral breath sounds were found to be normal and vesicular in nature, However, extensive rhonchi, which are low-pitched wheezing sounds, and occasional fine inspiratory crepitations, which are crackling sounds, were heard in bilateral lung fields.

Table 1: Lab Values of the Case

Investigations: Case 1	Investigations: Case 2
CBC HB 12.8 , TC: 41400, DC: 32/05/63 Platelets: 3,20,000	HB 11.5, TC: 28100, DC:53/06/41 Platelets: 4,21,000
HRCT Thorax Patchy ground glass opacities and mild bronchiolitis	Patchy ground glass opacities and bronchiectesis
MRI Whole Spine, CSF Analysis: Essentially normal	
Bone Marrow Aspiration Myeloid Hyperplasia with eosinophilia and its precursors	Hyperplastic marrow with erythroid hyperplasia and increase in eosinophilic precursors
ANCA: p ANCA strong positive	
Nerve Conduction Study Severe sensory motor peripheral neuropathy: Axonal type in both upper and lower limbs	
Sural Nerve Biopsy Chronic moderately severe axonal neuropathy with vascular changes; compatible with vasculitic neuropathy (small and medium vessels)	
2D Echo, RFT, LFT, Vit B12: WNL	

DISCUSSION

The diagnosis for both patients has been confirmed as Eosinophilic granulomatosis with polyangiitis (EGPA) presenting as peripheral neuropathy. They were both treated with pulse methylprednisolone therapy, followed by a course of immunomodulators and corticosteroids. The dosage of these medications was adjusted according to their weight. As a result of the treatment, both patients experienced a rapid improvement in their motor deficits.

CONCLUSION

Ascending paralysis in an acute setting might pose a clinical diagnostic challenge as the reliability on the history and precision on physical examination may provide an insight into the etiology of the disease added with the relative delay in immediate availability of specialized tests such as nerve conduction studies and neuro imaging. Acute inflammatory polyradiculoneuropathy may pose an imminent danger to the patients life with rapid progression of involvement to phrenic nerve and respiratory muscles, as compared to vasculitic neuropathy, which may present similarly but with slower progression and with no acute life threating complications.

Peripheral neuropathy is quite common in patients with EGPA.[1]

Mononeuritis multiplex is slightly more common than symmetric polyneuropathy, and the lower limbs are predominately affected.[2] Peripheral neuropathy alone is rarely life threatening as compared to renal and lung involvement but does significantly affect quality of life.[2] Less than 40% of patients with EGPA have positive ANCAs, this subgroup of patients presents most often with neuropathy. ANCA positivity has also been associated with lower mortality.[3] Both the asthmatic patients presenting with symmetric ascending paralysis and hyporeflexia, severe eosinophilia, and skin purpuric lesion which raised our suspicion of EGPA. EGPA and other vasculitis should always be part of the differential diagnosis of GBS, as the first line treatments may differ; while steroids are of no use (may even be harmful) in GBS, they are the mainstay of treatment in EGPA.[4]

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