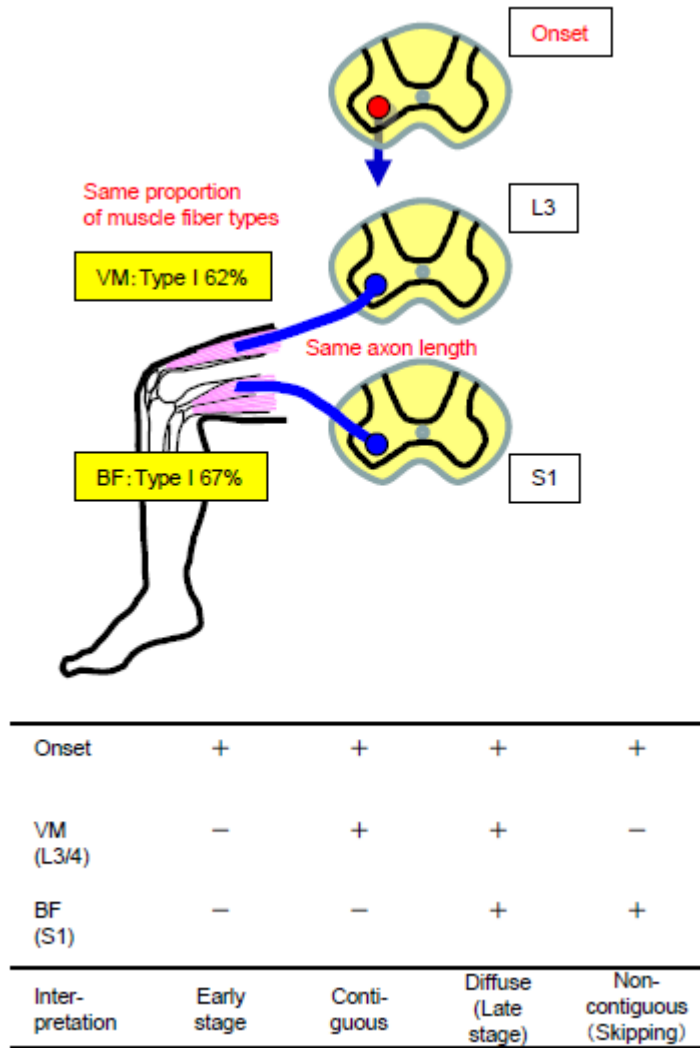


SUPPLEMENTARY MATERIAL

Supplementary Figure 1.

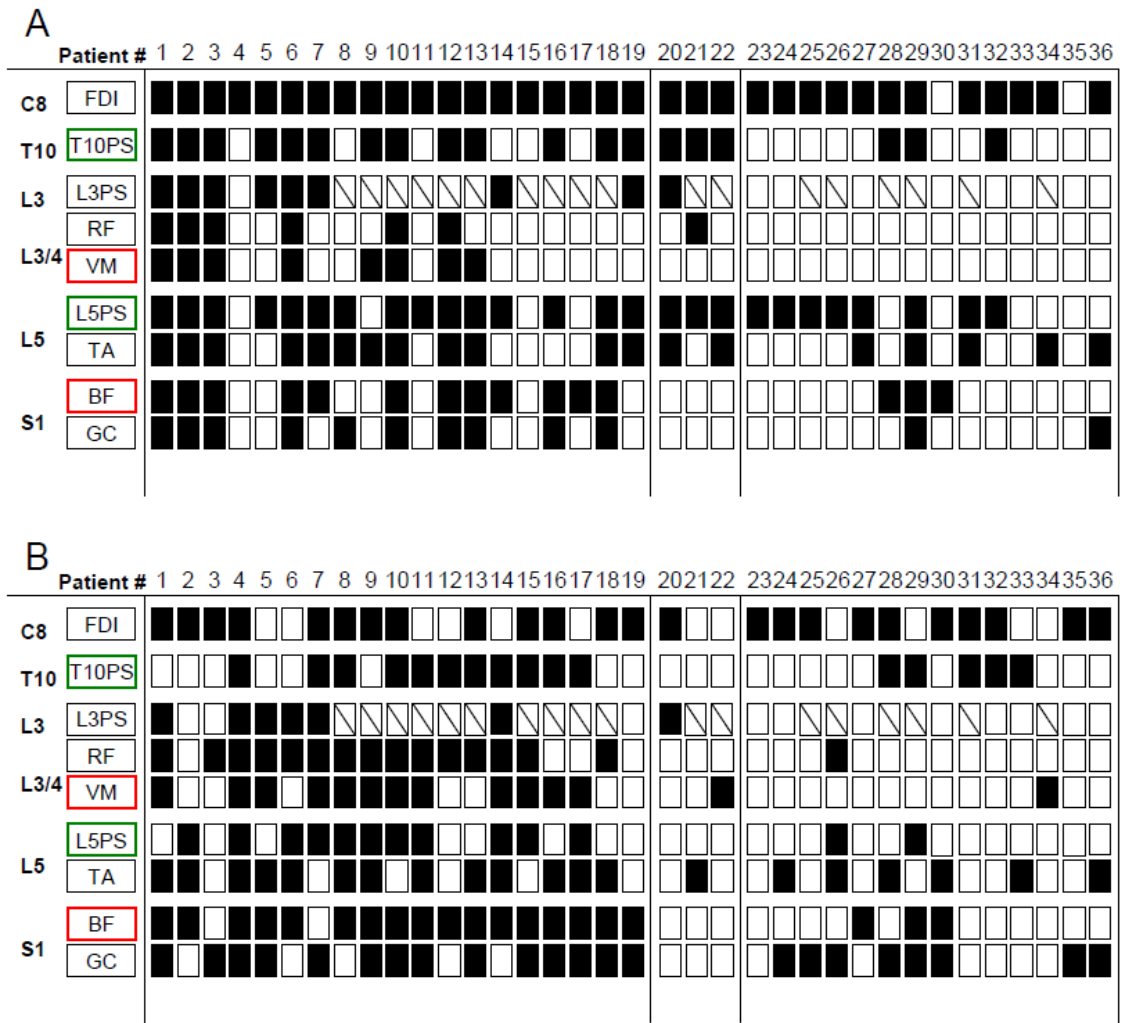
Title. Schema for evaluation of lesion spreading pattern by using the paired muscles.



Legend. The proportion of muscle fibre types and motor axon lengths are both related to the motoneuronal vulnerability in ALS. Two muscles, VM and BF, have almost the same proportion of muscle fibre types and motor axon lengths. A difference in the presence (+) or absence (-) of spontaneous EMG activities between these paired muscles thus cannot be explained by the difference in motoneuronal vulnerability. Therefore, examining pairs of muscles like these enables the reliable evaluation of the pattern of lesion spreading.

Supplementary Figure 2.

Title. Distribution patterns of fibrillation potentials, positive sharp waves and fasciculation potentials in all patients.

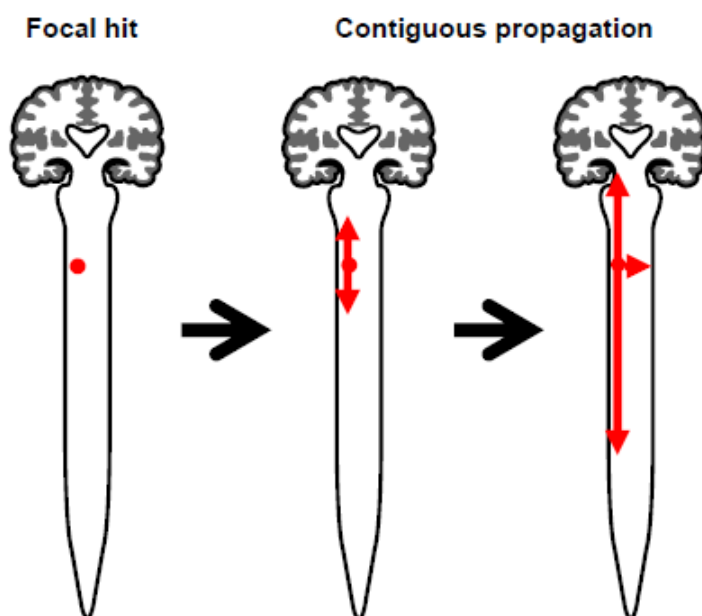


Legend. A. Distribution patterns of fibrillation potentials and positive sharp waves. B. Distribution patterns of fasciculation potentials. Patients #1-36 correspond to those in figure 1. Closed squares: abnormal spontaneous EMG activity present. Open squares: abnormal spontaneous EMG activity absent. Squares with oblique line: data not available.

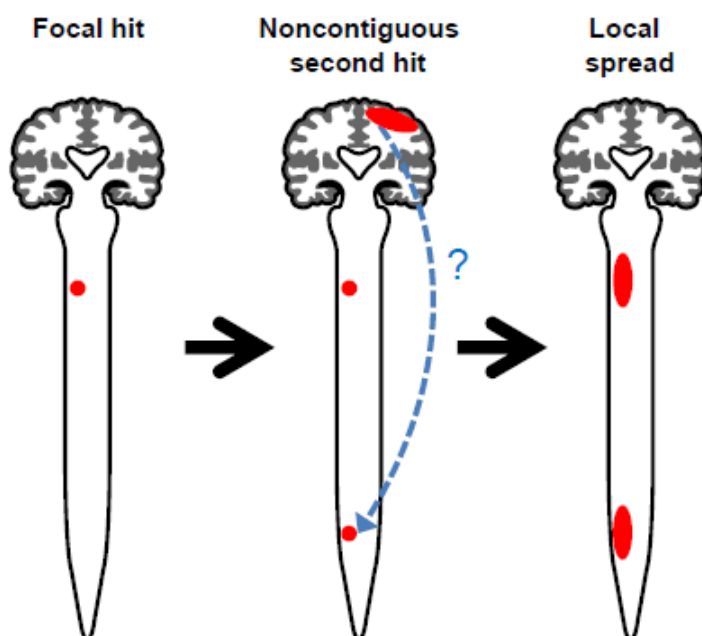
Supplementary Figure 3.

Title. Regional spread mechanisms of ALS lesion from cervical to lumbosacral cord.

A) "Single seed and simple propagation" hypothesis



B) Multifocal hits and local spread



Supplementary Table 1.

Title. The clinical UMN signs of the 14 patients who showed “noncontiguous pattern” at nEMG examination.

Patient number	23	24	25	26	27	28	29	30	31	32	33	34	35	36
BB (C5/6)	+/-	+/-	+++	+	++	+	++	+/-	++	+/-	+	+++	+	++
BR (C5/6)	+/-	+/-	+	++	++	++	++	+/-	+++	+/-	+	+++	+	++
TB (C6/7)	+/-	+/-	+	+	++	+	++	+/-	++	+/-	++	+++	+	++
FFR	-	-	+	+	-	-	+	-	+	-	+	+	-	+
PTR (L3/4)	++	+/-	+++	+++	++	++	++	+	++	++	+/-	+++	+	++
ATR (S1)	+	+/-	+++	++	++	++	++	+	++	++	+/-	+++	+	++
Plantar	-	-	-	-	+	-	+	-	-	-	-	+	+	+

BB: biceps brachii tendon reflex; BR: brachioradialis tendon reflex; TB: triceps brachii tendon reflex; FFR: finger flexion reflex; PTR: patella tendon reflex; ATR: Achilles tendon reflex; Plantar: abnormal planter reflex.