A Case of Acute Pure Sensory Guillian – Barr Syndrome

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Abstract: Sensory Guillian – Barr syndrome (GBS) is an acute neuropathy that involves sensory peripheral nerves. As very few such cases have been reported till date, its clinical, neurophysiological and pathological features are still not well described and there is no consensus to consider acute sensory ataxic neuropathy (ASAN) as a variant of GBS. This study reports a 50 year old female with acute onset difficulty in walking preceeded by fever, without any motor weakness and with a sensory level over the thoracic region with positive romberg sign. Nerve conduction studies (NCSs) were suggestive of decreased sensory nerve action potential(SNAPs) in bilateral median and superficial peroneal nerves and CSF study was suggestive of albumino cytological dissociation. MRI cervico dorsal lumbar spine with contrast was not suggestive of compressive or non compressive myelopathy. Patient was then given IV immunoglobulin and had satisfactory improvement.

Keywords: Guillian Barr syndrome, sensory nerve action potentials, acute sensory ataxic neuropathy, nerve conduction studies

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I. 1. Introduction

Sensory GBS is an acute demyelinating neuropathy that presents with involvement of either predominantly sensory nerves or only sensory nerves. However, the existence of a purely sensory form of GBS remains subject of controversy, since these cases always demonstrate a degree of motor weakness and abnormalities in motor NCSs and are difficult to distinguish from acute sensory neuropathy(1). Till date only a few cases of pure sensory GBS have been reported (2-4), describing its clinical, neurophysiological and pathological features and response to iv immunoglobulins, leading to delay in treatment. This study reports a 50 year old female with acute onset difficulty in walking preceded by fever without any motor weakness and with a faint sensory level in the thoracic region with positive romberg sign. The presence of sensory level was highly suggestive of a cord lesion but MRI cervico dorsal lumbar cord with contrast was not suggestive of compressive or non compressive myelopathy. NCSs were suggestive of decreased SNAPs in bilateral median and superficial peroneal nerves and CSF study was suggestive of albumino cytological dissociation. Patient was then given IV immunoglobulin and had satisfactory improvement. Diagnosing GBS early in its course is therefore important in view of early initiation of immunotherapy which can markedly improve the outcome and recovery. Sometimes a subjective sensory level in a patient of sensory predominant GBS may be misleading and suggest a cord pathology but GBS should be ruled out by careful evaluation of the patient.

II. CASE REPORT

50 year old female developed flu like symptoms and fever for 2 days, after which she developed difficulty in walking, which was mainly in the form of imbalance which worsened during dark. Within 3 days of onset of neurological symptoms, patient was not able to walk independently and needed support of two people. There was no history suggestive of bladder bowel involvement. She was admitted to Grecian hospital on December 26, 2017 at 7th day of her illness and her neurological symptoms were still progressing. On examination the patient was markedly unsteady which worsened on closing eyes. Power at each joint was 5/5. Tone was decreased at all joints. Reflexes were 1+ in upper limbs and absent in lower limbs. Planter was mute bilaterally. On sensory examination there was impaired fine touch sensation below T4 (upto 40% loss), however, pain and temperature sensation was intact. She also had impaired joint and position sense below anterior superior iliac spine. On examination there was no cranial nerve involvement or ophthalmoplegia. In view of subjective sensory level, MRI cervical dorsal and lumbar spine with contrast was done which was normal except presence of few osteophytes and insignificant disc bulges without any evidence of compressive or non compressive myelopathy. All routine tests and electrolytes were normal. NCS was suggestive of decreased SNAPs (<80% and >70%) in bilateral median nerve and superficial peroneal nerve with mild decrease in sensory conduction velocities (>80%) of the similar nerves with normal latencies and without any evidence of conduction block. Motor NCSs were unremarkable and F waves were normal. CSF examination was then performed on the 8th day of illness and was suggestive of albumino cytological dissociation (protein – 150mg/dl
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and acellular). Patient was unaffordable for anti ganglioside antibody and refused for sural nerve biopsy. On day 9th of illness intravenous immunoglobulins were started (2gm/kg given over 5 days). Clinical symptoms started improving and after 2 weeks of giving iv immunoglobulin therapy there was satisfactory neurological improvement and the patient was able to walk independently.

III. Discussion

However, Wartenberg (3) and Asbury (5) have discussed concept and criteria of sensory GBS, but pure sensory variant of GBS still remains a controversial entity. Usually sensory cases of GBS also have mild motor weakness clinically so most of these cases are predominately sensory instead of pure sensory variant of GBS, therefore pure sensory GBS remains a rare entity. Till date no variant of GBS has been reported with a sensory subjective level as in our patient, which may be misleading enough to delay the diagnosis and management of GBS.

References