Intact thumb reflex in areflexic Guillain Barré syndrome: A novel phenomenon

Karkal Ravishankar Naik1,2, Aralikatte Onkarappa Saroja1,2, Manik Mahajan2
1Departments of Neurology, KLES’ Dr Prabhakar Kore Hospital and Medical Research Centre, 2KLE University’s Jawaharlal Nehru Medical College, Belgaum, Karnataka, India

Abstract

Introduction: Areflexia is one of the cardinal clinical features for the diagnosis of Guillain Barré syndrome. However, some patients may have sluggish proximal muscle stretch reflexes. Presence of thumb reflex, a distal stretch muscle reflex has not been documented in Guillain Barré syndrome. Materials and Methods: We prospectively evaluated thumb reflex in Guillain Barré syndrome patients and age matched controls from April to September 2013. Results: There were 31 patients with Guillain Barré syndrome in whom thumb reflex could be elicited in all (24 brisk, 7 sluggish), whereas all the other muscle stretch reflexes were absent in 29 patients at presentation and the remaining two had sluggish biceps and quadriceps reflexes ($P = 0.001$). Serial examination revealed gradual diminution of the thumb reflex ($P < 0.001$). Rapid progression of weakness was associated with early loss of the thumb reflex. Conclusion: Thumb reflex, a distal stretch reflex is preserved in the early phase of Guillain Barré syndrome.

Key Words
Acute flaccid paralysis, Guillain Barré syndrome, muscle stretch reflex, thumb reflex

For correspondence:
Karkal Ravishankar Naik, Professor, Department of Neurology, KLE University’s Jawaharlal Nehru Medical College and KLES’ Dr Prabhakar Kore Hospital and Medical Research Centre, Belgaum, Karnataka - 590 010, India.
E-mail: krnaik60@yahoo.com

Ann Indian Acad Neurol 2014;17:199-201

Introduction
Guillain Barré syndrome (GBS) is the most common cause of immune-mediated acute flaccid paralysis. Areflexia is one of the definitive diagnostic features for GBS. Some patients with GBS may have sluggish proximal reflexes with distal areflexia and rarely may have uniformly brisk reflexes. Thumb reflex consists of flexion of distal phalanx of the thumb upon tapping the flexor pollicis longus tendon in distal forearm. Sudden pressure over the volar lateral forearm during examination of a patient with otherwise areflexic GBS produced brisk flexion at the interphalangeal joint of the thumb. This phenomenon prompted us to evaluate prospectively the thumb reflex in GBS patients.

Materials and Methods
Patients with acute flaccid paralysis presenting from April to September 2013 fulfilling the National Institutes of Neurological and Communicative Disorders and Stroke (NINCDS) criteria for GBS were included in the study. Demographic data and details of antecedent illness, symptom duration and disease progression were recorded. All patients underwent detailed neurological examination. Muscle power was assessed by the Medical Research Council (MRC) scale and the clinical disability was graded by Hughes score. Thumb reflex was elicited on both sides independently by percussing the flexor pollicis longus tendon in the distal forearm, which was in supinated position. The finger placed over the tendon was percussed at the junction of distal one-third and proximal two-third of the forearm just lateral to the flexor carpi radialis resulting in flexion at interphalangeal joint of the thumb. [Video 1 and Figure 1] Nerve conductions were performed within two days after admission. Motor and sensory nerve conductions with elicitation of F-waves were performed in at least two upper and two lower limb nerves. Electrophysiological classification of GBS was done using earlier published criteria. ANOVA with post-hoc analysis was done using Statistical Package for the Social Sciences software (SPSS for Windows version 17.0).

Video files available on www.annalsofian.org

Quick Response Code: Website: www.annalsofian.org
DOI: 10.4103/0972-2327.132628

Ann Indian Acad Neurol. April-June 2014, Vol 17, Issue 2
Results

There were 31 patients present during the study period (17 males and 14 females) with age ranging from 13 to 78 years (40.9 ± 18.1). Twenty-two patients presented within one week of illness (71%), four during second week (12.9%) and five during third and fourth weeks (16.1%). Eighteen patients (58%) had antecedent events among whom 15 had diarrhea (83.34%) and three patients had fever. Twenty-five patients had lower limb onset of weakness (80.64%) and six had upper limb onset. Weakness progressed to upper or lower limbs within six hours to seven days from onset of weakness (median 1 day). Maximum weakness occurred 2 to 25 (9.36 ± 5.11; median 9) days from the onset. Twenty-one patients (68%) had sensory symptoms among whom two had impaired sensations. Facial weakness was present in seven patients (22.6%) and six had bulbar dysfunction (19.36%). Autonomic dysfunction with variations in blood pressure and heart rate was seen in eight patients. Five patients required mechanical ventilation for 7 to 60 days (mean 26.4 days).

Nerve conduction studies were available in 29 patients among whom 20 patients (69%) had features of demyelination and nine had axonal motor neuropathy. Nerve conduction could not be done in two patients, who required mechanical ventilation at admission and succumbed later due to respiratory infection. Electromyographic recording of the thumb reflex using surface electrodes (active electrode over the flexor pollicis longus and reference electrode over its tendon) revealed consistent response [Figure 2].

At admission, lower limb muscle stretch reflexes were absent in all patients. Biceps, supinator and triceps reflexes were absent in 29 patients and were depressed in two patients. Thumb reflex could be elicited in all the 31 patients at presentation; brisk in 24 patients and depressed in seven patients Among healthy control subjects, thumb reflex was brisk in 13, depressed in 14, and absent in four [Table 1]. The sensitivity of the test was 100 percent and the specificity was 53.45 percent. Serial examinations of thumb reflex were available in 27 patients among whom the thumb reflex continued to be brisk in four patients, depressed in 18 and became absent in five patients after one week. Among the five patients in whom thumb reflex became absent in the first week, three required mechanical ventilation.

Discussion

Acute flaccid paralysis of more than one limb with areflexia constitutes the core clinical criteria for GBS. However, some patients may have sluggish biceps and quadriceps muscle stretch reflexes.[5] Several electrophysiological criteria have been proposed for classification of GBS as demyelinating and axonal forms.[6,7] All the patients in the present study conformed to clinical and electrodagnostic features of GBS. Demographic variables, antecedent events, clinical course and progression of disease in our patients were consistent with previous studies.[7,8] Facial and bulbar involvements were similar to the case series from Japan and China where axonal GBS is more common[9] whereas it was less frequent compared to previous publications from India[10] and western countries.[8]

Thumb reflex is a distal muscle stretch reflex in the upper limb and consists of contraction of flexor pollicis longus on percussion of its tendon. It is mediated through the anterior interosseous nerve, a branch of median nerve. Peripheral and central responses to passive mechanical stretching of the thumb have been evaluated in few central nervous system disorders.[11,12] Literature survey did not reveal publications on the presence of the thumb reflex in peripheral nervous system disorders like GBS.

In the present cohort of 31 GBS patients, thumb reflex was universally present at admission in all the patients irrespective

Table 1: Showing the incidence of thumb reflex among control subjects and patients with Guillain Barré syndrome. Control vs. patients at admission P = 0.001, patients at admission vs. at one week P < 0.001

| Controls (%) | Patients |
|--------------|----------|
|              | At admission (%) | At one week (%) |
| Brisk        | 13 (41.94) | 24 (77.4) |
| Diminished   | 14 (45.16) | 7 (22.6)  |
| Absent       | 4 (12.90)  | 0          |

Figure 1: Photograph showing the method for elicitation of the thumb reflex. The examiner’s thumb placed over flexor pollicis longus on the volar aspect of the forearm (junction between distal 1/3 and proximal 2/3) is struck firmly with reflex hammer

Figure 2: The surface showing recorded electromyographic (EMG) signals from flexor pollicis longus. EMG signals triggered the sweep with delay of -5 divisions. Waves in the top are the superimposed eight trials and the bottom trace is the averaged response
of the muscle power while all the other muscle stretch reflexes were absent in 29 patients. Though the thumb reflex was brisk in the initial period in over three-fourth of cases, there was progressive diminution as the disease course progressed. Patients with early diminution of the thumb reflex had rapid worsening of weakness. The presence of this reflex could indicate relative sparing of anterior interosseous nerve in the initial phase of GBS, which gets involved later as reflected by subsequent diminution of the reflex. However, this was not clinically supported as the power in flexor pollicis longus did not correlate with briskness of the reflex. None of the patients had other clinical features of upper motor neuron dysfunction. Preservation of the thumb reflex when other muscle stretch reflexes are absent has not been documented to the best of our knowledge in patients with GBS. Patients with GBS have been documented to have isolated or generalized hyperactivity of muscle stretch reflexes during the course of the illness. Central mechanism, possibly caused by dysfunction of spinal inhibitory interneurons, was proposed by some authors. At this juncture, the pathophysiological basis for the persistence of this reflex in GBS patients is unclear.

Electrophysiological correlation of the muscle stretch reflexes which are conventionally examined during clinical evaluations have been evaluated in few studies. The sweep in the electromyographic equipment is triggered by a sensor in the specially constructed reflex hammer thereby enabling the examiner to assess the latency and the projected conduction velocity in the muscle stretch reflex. Tendon reflex (T-reflex) can be recorded in commercially available electromyographic equipment using dedicated reflex hammer incorporating similar method to initiate the data acquisition. This could help measuring the latency to the onset of the reflex, which should be able to document the physiological and electromyographic correlation of the thumb reflex. Further studies on larger cohort of patients along with electrophysiological recording of the reflex may define the utility of the thumb reflex in GBS. We propose that preservation of thumb reflex should not be a clinical feature against the clinical diagnosis of GBS in early phase of the illness.

Conclusion

To the best of our knowledge, this is the first report documenting the presence of thumb reflex in areflexic GBS patients. Thumb reflex was present in all the cases of Guillain Barré syndrome during initial period and waned off during the later phase of the illness. Rapid progression of weakness was associated with early loss of thumb reflex. We reiterate that thumb reflex is a novel clinical sign seen in early phase of GBS.

References

1. Asbury AK, Cornblath DR. Assessment of current diagnostic criteria for Guillain Barré syndrome. Ann Neurol 1990;27:S21-4.
2. Podnar S, Vodusek DB. Hyperreflexia in a patient with motor axonal Guillain-Barre syndrome. Eur J Neurol 2000;7:727-30.
3. Campbell WW. Dejong’s The Neurological Examination. 7th ed. New Delhi: Wolter Kluwer Pvt Ltd; 2013.
4. Hughes RA, Newson-Davis JM, Perkin GD, Pierce JM. Controlled trial prednisolone in acute polyneuropathy. Lancet 1978;2:750-3.
5. Hadden RD, Cornblath DR, Hughes RA, Zielasek J, Hartung HP, Toyka KV, et al. Electrophysiological classification of Guillain Barre syndrome: Clinical associations and outcome. Plasma Exchange/Sandoglobulin Guillain-Barré Syndrome Trial Group. Ann Neurol 1998;44:780-8.
6. Cornblath DR. Electrophysiology in Guillain Barre syndrome. Ann Neurol 1990;27:S17-20.
7. van Doorn PA, Ruts L, Jacobs BC. Clinical features, pathogenesis, and treatment of Guillain-Barré syndrome. Lancet Neurol 2008;7:939-50.
8. Winer JB, Hughes RA, Osmond C. A prospective study of acute idiopathic neuropathy. I. Clinical features and their prognostic value. J Neurol Neurosurg Psychiatry 1998;51:605-12.
9. Ho TW, Mishu B, Li CY, Gao Cy, Cornblath Dr, Griffin JW, et al. Guillain-Barre syndrome in northern China: Relationship to Campylobacter jejuni infection and anti-ganglioside antibodies. Brain 1995;118:597-605.
10. Kannan MA, Ch RK, Jabeen SA, Mridula KR, Rao P, Borgohain R. Clinical and Electrophysiological subtypes and antiganglioside antibodies in childhood Guillain-Barre Syndrome. Neurol India 2011;59:727-32.
11. Rothwell JC, Oseso JA, Traub MM, Marsden CD. The behaviour of the long-latency stretch reflex in patients with Parkinson’s disease. J Neurol Neurosurg Psychiatry 1983;46:35-44.
12. Cody WJ, Richardson CH, MacDermott N, Ferguson IT. Stretch and vibration reflexes of wrist flexor muscles in spasticity. Brain 1987;110:433-50.
13. Kuwabara S, Ogawara K, Koga M, Mori M, Hattori T, Yuki N. Hyperreflexia in Guillain Barre syndrome: Relation with acute motor axonal neuropathy and anti-GM1 antibody. J Neurol Neurosurg Psychiatry 1999;67:180-4.
14. Péron Y, Nguyen The Tich S, Fournier E, Genet R, Guihéneuc P. Electrophysiological recording of deep tendon reflexes: Normative data in children and in adults. Neurophysiol Clin 2004;34:131-9.

How to cite this article: Naik KR, Saroja AO, Mahajan M. Intact thumb reflex in areflexic Guillain Barré syndrome: A novel phenomenon. Ann Indian Acad Neurol 2014;17:199-201.

Received: 31-12-13, Revised: 03-02-14, Accepted: 05-03-14

Source of Support: Nil, Conflict of Interest: Nil