Austin Journal of Neurological Disorders & Epilepsy

Open Access

Austin Publishing Group

Case Report

Pharyngeal-Cervical-Brachial Variant of Guillain–Barré Syndrome in Pregnancy

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Received: April 24, 2017; **Accepted:** May 19, 2017; **Published:** May 26, 2017

Abstract

Pharyngeal-cervical-brachial (PCB) variant is a rare subtype of Guillain Barre's Syndrome (GBS) with typically rapidly progressive oropharyngeal and cervicobrachial muscle weakness and areflexia in the upper extremities. 31-year-old female patient at the 24th week of pregnancy was admitted to our clinic with a one week old rapid onset weakness in the right hand, difficulty in speech and swallowing. She had bilateral facial and upper extremities weakness and her pharyngeal reflex was decreased. In the EMG performed bilateral upper extremities, local motor axonal polyneuropathy was observed in the motor fibers. CSF protein level was found to be 89mg/dl. No cell was seen. In a study done on 250GBS patients, the incidence of PCB variant was 3%. PCB variant of GBS has never been reported during pregnancy previously. It should be kept in mind in patients with oropharyngeal and cervicobrachial weakness for utilization of early prompt treatment.

Keywords: Guillain Barre Syndrome; Pregnancy

Introduction

Guillain Barre's Syndrome (GBS) is an inflammatory demyelinising polyradiculoneuropathy with acute onset, rapidly progressive muscle weakness and paresthesias [1]. Its incidence during pregnancy is 6-24/100,000 [2]. Its incidence is 13% in the first trimester, 47% in the second trimester and 40% in the third trimester [3]. GBS encompasses a heterogenous group of diseases with different subtypes and clinical variants [4]. Pharyngeal-cervical-brachial (PCB) variant is a rare subtype of GBS with typically rapidly progressive oropharyngeal and cervicobrachial muscle weakness and areflexia in the upper extremities [5]. Weakness in the lower extremities is not commonly seen and when present, it is quite mild [4,6]. PCB variant is defined as a localized axonal form of GBS and a part of Fisher Syndrome (FS) [7]. PCB variant of GBS has never been reported during pregnancy previously.

Case Presentation

31-year-old female patient at the 24th week of pregnancy was admitted to our clinic with a one week old rapid onset weakness in the right hand, difficulty in speech and swallowing. The patient was conscious, cooperative and oriented. Her speech was hypophonic and had mild dysarthria. Her pupillae were isocoric, IR +/+ and her ocular movements were within normal limits. She had bilateral facial weakness. Palatal arcs showed little elevation bilaterally and her pharyngeal reflex was decreased. Bilateral biceps, triceps and styloradial deep tendon reflexes (DTR) were decreased upper extremities while her patella and achilles DTR were bilaterally normal in the lower extremities. Muscle strength in the upper extremities was more apparent in the right extremity. The muscle strength was found to be 2 distally, 3 proximally in the upper extremity, 5 in bilateral lower extremities, 3 in neck flexion according to 0-5 Medical Research Council [MRC] scale. Cranial and Cervical magnetic resonance imaging (MRI) was done on the patient to rule out central nervous system pathologies, no lesion was detected. Liver and kidney function testes, thyroid hormone levels, vitamin B12 and folic acid levels were within normal ranges. Electromyography (EMG) was ordered to rule out presynaptic or postsynaptic pathologies and to make differential diagnosis of GBS. In the repetitive EMG performed on Trapezius and Abductor digiti minimi muscles no decrementel response was observed. In the EMG performed bilateral upper extremities, local motor axonal polyneuropathy was observed in the motor fibers (Table 1). Lumbal punction was performed on the patient, suspecting the PCB variant of GBS. CSF protein level was found to be 89 mg/dl. No cell was seen. Antiganglioside antibodies were negative (GM1, GQ1b, GD1b, GT1b, GD1a, GM3, GM2). 2gr/kg intravenous immunoglobulin (IVIG) treatment was initiated. At the one-month follow-up visit partial recovery in patient's speech and distal weakness in the hands was observed.

Discussion

GBS is an acute inflammatory demyelinising polyneuropathy (AIDP) that affects motor and sensory fibers. Autoimmunity is suspected in its pathogenesis. It predominantly manifests with muscle weakness, sensory loss and hyporeflexia. It is rarely seen during pregnancy. Although it could be seen all through pregnancy its risk is increased in the 3rd trimester and postpartum period [8]. GBS seen during pregnancy commonly occurs as AIDP with the other forms seldom seen [9,10]. In our case the diagnosis was made in the 3rd trimester in accordance with the literature, however in contrast with the literature our case was not the commonly seen demyelinising form of GBS. It was the axonal form and PCB variant. In a study done on 250GBS patients, the incidence of PCB variant was 3% [5], it was never reported during pregnancy. Our case is the first reported case in the literature.

Before establishing PCB diagnosis spinal cord pathologies with oropharyngeal and cervicobrachial weakness, pathologies in the

Citation: Ekmekçi B, Sağlam S, Tak ZA, Altun Y, Çağ I and Gedik E. Pharyngeal-Cervical-Brachial Variant of Guillain–Barré Syndrome in Pregnancy. Austin J Neurol Disord Epilepsy. 2017; 4(1): 1031.

Table 1: Electromyography results.

Sensory NCS	Latency (ms)	Peak Amplitude (μV)	Velocity(m/s)
R MEDIAN-Digit II	2,24	57,2	53,6
L MEDIAN- Digit II	2,19	61,3	54,9
R ULNAR- Digit V	1,98	37	50,5
L ULNAR- Digit V	1,96	38,8	51,3
L SURAL- Lat Malleolus	2,81	26,3	56,9
Motor NCS	**Latency (ms)**	**Amplitude (mV)**	**Velocity(m/s)**
R MEDIAN - APB	3,3	2,1	52,6
	7,1	1,8	
L MEDIAN - APB	3,6	3,3	51
	6,9	4,1	
R ULNAR - ADM	2,8	2,4	54,1
	6,5	3,2	
L ULNAR - ADM	2,2	3,2	51,2
	6,2	3,7	
R TIBIAL (KNEE) - AH	3,6	4,2	46,2
	11,8	4,7	
L TIBIAL (KNEE) - AH	3,9	5,7	45
	12,4	4,8	

NCS: Nerve Conduction Study; APB: Abductor Pollicis Brevis; ADM: Adductor Digiti Minimi; AH: Abductor Hallucis.

neuromuscular junction and myopathies should be considered in the differential diagnosis. Our case was evaluated with cranial and cervical MRI to rule out brain stem and cervical pathologies. No pathological finding was discovered. Subacute myositis was ruled out as the patient had no muscle pain and no proximal muscle weakness in the physical examination, serum CK levels were within normal range. For the diagnosis of myositis short duration low amplitude polyphasic MUPs are seen in injection EMG, this finding was not seen in our patient. The patient did not have fluctuating weakness that is commonly seen in neuromuscular junction pathologies. Pytosis, fluctuating proximal muscle weakness which worsens with the fatigue test, decrementel response in the repetitive EMG with 3-5-10-20 Hz consecutive impulses are commonly seen in the physical examination of Myastenia Graves (MG). These findings were not present in our patient leading us away from the diagnosis of MG. Botulism was ruled out as the patient did not have pytosis and ophtalmoplegia that are usually seen in that disease. Motor neuron disease could start with local muscle weakness and bulbar involvement. However our patient had decrease in BKAP in EMG without muscular atrophy and high CSF protein levels, making motor neuron disease unlikely.

Autoantibodies against specific neuronal gangliosides were discovered to be associated with different GBS variants [5,11]. Anti-GM1 and anti-GD1a antibodies were associated with Acute Motor Axonal Polyneuropathy (AMAN) [12]. Anti-GQ1b antibody was found to be associated with FS that manifests with ataxia and ophtalmoplegia, and Bickerstaff Brain Stem Encephalitis (BBE) [13]. Anti-GT1a antibody was found to be positive in the PCB variant [5]. In a study done in 220 GBS patients, anti-GT1a was found to be positive in 23 patients. 5 of them had weakness in oropharynx and neck flexion [14]. Anti GT1a is found to be positive in 50% of the PSB variant [2]. In our case anti-GT1a and other ganglioside antibodies were found to be negative. Antibody negativity does not rule out the diagnosis, it is supportive in cases which are diagnosed clinically and neurophysiologically. However we could evaluate the antibody levels after IVIG treatment because of social reasons. This could have affected the antibody levels.

PCB variant is not a pathological diagnosis but the neuronal injury occurs as AMAN type. In a case report with two patients, axonal injury was discovered in the signal transduction study [6]. In another study with two patients, signal conduction speeds were evaluated. Axonal injury was detected, no demyelinisation was observed and temporal dispersion was not found during transduction [15]. Axonal degeneration characterized with local decrease in BKAP amplitude in the upper extremity was not present in our case in accordance with the literature. Demyelinisation and signal block was not observed.

PCB is a rare variant of GBS. PCB variant of GBS has never been reported during pregnancy previously. It should be kept in mind in patients with oropharyngeal and cervicobrachial weakness for utilization of early prompt treatment.

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