

## Case Report

# Guillain-Barré Syndrome in a Patient with Sickle Cell Anemia

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## Abstract

### Description

A 24-year-old African American male with a history of sickle cell anemia (Hb S/S) presented to an outside hospital with acute colitis, acute renal failure and sickle cell crisis and was treated with supportive measures. On day 3 of hospitalization, he developed bilateral ascending paralysis with sacral numbness. Magnetic resonance imaging (MRI) demonstrated epidural lipomatosis, which was attributed as the cause of his paralysis. He was transferred to our facility for neurosurgery evaluation. Based on the physical examination, Guillain-Barré Syndrome (GBS) was suspected. This conclusion led to a lumbar puncture with cerebrospinal fluid (CSF) analysis that confirmed the diagnosis. He was then treated with intravenous immunoglobulin (IVIg), which resolved his symptoms.

We present this case to highlight the importance of a physical exam rather than relying heavily on imaging studies. Physical exam findings lead to a diagnosis, which was then confirmed with appropriate testing.

### Keywords

sickle cell anemia; Guillain-Barré syndrome; autoimmune diseases of the nervous system; ascending paralysis; acute autoimmune neuropathy; physical examination; male

## Introduction

Sickle cell anemia (SCA) can have a myriad of central and peripheral nervous system manifestations. In patients with recurrent admissions for painful crises and neuropathic pain, potentially life-threatening diagnoses may be inadvertently overlooked. Guillain-Barré syndrome (GBS), an acute immune-mediated polyneuropathy, is believed to result from molecular mimicry as an immune response to a preceding infection that cross-reacts with peripheral nerves.<sup>1,2</sup> Symmetrical ascending paralysis and absent deep tendon reflexes are pathognomonic features of GBS.<sup>2,3</sup>

## Case Presentation

A 24-year-old African American man with SCA status post splenectomy, chronic back pain and hypertension was admitted for intractable nausea, vomiting and diarrhea for approximately 1 week. On admission at an outside hospital,

he was diagnosed with acute colitis, sickle cell crisis and acute renal failure with a serum creatinine of 2.7. He was treated with intravenous hydration, opiates, antibiotics (clindamycin and levofloxacin) and anti-emetics. On day 3 of his admission, he experienced bilateral lower extremity weakness and pain in the buttocks that radiated to the soles of the feet with associated numbness. He reported an inability to stand as well as urinary and fecal incontinence. As he went to sleep feeling well, he was unable to specify if the weakness was ascending versus descending or symmetrical versus asymmetrical. A physical examination revealed absent bilateral Achilles reflexes, decreased sensation and mild weakness. An MRI of the lumbosacral spine (**Figure 1**) did not reveal any cord compromise but did demonstrate epidural lipomatosis likely related to his history of sickle cell disease. He was transferred to our facility for a neurosurgery evaluation and escalation of care.



**Figure 1.** MRI of the lumbosacral spine showed epidural lipomatosis and spinal canal stenosis, most notable at L4-5 and L5-S1 with disc bulging and no evidence of spinal cord compromise (orange arrows).

On arrival to our hospital, his vital signs were within the normal range. Upon a physical exam, he had absent Achilles and patellar reflexes, absent Babinski reflex, decreased sensation and proprioception in his lower extremities—4/5 strength on ankle dorsiflexion bilaterally and 3+/5 strength on hip flexion/extension and knee extension. The remainder of the exam was unremarkable. His complete blood count was notable for neutrophilic dominant leukocytosis, microcytic anemia and thrombocytosis. A comprehensive metabolic profile revealed an elevated serum creatinine, total bilirubin and lactate dehydrogenase. GBS was suspected. A lumbar puncture was performed, and CSF studies revealed cytoalbuminologic dissociation (**Table 1**), reinforcing our diagnosis. He was treated with a 5-day course of IVIg 400 mg/kg/day for 5 days, which resulted in the resolution of his symptoms. It was noted that he had a restored strength of 4/5 in bilateral proximal and distal lower extremities. He was discharged with home health services for continued rehabilitation and close outpatient neurology follow-up.

## Discussion

Clinically, sickle cell disease has been associated with vaso-occlusive crises, increased susceptibility to certain infections and hemolysis.

Repeated vaso-occlusive crises lead to multiple complications, including chronic pain, ischemic or hemorrhagic strokes, acute chest syndrome, splenic infarction and renal infarction.<sup>4</sup> Neurological complications include chronic headaches, epilepsy and cognitive impairment secondary to anemia, hypoxia and silent infarcts.<sup>5</sup>

Historically, GBS was considered a disorder, but it is now consistent with multiple variants, including acute inflammatory demyelinating polyneuropathy, acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (ASMAN).<sup>6</sup> Acute inflammatory demyelinating polyneuropathy (AIDP) is the most common form of GBS in the developed world, accounting for about 85% of cases.<sup>7,8</sup> The majority of patients develop neurologic symptoms 2 to 4 weeks after a respiratory or gastrointestinal infection.<sup>6,9</sup> Diagnostic criteria for GBS includes progressive weakness in at least 2 extremities, areflexia and symptoms progressing for less than 4 weeks. Patients may also complain of sensory deficits and, in the acute phase, patients may report pain due to nerve root inflammation.<sup>3,10,11</sup>

Our patient's presentation was initially thought to be related to acute sickle cell crisis and lipomatosis, causing cord compromise. Anchoring bias further affected clinical decision making.

**Table 1.** CSF analysis consistent with cytoalbuminologic dissociation.

Color	Yellow
Appearance	Clear
Xanthochromia	Present
WBC	12 leukocytes/mm <sup>3</sup>
Polymorphonuclear cells	4%
Lymphocytes	86%
Monocytes/macrophages	9%
Eosinophils	1%
RBC	68 erythrocytes/mm <sup>3</sup>
CSF Total Protein	330 mg/dL

A thorough physical exam and its discordance with the imaging studies raised concern about other possible etiologies. GBS was suspected, given the patient’s antecedent history of gastroenteritis, more so than other forms of acute inflammatory demyelinating polyradiculoneuropathy. A lumbar puncture was performed, and CSF studies revealed a cytoalbuminologic dissociation indicative of GBS. The patient has not returned to our hospital with similar symptoms.

We present this case to highlight the importance of maintaining a broad differential and low threshold to consider alternative diagnoses when the patient’s clinical symptoms do not correlate with the initial diagnosis. The final diagnosis of GBS was delayed given our bias since we clung to an acute sickle cell crisis and epidural lipomatosis as the etiology for this patient’s symptoms. Anchoring bias lead to a postponement in diagnosis and initiation of appropriate treatment. Further delay could have led to devastating results.

## Conclusion

GBS should be considered in an appropriate clinical setting, particularly in patients who report an infection preceding the onset of neurologic symptoms, such as paralysis and paresthesia. Prompt recognition and management is crucial to avoid a catastrophic outcome. Detailed bedside exams should be an integral part of everyday practice.

## Conflicts of Interest

The authors declare they have no conflicts of interest.

Drs. D. Patel, P. Patel and K. Udani are employees of Grand Strand Medical Center, a hospital affiliated with the journal’s publisher.

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