Guillain-Barre Syndrome During Pregnancy: A Rare Presentation of the Axonal Variant

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CASE DESCRIPTION

A 30-year-old G6 P5 female with no significant past medical history, currently pregnant at 6 months, who presented to the hospital with a three-day history of ascending paralysis. She was in her usual state of health several days prior when she complained of upper respiratory symptoms. The following day she had difficulty walking and experienced a heavy feeling of her hands.

Review of systems were negative for sensory impairment as well as bowel or bladder incontinence. She denied any alcohol, tobacco, illicit drug use, or known chemical exposure. Her last flu vaccination was several years ago. She also reported no recent travel outside of the state. Due to a lack of insurance, however, she reported she had no established prenatal care visits during this pregnancy but was taking "prenatal care pills" that were recommended to her from the prior pregnancies.

No gross atrophy was noted on physical examination. Muscle strength testing of the right upper extremity revealed 2/5 shoulder abduction and 3/5 distally. The left upper extremity was slightly weaker with a 1/5 shoulder abduction and 2/5 distally. Lower extremity muscle strength testing was a 1/5 throughout bilaterally. Sensation to light touch, pain, and temperature were grossly intact bilaterally. Deep tendon reflexes were 1+ in the bilateral upper extremities and absent at the patellar and achilles. Babinski and Hoffman reflexes were absent and no clonus was detected.

Metabolic etiologies were initially ruled out with routine laboratory testing which revealed a normal B12, folate, and thyroid. Infectious workup was negative.

Motor Nerve Study

Left Median Nerve						
Rec Site: APB	Lat (ms)	Dur (ms)	Amp (mV)	Area (mVms)	Dist (mm)	C.V. (m/s)
STIM SITE						
Wrist	4.3	5.2	0.700	2.4	0	
Elbow	8.1	5.8	0.517	2.1	180	48.0

Left Ulnar Nerve						
Rec Site: ADM	Lat (ms)	Dur (ms)	Amp (mV)	Area (mVms)	Dist (mm)	C.V. (m/s)
STIM SITE						
Wrist	4.3	5.3	0.133	0.4	0	
B.Elbow	7.2	4.8	0.117	0.3	140	49.4
A.Elbow	9.4	3.9	0.043	0.1	100	44.4

Peroneal/Fib Nerve										
Rec Site: EDB	Lat (ms)		Dur (ms)		Amp (mV)		Area (mVms)		Dist (mm)	C.V. (m/s)
STIM SITE	L	R	L	R	L	R	L	R	L R	L R
Ankle	NR	NR							0 0	
Fib.Head	NR	NR							0 0	
Pop.Fos.	NR	NR							0 0	

Tibial Nerve										
Rec Site: AH	Lat (ms)		Dur (ms)		Amp (mV)		Area (mVms)		Dist (mm)	C.V. (m/s)
STIM SITE	L	R	L	R	L	R	L	R	L R	L R
Ankle	NR	6.2		4.6		0.225		0.1	0 0	
Pop.Fos.	NR	13.2		4.6		0.042		0.1	0 300	42.9

Sensory Nerve Study

Left Sural Nerve				
Rec Site: Ankle	Lat (ms)	Pk Lat (ms)	Amp (uV)	Dist (mm)	C.V. (m/s)
STIM SITE					
mid calf	1.8	2.4	53.7	0	

Left Med/Uln/Rad Nerve					
Stim Site: Wrist	Lat (ms)	Pk Lat (ms)	Amp (uV)	Dist (mm)	C.V. (m/s)
REC SITE					
R Thumb	1.8	2.2	17.7	0	
M Thumb	1.9	2.4	64.7	0	
Index	2.1	2.8	40.7	0	
5th dig	2.0	2.6	36.0	0	

EMG Study

Name	Ins Act	Fibs	PSW	Fascics	Polyph	MU Amp	MU Dur	Config	Pattern	Recruit
L. Deltoid	norm	none	none	none	none	norm	norm	norm		dec
L.	norm	none	none	none	none	norm	norm	norm		dec
L. Biceps Brac	norm	none	none	none	none	norm	norm	norm		dec
L. Triceps	norm	none	none	none	none	norm	norm	norm		dec
L. Dors.Int.1	norm	none	none	none	none	norm	norm	norm		dec
L. Pronator Te	norm	none	none	none	none	norm	norm	norm		dec
L. Rectus Fem.	norm	none	none	none	none	norm	norm	norm		dec
L. Biceps Ln.H	norm	none	none	none	none	norm	norm	norm		dec
L. Tibialis An	norm	none	none	none						
L. Peroneus Ln	norm	none	none	none						
L. Gastroc.Med	norm	none	none	none						

RESULTS

Due to the patient's pregnancy, computerized topography had to be avoided due to the added risk of radiation. Neuroimaging of the brain was also unremarkable for any acute intracranial pathology. MRI of the spine was negative for any cord lesion or cord signal alteration which would be suggestive of a possible neoplasm or transverse myelitis. Due to the patient's lack of prenatal care, a transabdominal ultrasound was performed which showed appropriate gestational size and biophysical profile. Imaging of the brain and spine were unrevealing. Lumbar puncture showed no significant albuminocytologic dissociation. Due to inconclusive results, electrodiagnostics was ordered and showed evidence of an axonal variant of Guillain-Barre syndrome (GBS).

Patient underwent two rounds of intravenous immunoglobulin with improvement in her upper and lower extremity strength. Following intensive inpatient therapy, she was discharged as contact guard for functional transfers and gait.

Findings of NCS/EMG: Normal left sural, radial, ulnar, and median sensory responses. Severely reduced amplitudes in the left median, ulnar, and right tibial responses with normal velocities and absent F waves. No responses to the Left tibial and bilateral peroneal motor stimulation. EMG of the left upper and lower extremities revealed severely decreased motor recruitment with no motor units seen firing below the knee.

DISCUSSION

GBS is rare in pregnancy with an estimated incidence between 1.2 and 1.9 cases per 100,000 annually and it carries a high maternal risk. Several variants of GBS exist depending on the predominant nerve fiber involved (motor versus sensory) and type of injury (demyelinating versus axonal). Axonal variants of GBS include acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN). Unlike the classical presentation of GBS, the acute motor axonal neuropathy (AMAN) variant, that our patient incurred, typically presents with rapidly progressing ascending weakness with a prolonged period of paralysis and respiratory failure. Electrodiagnostic testing provide a useful tool in differentiating GBS subtypes. However, results may be equivocal in the early phases so serial testing is necessary. Antibody identification is also helpful in the classification of axonal GBS.

CONCLUSION

AMAN is an uncommon variant of GBS. This case highlights the importance of early detection of AMAN due to the rapid progression and severity of its symptoms, especially in pregnancy. Electrodiagnostic testing provides valuable information distinguishing AMAN from other acute pathology. With early identification, our patient was able to receive timely treatment and avoid a worsening outcome.

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