

Coincidence of Guillain-Barre Syndrome in a Patient with Cervical Spondylotic Myelopathy, A Case Report

Talal M. Al-Harbi^{1*}, Sameeh O. Abdulmana¹, Mohammed Bin Falah² and Reem F. Bunyan¹

¹Department of Neurology, Neurosciences Centre, King Fahad Specialist Hospital, 6830 Ammar Bin Thabet St, Al Muraikabat, Dammam 32253-3202, Saudi Arabia

²University Medical Center, King Abdullah Medical City, P.O. Box 26671, Adliya, Kingdom of Bahrain

*Corresponding author: Talal M. Al-Harbi, Department of Neurology, Neuroscience Centre, King Fahad Specialist Hospital, 6830 Ammar Bin Thabet St, Al Muraikabat, Dammam 32253-3202, Saudi Arabia, Tel: 00966138442222 Ext: 2278/2270, 00966565836683; E-mail: talal563@yahoo.com

Rec date: Jan 28, 2016; Acc date: Feb 17, 2016; Pub date: Feb 19, 2016

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Abstract

We report a 50-year-old lady who is well known to have cervical spondylotic myelopathy presented to the emergency department with worsening neck pain, numbness and weakness in the hands initially thought to be secondary to progressive cervical myelopathy. However, her symptoms rapidly progressed to flaccid areflexic quadriplegia and respiratory difficulty within few days. Electrophysiological studies and cerebrospinal fluid analysis were consistent with an acquired demyelinating polyradiculoneuropathy. She improved after immunotherapy with intravenous immunoglobulin. Considering this is a rare co-occurrence, neurologists and neurosurgeons should be aware of the coincidence of Guillain-Barré syndrome in a patient who has compressive spondylotic myelopathy to avoid unwanted devastated consequences.

Keywords: Guillain-Barre syndrome; Cervical spondylotic myelopathy; Myelopathy

Introduction

Guillain-Barré syndrome (GBS)-Acute Inflammatory Demyelinating Polyradiculoneuropathy, (AIDP) is an acute polyneuropathy that commonly manifests with areflexic flaccid paralysis associated with variable sensory and autonomic disturbances [1]. Albuminocytologic dissociation; elevated cerebrospinal fluid (CSF) protein without pleocytosis is a typical finding in GBS [1,2]. GBS is a neurological emergency that affects 1:100,000 person-years in the Western World [1,3]. Symptoms progress over a period of up to 4 weeks. During and after the acute phase of GBS, the prognosis is extremely variable, nearly 25% of patients develop respiratory impairment, 20% remain severely disabled, and 5 % may die, despite immunotherapy [1].

Cervical spondylotic myelopathy (CSM) is one of the most common causes of spinal cord dysfunction in older persons [4]. The aging process results in degenerative changes in the cervical spine that in advanced stages can cause compression of the spinal cord [4]. On the other hand, GBS and CSM may both present as a subacute progressive paraparesis or quadriplegia [5]. Patients with GBS typically have reduced or absent reflexes and may have cranial nerve involvement [1]. While patients with CSM are usually present with neck and arm pain with weakness and hyperreflexia and do not have cranial nerve involvement [4]. However, these clinical signs could be absent early at the time of the presentation. Therefore, neuroimaging and electrophysiologic studies are often required in solving the diagnostic complexity [5].

Here, we present a case to report of GBS in a patient with chronic CSM. We encourage detailed neurologic assessment in patients with CSM especially in atypical cases before consideration for surgical intervention.

Case Report

A 50-year-old woman presented to the emergency department (ED) complaining of chronic neck pain that was getting worse associated with tingling sensation and weakness in the upper limbs of seven days duration. There was no history of bulbar dysfunction, sphincter abnormality, or autonomic symptoms. She had no history of preceding upper respiratory tract or gastrointestinal infection. On physical examination, the neurological examination revealed no cranial nerve dysfunction, generalized hypotonia, weakness of the proximal and distal upper and lower limbs muscle power was grade 4+/5 (Medical Research Council scale), and the deep tendon reflexes showed diffuse hyporeflexia. Plantar responses were flexor on both sides, sensory and cerebellar examinations were normal.

Cervical spine MRI showed multilevel degenerative changes with moderate cord compression and myelomalacia, most prominent at the C4-C5 level, with dilated central canal above and below the level of compression, likely as a result of venous congestion and impaired CSF flow (Figure 1). The initial working diagnosis was CSM with subacute worsening.

On the third day of admission, the patient's condition became progressively worse; she developed moderate flaccid quadriplegia (power grade 3/5), absent deep tendon reflexes, and swallowing difficulty. Nerve conduction studies and electromyography demonstrated prolonged distal latencies, temporal dispersion, slowing of the conduction velocities, and absent F waves of all four limbs, consistent with a generalized sensorimotor acquired demyelinating polyradiculoneuropathy. Needle examination revealed denervation activity in the distal muscles and decreased recruitment of the motor units. Furthermore, CSF analysis showed albuminocytologic dissociation with a normal glucose (4.7 mmol/L), high protein (5.6 g/L), and normal WBC count (4; 100% lymphocytes).

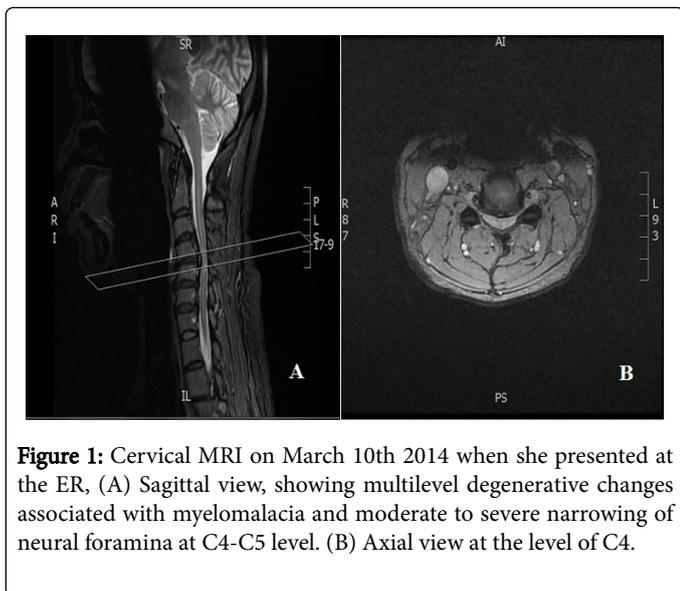


Figure 1: Cervical MRI on March 10th 2014 when she presented at the ER, (A) Sagittal view, showing multilevel degenerative changes associated with myelomalacia and moderate to severe narrowing of neural foramina at C4-C5 level. (B) Axial view at the level of C4.

At that time, her breathing was also affected, with low forced vital capacity (1.4 L). She was transferred to the intensive care unit (ICU) and electively intubated. She was started on immunotherapy with intravenous immunoglobulin (IVIg) 0.4/kg/day for a total of 5 days. On the 5th day of ICU admission, the patient was successfully extubated and transferred to the floor in a stable condition for extensive rehabilitation. Her hematological and routine metabolic workup was normal, and the autoimmune markers, including anti-GM1 antibodies, and Campylobacter serology, all were negative.

Three weeks later, she showed marked improvement. She was able to swallow normally, and her muscle strength improved to 4+/5 and 4-/5 in the upper and lower extremities, respectively. She was discharged home in a satisfactory condition, and prescribed daily physiotherapy. She is scheduled to undergo cervical spine decompression once her overall condition is stable.

Discussion

Acute myelopathy due to spinal cord compression or acute transverse myelitis can be clinically confused with GBS since the deep tendon reflexes may be depressed in the acute stage of spinal cord disease [4]. However, early bowel and bladder dysfunction and a sensory level point to myelopathy that is usually supported by finding a focal lesion in the MRI of the spine [4,5].

When our patient presented to the ED, she had neck pain and numbness in the fingers that were thought to be due to the progression of cervical myelopathy. Neck pain is not common symptoms of GBS; it is more prevalent in CSM. However, neck pain has been rarely reported in the pharyngeal-cervical-brachial variant, which accounts for approximately 3% of total GBS cases [2]. Our case may represent this variant that would also explain why she developed bulbar weakness soon after admission. Alternatively, her disease could have started with axial and neck hypotonia that had aggravated her preexisting cervical roots compression. The rapid progression of her upper limbs weakness followed by acute quadriplegia and areflexia were highly suspicious of Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP).

The diagnostic challenge, in this case, is that her cervical MRI was abnormal when she attended to the ED, showing CSM confirmed by signal changes within the cervical spinal cord at the C4 level (Figure 1). However, comparing the new cervical spine MRI with a previous one done ten months before admission helped in ruling out new or progressive spinal cord injury (Figure 2), since there was no significant difference between the two images.

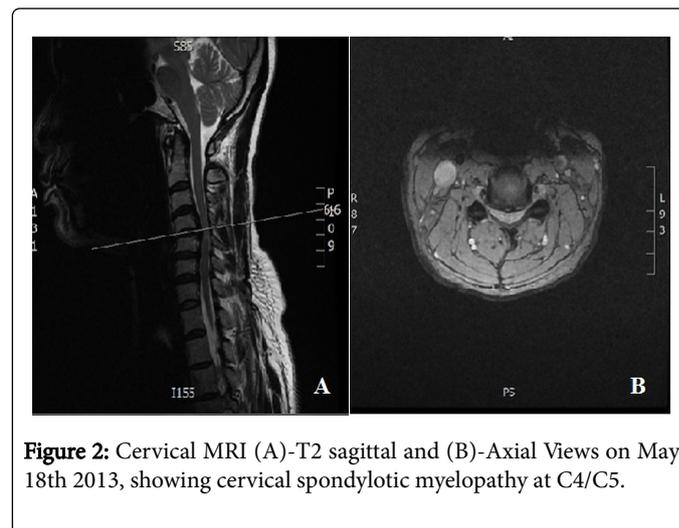


Figure 2: Cervical MRI (A)-T2 sagittal and (B)-Axial Views on May 18th 2013, showing cervical spondylotic myelopathy at C4/C5.

In this case, the diagnosis of GBS was confirmed by the electrophysiological study that demonstrated slowing of the nerve conduction velocities, abnormal temporal dispersion, partial motor conduction block and prolonged distal latencies that are not typical findings of CSM [6].

High protein level in CSF may be observed early in GBS course, although, protein level may not yet be prominent until 1-2 weeks after the onset of weakness, rarely they do remain persistently normal [1,2]. In this case, we diagnosed acute polyradiculoneuropathy superimposing a chronic CSM.

In reviewing the literature, we found no association between GBS and CSM. Only one similar case was reported previously by Abai et al., a 39-year-old lady developed progressive lower, and upper limbs weakness and her MRI neck demonstrated cervical spondylosis with cord compression at C5/C6 [5]. The diagnosis was confirmed by electrophysiological studies. The authors assumed that the patient developed GBS then cervical myelopathy commenced while our patient is known to have CSM then she had GBS.

Considering this case and the previously reported one, variable development of symptoms of GBS may coexist in a patient with CSM highlighting the importance of the making appropriate diagnosis especially in patients considered for surgery.

Conclusion

GBS and cervical myelopathy may co-exist. The clinicians should suspect GBS in a patient with cervical myelopathy who present with symptoms that cannot be explained by cervical cord compression alone.

Conflict of Interest

All authors contributed to the process of manuscript writing. None of the authors have any conflict of interest, which may arise from being named as an author on the manuscript.

Sources of Support

We disclose there are no sources of funding or financial support.

Consent for Manuscript and Figure

The patient and her husband gave written consent for the use of personal and medical information for the publication of this case report and any accompanying images.

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