excellent technical assistance. This work was supported by a grant-in-aid for scientific research from the Ministry of Education, Culture, Sports, Science and Technology (14570957) and a research grant from the Zikei Institute of Psychiatry.

REFERENCES

1. Nalini A, Thennarasu K, Gourie-Devi M, Shenoy S, Kulshreshtha D. Clinical characteristics and survival pattern of 1,153 patients with amyotrophic lateral sclerosis: experience over 30 years from India. *J Neurol Sci* 2008; **272**: 60–70.
2. Ravits J, Laurie P, Fan Y, Moore DH. Implications of ALS focality: rostral-caudal distribution of lower motor neuron loss postmortem. *Neurology* 2007; **68**: 1576–1582.
3. Ince PG. Neuropathology. In: Brown RJ, Meininger V, Swash M, eds. *Amyotrophic Laeral Sclerosis*. London: Martin Dunitz, 2000; 83–112.
4. Patrikios JS. *Contribution À l'Étude Des Formes Cliniques Et De l'Anatomie Pathologique De La Sclérose Latérale Amyotrophique*. Paris University, 1918.
5. Bonduelle M. Amyotrophic lateral sclerosis. In: Vinken PJ, Bruyn GW, eds. *Handbook of Clinical Neurology*. 22. Amsterdam: North-Holland, 1975; 281–338.
6. Cappellari A, Ciammola A, Silani V. The pseudopolyneuritic form of amyotrophic lateral sclerosis (Patrikios' disease). *Electromyogr Clin Neurophysiol* 2008; **48**: 75–81.
7. Guidetti D, Bondavalli M, Sabadini R *et al.* Epidemiological survey of amyotrophic lateral sclerosis in the province of Reggio Emilia, Italy: influence of environmental exposure to lead. *Neuroepidemiology* 1996; **15**: 301–312.
8. Mortara P, Bardelli D, Leone M, Schiffer D. Prognosis and clinical varieties of ALS disease. *Ital J Neurol Sci* 1981; **2**: 237–242.
9. Salemi G, Fierro B, Arcara A, Cassata M, Castiglione MG, Savettieri G. Amyotrophic lateral sclerosis in Palermo, Italy: an epidemiological study. *Ital J Neurol Sci* 1989; **10**: 505–509.
10. Wijesekera LC, Mathers S, Talman P *et al.* Natural history and clinical features of the flail arm and flail leg ALS variants. *Neurology* 2009; **72**: 1087–1094.
11. Nishigaki S, Ando K, Nagata Y, Hirose K. Pseudopolyneuritic form of amyotrophic lateral sclerosis. *Rinsho Shinkeigaku* 1973; **13**: 377–384.
12. Terao S, Sobue G, Hashizume Y, Mitsuma T, Takahashi A. Disease-specific patterns of neuronal loss in the spinal ventral horn in amyotrophic lateral sclerosis, multiple system atrophy and X-linked recessive bulbospinal neuronopathy, with special reference to the loss of small neurons in the intermediate zone. *J Neurol* 1994; **241**: 196–203.
13. Tsuchiya K, Shintani S, Kikuchi M *et al.* Sporadic amyotrophic lateral sclerosis of long duration mimicking spinal progressive muscular atrophy: a clinicopathological study. *J Neurol Sci* 1999; **162**: 174–178.
14. Hasegawa M, Arai T, Nonaka T *et al.* Phosphorylated TDP-43 in frontotemporal lobar degeneration and amyotrophic lateral sclerosis. *Ann Neurol* 2008; **64**: 60–70.
15. Neumann M, Rademakers R, Roeber S, Baker M, Kretzschmar HA, Mackenzie IR. A new subtype of frontotemporal lobar degeneration with FUS pathology. *Brain* 2009; **132**: 2922–2931.
16. Hayashi S, Sakurai A, Amari M, Okamoto K. Pathological study of the diffuse myelin pallor in the anterolateral columns of the spinal cord in amyotrophic lateral sclerosis. *J Neurol Sci* 2001; **188**: 3–7.
17. Nishihira Y, Tan CF, Hoshi Y *et al.* Sporadic amyotrophic lateral sclerosis of long duration is associated with relatively mild TDP-43 pathology. *Acta Neuropathol* 2009; **117**: 45–53.
18. Brownell B, Oppenheimer DR, Hughes JT. The central nervous system in motor neurone disease. *J Neurol Neurosurg Psychiatry* 1970; **33**: 338–357.
19. Ince PG, Evans J, Knopp M *et al.* Corticospinal tract degeneration in the progressive muscular atrophy variant of ALS. *Neurology* 2003; **60**: 1252–1258.
20. Sasaki S, Iwata M. Immunocytochemical and ultrastructural study of the motor cortex in patients with lower motor neuron disease. *Neurosci Lett* 2000; **281**: 45–48.
21. Tan CF, Eguchi H, Tagawa A *et al.* TDP-43 immunoreactivity in neuronal inclusions in familial amyotrophic lateral sclerosis with or without SOD1 gene mutation. *Acta Neuropathol* 2007; **113**: 535–542.
22. Mackenzie IR, Bigio EH, Ince PG *et al.* Pathological TDP-43 distinguishes sporadic amyotrophic lateral sclerosis from amyotrophic lateral sclerosis with SOD1 mutations. *Ann Neurol* 2007; **61**: 427–434.
23. Aoki M, Ogasawara M, Matsubara Y *et al.* Familial amyotrophic lateral sclerosis (ALS) in Japan associated with H46R mutation in Cu/Zn superoxide dismutase gene: a possible new subtype of familial ALS. *J Neurol Sci* 1994; **126**: 77–83.
24. Ohi T, Saita K, Takechi S *et al.* Clinical features and neuropathological findings of familial amyotrophic lateral sclerosis with a His46Arg mutation in Cu/Zn superoxide dismutase. *J Neurol Sci* 2002; **197**: 73–78.

25. Ohi T, Nabeshima K, Kato S, Yazawa S, Takechi S. Familial amyotrophic lateral sclerosis with His46Arg mutation in Cu/Zn superoxide dismutase presenting characteristic clinical features and Lewy body-like hyaline inclusions. *J Neurol Sci* 2004; **225**: 19–25.

26. Arisato T, Okubo R, Arata H *et al.* Clinical and pathological studies of familial amyotrophic lateral sclerosis (FALS) with SOD1 H46R mutation in large Japanese families. *Acta Neuropathol* 2003; **106**: 561–568.