

Introduction

Diagnosis of Guillain-Barré Syndrome (GBS) is dependent upon high degree of clinical suspicion. Late recognition can delay treatment, negatively impacting recovery and prognosis. We discuss a case of unilateral weakness, urinary incontinence, and normoreflexia with non-diagnostic initial work-up.

Findings suggestive of GBS on electromyography (EMG) eventually prompted initiation of proper treatment. This case highlights the importance of clinical suspicion despite atypical presentation

Case Presentation

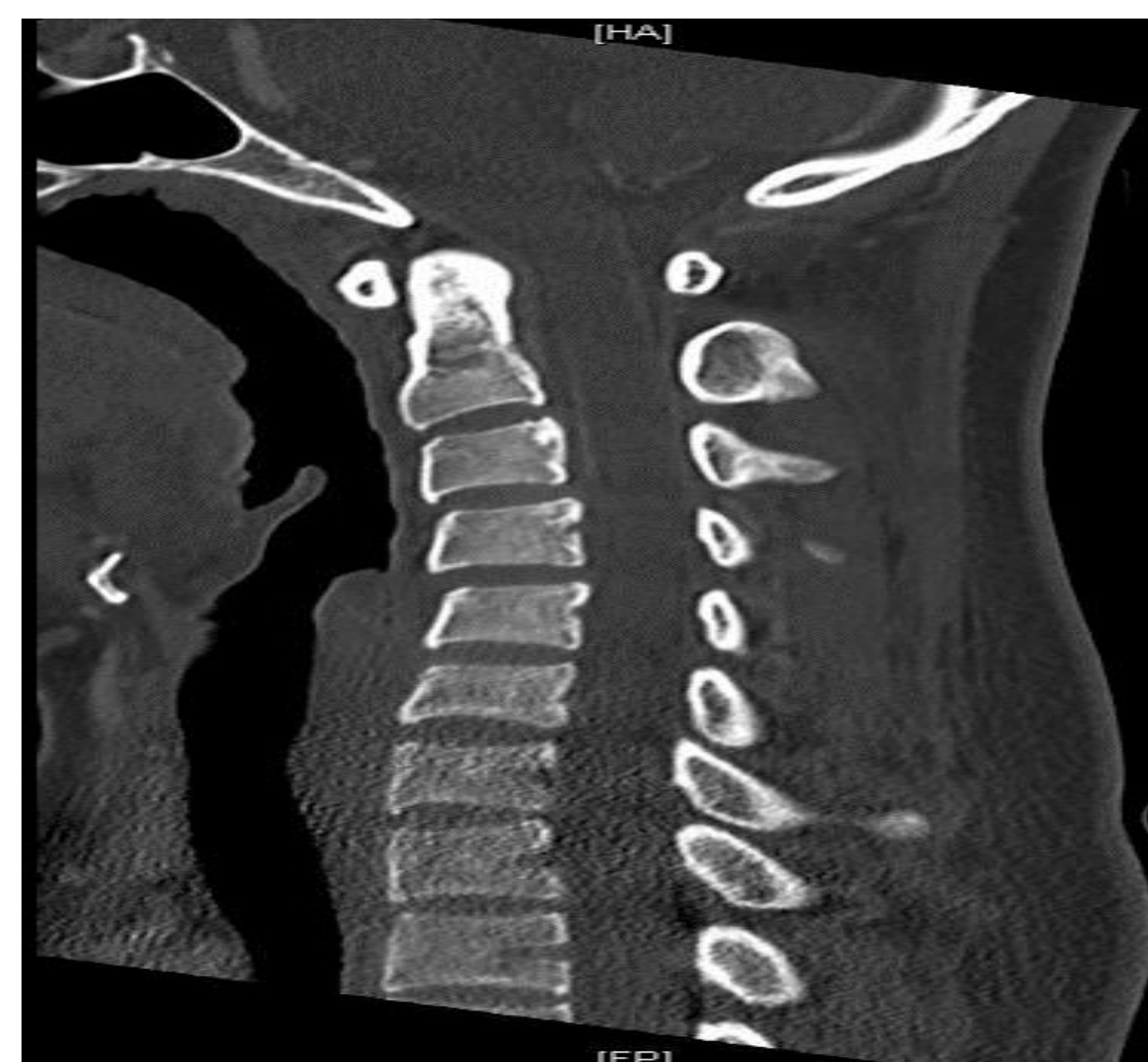
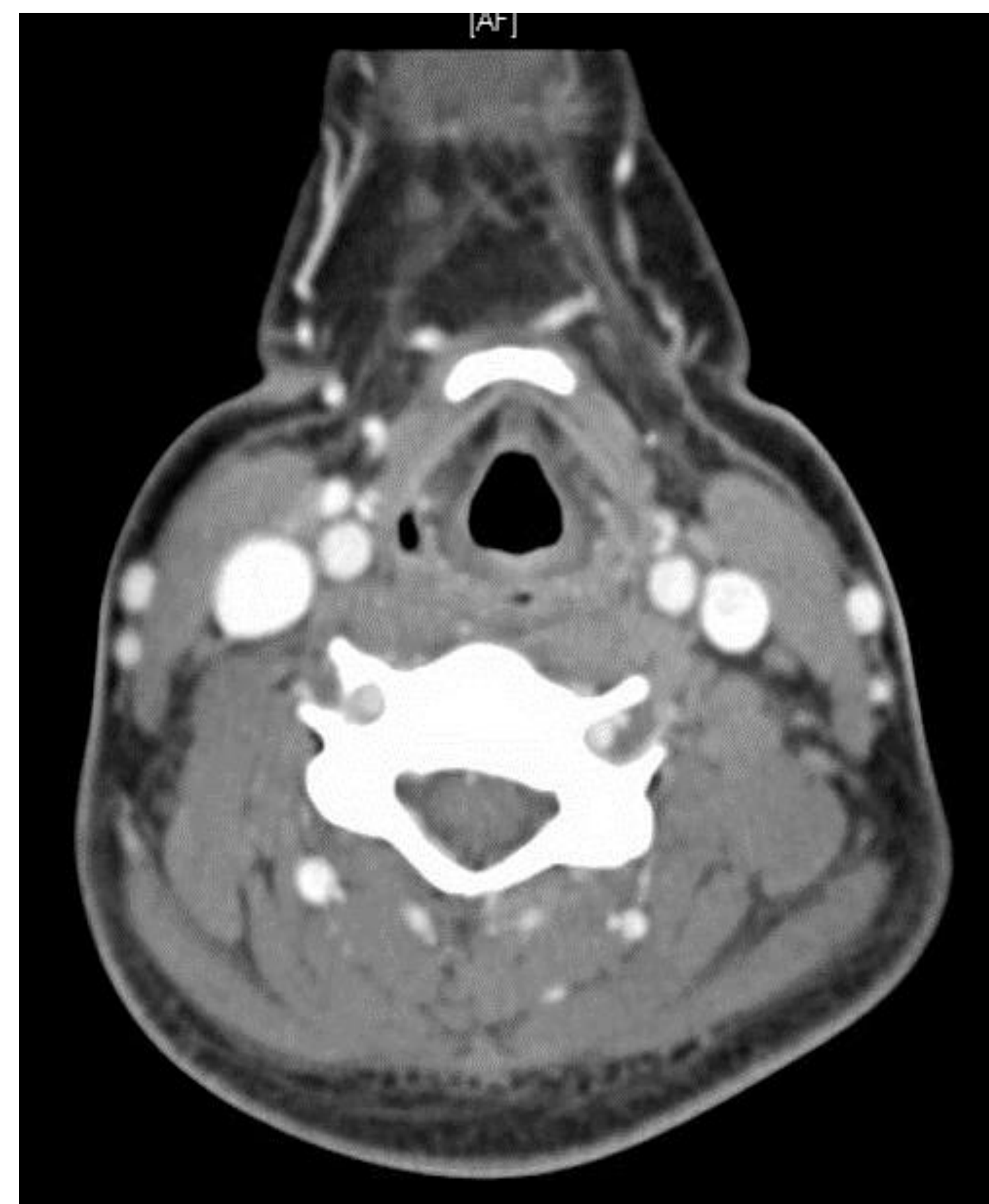
Our patient is a 19-year-old female presenting with left lower extremity weakness, associated tingling, and decreased sensation for four days.

Physical exam showed decreased proximal left lower extremity motor strength and sensation with distal tendon reflexes (DTRs) intact. She reported inability to urinate, requiring catheterization.

Initial work-up and imaging were unremarkable, including MRI of the brain, cervical and thoracic spine. Lumbar puncture was performed with normal cerebrospinal fluid (CSF) analysis.

Intravenous steroids were given for two days with subjective improvement of symptoms, after which the patient left against medical advice. However, she returned one week later with worsened left lower extremity motor strength, left foot drop, and absent DTRs. EMG was performed demonstrating severe peripheral neuropathy and clinical diagnosis of GBS was made. Confirmatory lumbar puncture was deferred and intravenous immunoglobulin (IVIg) administration resulted in complete symptomatic resolution.

Imaging



Electrodiagnostic Studies

Motor Summary Table

Site	NR	Onset (ms)	Norm Onset (ms)	O-P Amp (mV)	Norm O-P Amp	Site1	Site2	Delta-0 (ms)	Dist (cm)	Vel (m/s)	Norm Vel (m/s)
Left Peroneal Motor (Ext Dig Brev)											
Ankle	NR	<6.0	<6.0	>2.5	>2.5	B Fib	Ankle		0.0		>42
B Fib	NR										
Right Peroneal Motor (Ext Dig Brev)											
Ankle	NR	<6.0	<6.0	>2.5	>2.5	B Fib	Ankle		0.0		>42
B Fib	NR										
Left Tibial Motor (Abd Hall Brev)											
Ankle		6.4	<6.0	2.6	>5.0	Poplit	Ankle	12.4	42.0	33.9	>40
Poplit		18.8		1.7							
Right Tibial Motor (Abd Hall Brev)											
Ankle		7.0	<6.0	4.5	>5.0	Poplit	Ankle	9.6	42.0	43.7	>40
Poplit		16.6		1.2							

NR	F-Lat (ms)	Lat Norm (ms)	Contra F-Lat (ms)	L-R F-Lat (ms)	L-R Lat Norm	M-Lat (ms)	F-Lat-MLat (ms)
Left Peroneal (Mrkrs) (EDB)							
NR	<60	<60			<4		
Right Peroneal (Mrkrs) (EDB)							
NR	<60	<60			<4		
Left Tibial (Mrkrs) (Abd Hallucis)							
	59.75	<61	59.96	0.21	<4	7.35	52.40
Right Tibial (Mrkrs) (Abd Hallucis)							
	59.96	<61	59.75	0.21	<4	7.35	52.61

EMG

Side	Muscle	Nerve	Root	Ins Act	Fibs	Psw	Amp	Dur	Poly	Reert	Int Pat	Comment
Right	VastusMed	Femoral	L2-4	Nml	0	0	Nml	Nml	Nml	Nml	Complete	
Right	AntTibialis	Dp Br Peron	L4-5	Nml	0	1+	Nml	Nml	Nml	Nml	Complete	
Right	Peroneus Long	Sup Br Peron	L5-S1	Nml	0	0	Nml	Nml	Nml	Nml	Complete	
Right	MedGastroc	Tibial	S1-2	Nml	0	0	Nml	Nml	Nml	Nml	Complete	

FINDINGS:

Evaluation of the Left Peroneal Motor and the Right Peroneal Motor nerves showed no response (Ankle) and no response (B Fib). The Left Tibial Motor nerve showed prolonged distal onset latency (6.4 ms), reduced amplitude (2.6 mV), and decreased conduction velocity (Poplit-Ankle, 33.9 m/s). The Right Tibial Motor nerve showed prolonged distal onset latency (7.0 ms) and reduced amplitude (4.5 mV).

Evaluation of the Left Peroneal F Wave showed no response. The Right Peroneal F Wave showed no response. All remaining F Wave latencies were within normal limits. All F Wave left vs. right side latency differences were within normal limits.

EMG needle evaluation of the Right AntTibialis and the Left AntTibialis showed slightly increased spontaneous activity. All remaining muscles (as indicated in the EMG scoring table) showed no evidence of electrical instability.

IMPRESSIONS:

The above electrodiagnostic study reveals evidence of severe peripheral neuropathy, suggested an acute Guillain Barre syndrome.

Discussion

GBS has an incidence of 1 to 2 cases per 100,000 annually, with typical symptoms of progressive symmetrical muscle weakness accompanied by depressed or absent DTRs in 90% of patients, however reflexes may be intact early in presentation [1, 2]. Paresthesias may occur with mild sensory symptoms. Dysautonomia occurs in 70% of patients, presenting as tachycardia, incontinence, blood pressure dysregulation, arrhythmia, and ileus [3]. Posterior reversible leukoencephalopathy syndrome has been reported in patients with dysautonomic hypertension [5]. Lumbar puncture normally shows elevated CSF protein and normal white blood cell count (i.e. albuminocytologic dissociation) in more than 60% of patients one week from onset of symptoms [1]. EMG studies can show demyelination or an axonal picture, further classifying variants of GBS. Nerve conduction studies in GBS may not become positive until two weeks after onset of symptoms [4]. MRI may show enhancement of intrathecal spinal nerve roots and cauda equina. Treatment includes supportive care, IVIG, and plasmapheresis. Patients may require closer monitoring in an intensive care unit for increasing weakness, respiratory depression, and dysautonomia.

Although rare, atypical presentation of GBS can hinder diagnosis. Typical features of symmetrical weakness, absent DTRs, and albuminocytologic dissociation may be absent, particularly early in disease course. Additional features such as dysautonomia and parasthesia may be present. Electrodiagnostic studies and repeat CSF analysis may be useful if diagnosis is unclear. Diagnosis should not be excluded based on atypical findings, and clinical suspicion should remain to prevent progression of a potentially fatal disease.

References

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5. Elahi A, Kelkar P, St Louis EK. Posterior reversible encephalopathy syndrome as the initial manifestation of Guillain-Barré Syndrome. Neurocrit Care 2004; 1:465.