

Isolated H Reflex Abnormality as the Sole Electrophysiological Marker of Demyelinating Neuropathy at Day -14- A Novel Observation

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Abstract

Review Article

Guillain Barre Syndrome (GBS) is a clinical diagnosis with typical electrophysiological features which evolve over 1-2 weeks. H reflex and F wave abnormalities are the earliest to appear. In this report, H reflex abnormality as an isolated feature even after two weeks of illness is an unusual feature which is highlighted by this case report. 38 year old male patient presented with history of antecedent febrile illness presented with progressive weakness of lower limbs of three days duration. He had areflexic proximal more than distal weakness of lower limbs more than upper limbs. Cerebrospinal fluid analysis showed albuminocytological dissociation. Nerve conduction study done on day 3 and day 13 showed an absent H reflex despite clinical progression. Patient responded well to immunotherapy. The importance of minor but definitive abnormalities on nerve conduction when collaborated with clinical findings is extremely useful in the clinical diagnosis of GBS.

Keywords: Guillain Barre Syndrome (GBS), electrophysiological features, H reflex abnormality, Nerve conduction.

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INTRODUCTION

A 38-year-old gentleman was seen in Neurology OPD with 3 days history of difficulty in walking. His wife noted that his gait had changed, and he appeared to walk with effort with a tendency to stumble. He also had difficulty in getting up from a chair, climbing stairs and in bearing his weight on the knees by day 2. He had no upper limb weakness, cranial nerve symptoms, sensory or bladder involvement. Three weeks ago, he had a febrile illness with a total duration of 10 days with transaminitis and lymphadenopathy which was later diagnosed as Infectious mononucleosis based on serology. He had an unremarkable general physical and systemic examination. Neurological examination showed normal higher mental functions and cranial nerves. Tone was decreased in all four limbs. Power was 4+/5 in proximal upper limbs and 4/5 in proximal lower limbs; 4+/5 in distal lower limbs and normal in distal upper limbs. He was able to walk without support but buckled his knees rarely. He was unable to walk on toes or on the heels even after repeated attempts. There was diffuse areflexia; plantar were flexor. Sensory examination and Romberg test were normal.

After admission, he underwent a nerve conduction study which showed bilateral absence of H reflex. There was no conduction slowing or blocks, or F wave abnormalities. Detailed blood work up was noncontributory which included vasculitis profile, ANA, Thyroid function, electrolytes, serum ACE level, B12 level, HIV, Hep B and C. After 48 hours of admission, he reported a modest improvement of symptoms and was discharged from the hospital.

He returned to us 1 week after discharge with three days history of worsening weakness. By that time required mild support to walk and could not get up from a low stool. Muscle power was only 3/5 in proximal lower limbs and 4/5 in distal lower limbs while upper limb power was 4-/5 and 4/5 in proximal and distal lower limbs respectively. Reflexes were all absent. Bilateral and mild neck flexor weakness was also observed. Bulbar muscles were normal and single breath count was 34. Cerebrospinal fluid examination revealed high protein (140 mg/dl) with normal sugars and no cells (albuminocytological dissociation). The nerve conduction study was reported which did not show any new finding except absence of H reflex. He was treated with a five-day course of intravenous

gamma globulin (IVIG 400 mg/kg/d) to which he responded explicitly.

DISCUSSION

Electrophysiological studies play a crucial role in diagnosis and classification of GBS though non-specific patterns evolve over time [1]. Though not able to delineate pathologies, absence of H-reflex is one of the earliest neurophysiological abnormality in GBS. A monosynaptic reflex by-passing the muscle spindle, it points to the proximal segments of the peripheral nerve. Its tendency to be absent even before the clinical presentation, makes it an apt tool for an earlier diagnosis [2].

In clinical neurophysiological parlance, 2 distinct terminologies are widely used, namely VEGBS (<4 days) and EGBS (<10 days) with reference to the timeline in GBS. During both these stages of disease, electrophysiological abnormalities maybe detected in up to 2/3 of cases. But, may not be classical or conclusive to make a diagnosis of demyelinating neuropathy. Among this early electrodiagnostic findings, most studies have highlighted importance of changes in late responses. The F wave latency maybe prolonged or the F wave persistence maybe reduced to <20%. Reduction in distal CMAP amplitude, prolongation of distal motor latency and temporal dispersion of upper and lower limb nerves are other findings which can appear in the early stages [3].

Berciano et-al, in an analysis of 59 VEGBS patients observed F Wave abnormalities in 47.5% and CMAP amplitude reduction ranked second with 45.7%. They further described abnormalities in SNAP amplitude motor to distal amplitude ratios along with other features of demyelination in a lesser proportion of

cases. H reflex abnormality was not evaluated in these subjects. Nevertheless, evaluation and reproduction of H reflex abnormalities is easier and less time consuming, as well as less subject to inter observer variation, compared to other markers of EGBS [4].

Electrophysiological tests, though useful in classifying and prognosticating GBS are inconclusive by themselves. Varied patterns which can be obtained are quite unpredictable with respect to time or presentation. Absence of H-reflex is identified to be the earliest neurophysiological marker, and with time other abnormalities and patterns mark their presence. Rarely absence of H-reflex can be the sole neurophysiological manifestation and can be identified with an eye of suspicion in the appropriate clinical scenario, supported with adequate biochemical evidence.

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