



Sensory Guillain-Barre Syndrome-Unusual Case

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Abstract

Sensory Guillain-Barre syndrome (GBS) is an acute demyelinating neuropathy that presents clinically with involvement of the sensory peripheral nerve only. However, the existence of a purely sensory form of GBS remains subject to controversy, since these cases always demonstrate a degree of motor weakness or abnormalities in motor nerve conduction studies (NCS) and are difficult to distinguish from acute sensory neuronopathy. We described a cases of an acute sensory demyelinating neuropathy that met most of the proposed diagnostic criteria of a sensory variant of GBS was admitted in Dr. RPGMC TANDA. In this case sensory neuropathy was sudden at onset and peaked to maximal deficit within 3 weeks. No motor weakness was present at the time of admission to hospital. A diagnosis of Guillain-barre syndrome was considered when neurophysiological studies was completed showing evidence of demyelination on sural nerve conduction. We describe this case a sensory variant of GBS on the base of history, clinical examination, Nerve conduction study and pathological finding. Clinical improvement followed treatment within one course of intravenous immunoglobulin (IVIG).

Keywords: Guillain-Barre Syndrome, Intravenous immunoglobulin, Nerve conduction study and demyelinating neuropathy.

Introduction

Sensory Guillain-Barre syndrome (GBS) is an acute demyelinating neuropathy that presents clinically with involvement of the sensory peripheral nerve fibers. the existence of a purely sensory form of GBS remains subject to controversy, since these cases always demonstrate a degree of motor weakness or abnormalities in motor nerve conduction studies (NCS) and are

difficult to distinguish from acute sensory neuronopathy¹ To date, only a few cases of pure sensory GBS have been described.² The diagnostic criteria for sensory variant of GBS should include A rapid onset, widespread and symmetrical distribution, elevated CSF protein with few cells, and nerve conduction studies consistent of a demyelination process. Diagnosing sensory GBS is important since immunotherapy

may positively influence the prognosis, in contrast to the slow but steady progression associated with idiopathic sensory neuropathy or Para neoplastic sensory neuronopathy. Therefore, understanding the neurophysiologies and clinical features may help in the diagnosis of complicated clinical cases and prevent unnecessary procedures.

Case Report

A 30 year old male presented with two weeks of progressive numbness and parasthesia of distal lower limbs that extended to all the limbs over four days, and was admitted to DR.RPGMC KANGRA AT TANDA. The patient experienced acute diarrhea three weeks prior to admission. The patient developed numbness first of all on the soles of both feet, which progressed over three days to the knee; thus, the patient had difficulty with walking due to poor balance. Subsequently, the patient had a markedly unsteady gait and tingling sensations in the distal lower limbs, which increased in intensity and extended more widely. Numbness in the upper limbs was observed, and extended up to elbow within week. Neurological examination demonstrated no weakness but revealed sensory loss to all modalities. There were diminished reflexes in lower extremities and positive Romberg sign. Base line laboratory tests were conducted on the second day following admission. Electrolytes, RBS, HbA1c, blood counts, peripheral neuropathy and infection like HIV ruled out. Thyroid function, vitamin B12 and folic acid deficiency ruled out. Serum protein electrophoresis and autoimmune work up were unremarkable. There were no other known cause for neuropathy and no family history of neuropathy.

Cerebrospinal fluid (CSF) revealed albuminocytological dissociation, 240 mg/dl of protein and a cell count 3/mm.³ Routine NCSs showed prolonged median and peronealdistal motor latencies 8.5ms and 10.8 ms respectively, with prolonged median and ulnar F wave i.e 37ms. There were very small amplitude ulnar

nerve response and no amplitude response for median nerve.

Absent sensory potentials, while the motor NCS was normal. Sagittal T2-weighted magnetic resonance imaging (MRI) scans of the cervical spine revealed a normal appearance of the posterior column. Patient was shifted to icu for monitoring of his respiratory function that remained stable. IVIG had been given to the patient for five days and there was significant improvement in signs and symptoms.

Discussion

Most reported cases of 'sensory GBS' in the literature rely on criteria set forth by Asbury in 1981 for a sensory and are flexic variant of GBS.³ At that time, he stated that 'the precise diagnostic limits of GBS remain uncertain' and this is still true today. Although several variants of GBS are widely recognized, the question of whether patients presenting with predominantly sensory findings can truly be considered to have GBS is still unanswered, with some supporting its inclusion as a subtype and others arguing that it should be considered a separate clinical entity.^{4 5}

There are ongoing efforts to clarify whether there is a place on the GBS spectrum for a sensory variant and to characterize this clinical entity using clinical and pathological findings.⁶

The criteria put forward by Asbury include rapid onset of sensory symptoms, symmetric and widespread distribution, complete or nearly complete recovery, characteristic electro diagnostic results showing demyelination and elevated CSF protein. Despite this, a review of diagnostic criteria for GBS, which is still widely used today, does not formally include the sensory variant. An updated set of eight criteria for sensory GBS was proposed by Oh et al in 2001 and supported by a series of eight cases; however, they have not been widely adopted. The proposed criteria include: (1) acute onset of sensory symptoms, (2) peak deficit achieved within 4 weeks, (3) diminished or absent reflexes, (4) normal motor strength, (5) nerve conduction

evidence of demyelination in at least two nerves, (6) monophasic course, (7) no other known cause for neuropathy and (8) no family history of neuropathy.

Wartenberg⁷ discussed the concept of a sensory equivalent to the ascending paralysis of GBS in 1958. However, their clinical and electrodiagnostic features are pathognomonic of acute sensory neuropathy or sensory GBS remains controversial. To date, reported clinical cases meeting these criteria have been scarce, for which there are two main reasons. Firstly, these cases always demonstrate a degree of motor weakness or abnormalities in motor NCSs, which suggests that the cases are predominantly sensory GBS, rather than purely sensory GBS. Secondly, acute sensory neuropathy represents two clinical syndromes: Acute sensory neuropathy involving the dorsal root ganglia and sensory GBS, an acute demyelinating neuropathy that presents clinically with involvement of the sensory peripheral nerve only. Demonstrating electrophysiological evidence of sensory demyelination in GBS may be difficult with the exception of usual presentation, where there is evidence of demyelination on motor NCSs. Sensory neuropathy can be differentiated by the absent sensory nerve action potentials in the presence of normal motor conduction⁽⁶⁾. In the present case, the sagittal T2-weighted MRI scan of the cervical spine, which demonstrated a normal appearance of the posterior column and fulfills maximum criteria proposed by Asbury. The aforementioned observations indicated that the present case was most likely pure sensory GBS not sensory neuropathy.

Conclusion

GBS is remarkably diverse condition with varied clinical presentation including sensory variants. A sensory variant of GBS should be considered in the differential diagnosis of acutely evolving sensory symptoms even in absence of weakness. Further research into the relationships of acute peripheral neuropathy with specific anti-

ganglioside antibodies may prove helpful in better understanding the varied phenotypes of GBS.⁸ Neurological consultation and NCS and CSF analysis should be done in suspected case of sensory GBS. It has a good prognosis after treatment with intravenous immunoglobulin. However, mild numbness and generalized areflexia symptoms remained with the patient.

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