

“JAW CLONUS” – A Rare yet Fascinating Clinical Sign in Neurology

Bhawna Sharma^{1*}, Raghavendra Bakki Sannegowda¹, Ashok Panagariya¹ and Pankaj Gupta²

¹Department of Neurology, SMS Medical College Hospital, Jaipur, Rajasthan, India

²S M S Medical College, Jaipur, Rajasthan, India

*Corresponding author: Bhawna Sharma, Department of Neurology, SMS Medical College Hospital, Jaipur, Rajasthan, India, Tel: 9986144799; E-mail: sharmadrhawna@gmail.com

Rec date: March 20, 2014; Acc date: April 14, 2014; Pub date: April 16, 2014

Copyright: © 2014 Sharma B, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Clonus is one of the important clinical sign elicited in neurology. It is the rhythmic muscle contraction which usually occurs in patients with lesions involving descending motor pathways. Repetition of this sequence of events induces rhythmic oscillation of some joints, the frequency of which is generally between 5 and 8 Hz. Clonus usually elicited at ankle and knee. Jaw clonus is rarely reported. We report here jaw clonus in a patient who presented with vascular dementia and Parkinsonism.

Keywords: Jaw clonus; stretch reflex; corticobulbar fibres

Introduction

Clonus is a series of rhythmic involuntary muscular contractions occurring at a frequency of 5–7 Hz in response to an abruptly applied and sustained stretch stimulus [1]. It often accompanies the spasticity and hyperactive deep tendon reflexes seen in corticospinal tract disease. Clonus is usually elicited at ankle and patella [2]. Pronator clonus been described in literature and wrist clonus been reported rarely [2,3]. Clonus of jaw can occur occasionally and case reports with jaw clonus are very few.

Case Report

55 year old male chronic smoker and known case of hypertension with history of stroke in the form of right hemiparesis four months back with partial recovery, presented to us with abrupt onset, fluctuating course and stepwise progressive dementing illness with Parkinsonism. He had history of behavioral changes in the form of apathy, social incontinence for bladder and memory impairment often improved with clues. He also had history of slowness in all activities, recurrent falls and monotonous speech. There was no history of inappropriate laughing, crying or difficulty in swallowing.

On examination patient had hypomimetic face, low volume monotonous speech, frontal release signs in the form of glabellar tap, palmo-mental reflex. His Hutchinski score was 11. He had bradykinesia and cogwheel rigidity at wrist joint bilaterally. He also had bilateral pyramidal signs in the form of spasticity at elbow joints, pronator catch, spasticity in lower limbs, brisk reflexes with asymmetry (R>L). His plantars were extensor bilaterally. His gait was characterized by short steps, reduced arm swing admixed with right sided spastic hemiplegic gait. However an interesting clinical finding was noticed in our patient, upon eliciting jaw jerk it was not only brisk but also there was persistent clonic movement. Jaw clonus was persistent as long as finger was kept over jaw with sustained pressure. His MRI brain showed multiple infarcts in subcortical structure involving basal ganglia, internal capsule and periventricular areas (Figure 1a, b).

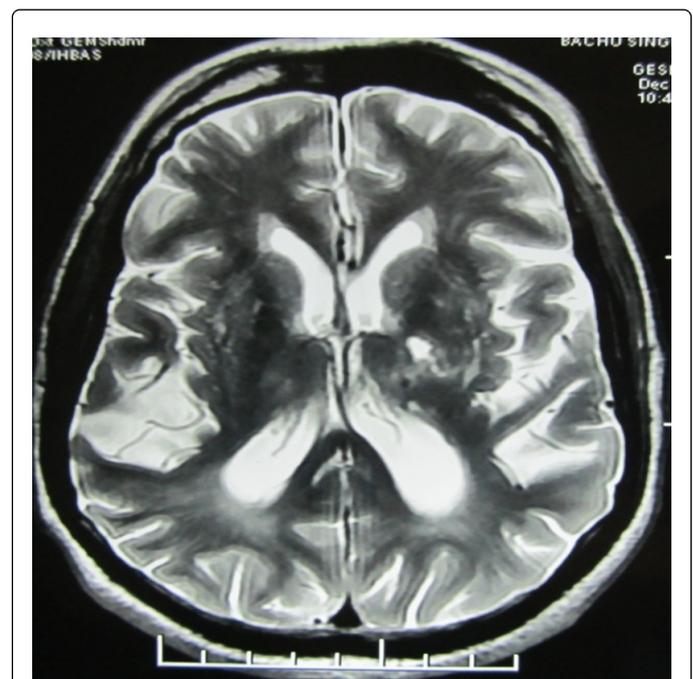


Figure 1a: T2W MRI brain showing infarcts in subcortical structure involving left basal ganglia, internal capsule and periventricular areas.

Discussion

Clonus depends for its elicitation on an appropriate degree of muscle relaxation, integrity of the spinal stretch reflex mechanisms, sustained hyperexcitability of the α -motor neurons and γ -motor neurons (suprasegmental effects) and synchronisation of the contraction-relaxation cycle of muscle spindles [1]. Simpler mechanisms of clonus are alternating stretch reflexes [2]. It is also hypothesised that clonus results from the action of the central

oscillator [4]. Ankle and patellar clonus are commonly described [2]. Pronator clonus, while eliciting pronator catch, may occur owing to severe hypertonus and wrist clonus though rare been reported [2]. Sharma B et al. [3] and sung et al. [5] reported a rare case of wrist clonus mimicking action induced tremors.

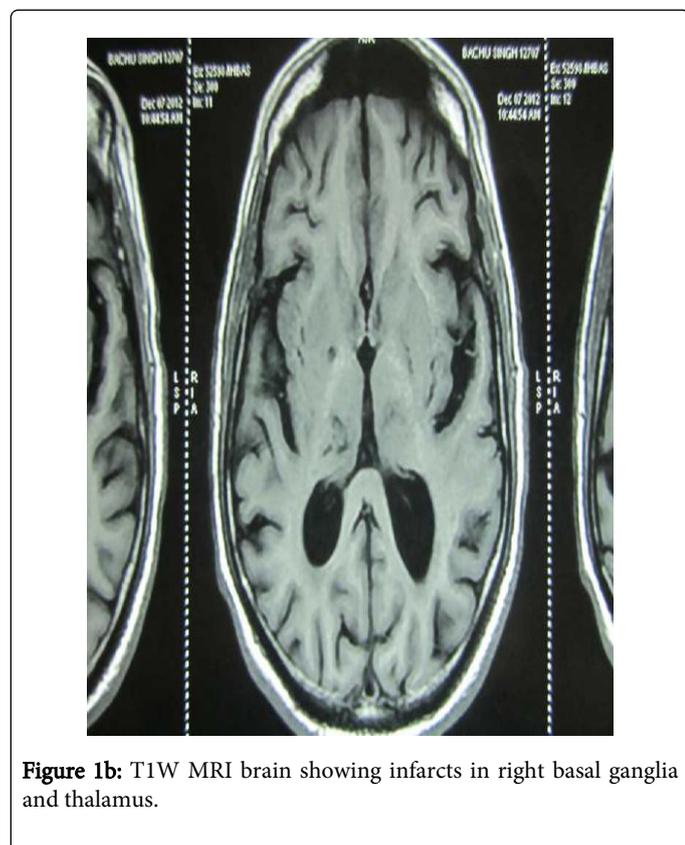


Figure 1b: T1W MRI brain showing infarcts in right basal ganglia and thalamus.

The jaw jerk was discovered by accident and was described in 1885-1886. It was a late sequel of Whytt's seminal description of the stretch reflex. The jaw jerk is a brisk, partial, upward jerk of the jaw caused by contraction of the temporalis, masseter and medial pterygoid muscles in response to striking the chin when the mouth is open. In patients with supranuclear lesions of the trigeminal nerve (eg, amyotrophic lateral sclerosis, extensive multiple sclerosis, bilateral corticobulbar infarction with pseudobulbar palsy), the jaw jerk is exaggerated and clonus may occur [6].

Jaw muscles have two types of spindle afferents based on their sensitivity. Afferents with high sensitivity during stretching of the jaw muscles and their silencing during the release phase of muscle stretch were classified as primary-like spindle afferents. A second type of tonic afferents was modulated only modestly during stretching of the jaw-elevator muscles classified as secondary-like spindle afferents because of their low dynamic sensitivity during ramp muscle stretch and their continued discharge during the release phase of muscle stretch [7]. The projection of jaw-muscle spindle afferents to caudal brainstem

regions plays a significant role in masticatory-muscle stretch reflexes and in the integration of trigeminal proprioceptive information and its transmission to higher centers [8]. There lies a multiple disynaptic jaw-muscle spindle afferent-motoneuron circuits and these pathways convey long-latency jaw-muscle stretch reflexes and may contribute to stiffness regulation of the masticatory muscles [9].

Beevor, et al. [10] described a case of amyotrophic lateral sclerosis with clonus of the lower jaw way back in 1886. Iodice et al reported a case of jaw tremor in a case of amyotrophic lateral sclerosis [11]. Exaggerated jaw jerk is considered due to supranuclear lesion of the trigeminal nerve and it may occur in ALS [12]. Worster- Drought [13] described 82 cases of varying degrees of congenital suprabulbar paresis in which they mentioned exaggerated jaw jerk in almost all patients. Though exact number of percentage was not mentioned, they described that often patients in that series had jaw clonus.. Here we describe a rare case of jaw clonus in a patient of vascular dementia with bilateral pyramidal and extrapyramidal features. Though there was no pseudobulbar effect or spastic tongue in our patient, possible hypothesis for brisk jaw jerk with jaw clonus was multiinfarct state involving corticobulbar fibres.

References

1. Ropper AH, Samuels MA (2009) Adams and Victor's Principles of neurology (9thedn) McGraw Hill, New York, USA.
2. Campbell WW (2005) Dejong's The neurologic examination (6thedn) Lippincott Williams and Wilkins, Philadelphia.
3. Sharma B, Sannegowda RB, Nagpal K, Jain R (2012) Wrist clonus mimicking as action-induced tremors: an important clinical lesson. BMJ Case Rep 2012.
4. Dimitrijevic MR, Nathan PW, Sherwood AM (1980) Clonus: the role of central mechanisms. J Neurol Neurosurg Psychiatry 43: 321-332.
5. Sung YH, Park KH, Lee YB (2010) A case of action-induced clonus that mimicked action tremors and was associated with cervical schwannoma. Journal of movement disorders 3:48-50.
6. Pearce JM (2011) The jaw jerk: an instance of misattribution. J Neurol Neurosurg Psychiatry 82: 351-352.
7. Dessem D, Donga R, Luo P (1997) Primary- and secondary-like jaw-muscle spindle afferents have characteristic topographic distributions. J Neurophysiol 77: 2925-2944.
8. Luo P, Wong R, Dessem D (1995) Projection of jaw-muscle spindle afferents to the caudal brainstem in rats demonstrated using intracellular biotinamide. J Comp Neurol 358: 63-78.
9. Luo P, Moritani M, Dessem D (2001) Jaw-muscle spindle afferent pathways to the trigeminal motor nucleus in the rat. J Comp Neurol. 435: 341-353.
10. Beevor CE (1886) A case of amyotrophic lateral sclerosis with clonus of the lower jaw. Brain 8: 516-519.
11. Iodice R, Manganelli F, Dubbioso R, Ruggiero L, Santoro L (2014) Teaching video neuroImages: Clonus of the lower jaw: an old sign that comes back.. Neurology 82: e96.
12. Kiernan MC, Vucic S, Cheah BC, Turner MR, Eisen A, et al. (2011) Amyotrophic lateral sclerosis. Lancet 377: 942-955.
13. Worster-Drought C (1956) Congenital suprabulbar paresis. J Laryng 70: 143.