The split hand sign

Rajesh Benny, Kishore Shetty[1]

Department of Neurology, Fortis and MGM Hospitals, [1]Neurology, Hiranandani Hospital, Mumbai, Maharashtra, India

Abstract

Amyotrophic Lateral sclerosis (ALS) is a disease characterized by pure motor asymmetric wasting of various muscles with associated upper motor neuron signs. The split hand sign, which is because of dissociated muscle weakness in the hands (thenar muscles disproportionately wasted as compared to the hypothenar muscles) is a useful clinical sign for bed side diagnosis of ALS.

Key Words

Amyotrophic lateral sclerosis, dissociated intrinsic hand muscle weakness, split hand sign

For correspondence:

Dr. Rajesh Benny, MGM Hospital, Sector 3, Vashi, Navi Mumbai, Maharashtra, India. E-mail: rajeshbenny@yahoo.com

Ann Indian Acad Neurol 2012;15:175-6

Introduction

Amyotrophic lateral sclerosis (ALS) is a pure motor neurodegenerative disease where there is asymmetric involvement of the upper and lower motor neurons. In the intrinsic muscles of the hands, there is preferential wasting of the abductor pollicis brevis (APB) and first dorsal interosseous muscle (FDI) (thenar muscles) as compared to the abductor digiti minimi (ADM) (hypothenar muscle) [Figures 1 and 2]. This peculiar pattern of dissociated intrinsic hand muscles was described by Dr Asa Wilbourn in 1996 as 'Split Hand'.[1,2] The clinical deficit is loss of the pincer grasp.

APB, FDI, and ADM are innervated by spinal motor neurons of the same segments (C8 and T1), and FDI and ADM have the same ulnar nerve supply. It is not known why APB and FDI are preferentially affected compared with ADM in those with ALS. Pure motor wasting of the intrinsic hand muscles is seen in the setting of various pathological states affecting the motor neurons [Spinal muscular atrophy (SMA), cervical spondylotic amyotrophy, as a component of peripheral neuropathy, spinocerebellar ataxia type 3 (SCA 3) and even in normal elderly individuals][3].

Specificity of the sign

Satoshi Kuwabara *et al,* and the Tokyo Metropolitan Electrodiagnosis Study Group[4] looked at the specificity of this dissociated muscle wasting and weakness in patients with ALS. The authors examined 77 patients with ALS and compared them with normal controls (n = 171) and disease controls (n = 196). The disease controls had lower motor neuron involvement other than ALS (SMA, bulbospinal muscular atrophy, cervical spondylotic amyotrophy, SCA 3 and peripheral polyneuropathy). The study looked at absolute CMAP amplitudes of APB, FDI and ADM. They also compared the ratios of APB/ADM and FDI/ADM. The authors found that patients with ALS had markedly reduced APB and FDI CMAP amplitudes (thenar involving) with higher ADM amplitudes (hypothenar sparing). APB/ADM and FDI/ADM ratios were therefore the lowest among ALS patients. Reduction in both the APB/ADM and FDI/ADM ratios was seen in 20% of patients of ALS, none of the 171 normal controls and in only 1 (0.5%) of the disease controls. This study demonstrated conclusively that the split hand sign was fairly specific for ALS.

Access this article online	
Quick Response Code:	Website: www.annalsofian.org
	DOI: 10.4103/0972-2327.99700

Pathophysiology

The exact cause of this preferential muscle involvement in ALS is not known. Both lower motor neuron and cortical dysfunction hypotheses have been postulated.[5,6] Humans use more of the pincer grasp (involving the APB and FDI muscle). This may place more oxidative stress/metabolic demand on the spinal motor neurons innervating these two muscles. Also, the corticospinal connections to APB and FDI far outnumber those to the ADM. This may result in more glutamate excitotoxicity to the APB and FDI spinal neurons.

However, Schelhaas *et al,* argued that the presence of this sign in diseases with different pathophysiological mechanisms like autosomal dominant spinal muscular atrophy, juvenile muscular atrophy and SCA 3 weakens the theory of the cortical origin of degeneration in ALS and supports the

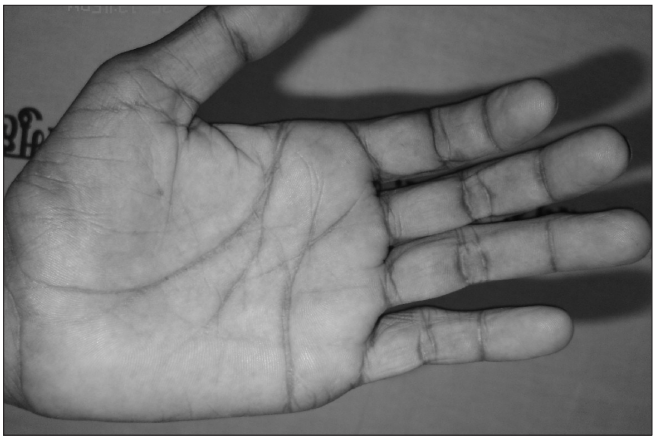

Figure 1: Wasting of the APB (thenar muscle) with sparing of the hypothenar muscles

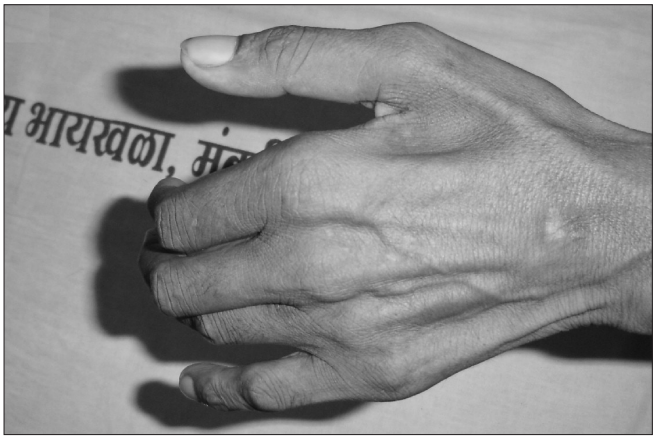

Figure 2: Involvement of the first dorsal interosseous

theory of an intrinsic vulnerability of spinal motor neurons sub serving the thenar complex.[7]

Conclusion

The presence of the 'split hand sign' should alert the clinician to the possible diagnosis of ALS.

References

1. Wilbourn AJ. Electrodiagnostic evaluation of the patient with possible ALS. In: Belsh JM, Schiffman PL, editors. Amyotrophic lateral sclerosis. Armonk, NY: Futura Books; 1996. p. 183.
2. Wilbourn AJ. The "split hand syndrome". Muscle Nerve 2000;23:138.
3. Eisen A, Kuwabara S. The split hand syndrome in amyotrophic lateral sclerosis. J Neurol Neurosurg Psychiatry 2012;83:399-403.
4. Kuwabara S, Sonoo M, Komori T, Shimizu T, Hirashima F, Inaba A, *et al.* and the Tokyo Metropolitan Electro-diagnosis Study Group. Dissociated small hand muscle atrophy in amyotrophic lateral sclerosis: Frequency, extent & specificity. Muscle Nerve 2008;37:426-30.
5. Einsen AA. Comment on the lower motor neuron hypothesis. Muscle Nerve 1993;16:870-1.
6. Kuwabara S, Mizobuchi K, Ogawara K, Hattori T. Dissociated small hand muscle involvement in amyotrophic lateral sclerosis detected by motor unit number estimates. Muscle Nerve 1999;22:870-3.
7. Schelhaas HJ, van de Warrenburg BP, Kremer HP, Zwarts MJ. The "split hand" phenomenon. Evidence of a spinal origin. Neurology 2003;61:1619-20.

How to cite this article: Benny R, Shetty K. The split hand sign. Ann Indian Acad Neurol 2012;15:175-6.

Received: 27-05-12 **Revised:** 01-06-12, **Accepted:** 03-06-12

Source of Support: Nil, **Conflict of Interest:** Nil