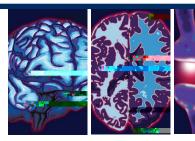
## **Case Report**



# Ultra-Structural Observation of the Focal Brain in a Patient with Motor Neuron Disease

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#### **ABSTRACT**

**Objectives:** To observe the ultra-structural changes of the brain tissue in a patient with motor neuron disease.

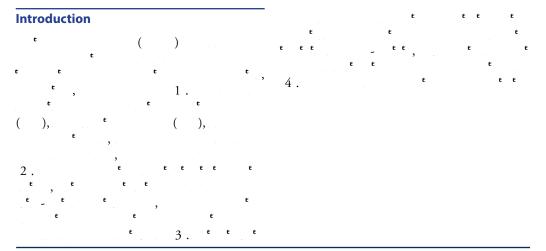
**Methods:** A stereotactic brain biopsy of the local lesions was performed based on the abnormal signals on magnetic resonance images, and the brain tissues were underwent light and electronic microscopic examination.

**Results:** Extensive extra-cellular edema and neuron necrosis were found in the internal capsule and thalamus. Metachromatic substances were found in the cytoplasma and were characterized by electronic dense granules, arranged in at plate layers or in at lines. Disorganized and degenerating mitochondria were observed in astrocytes and neurons.

**Conclusions:** Ultra-structural observation of the brain tissues might be helpful in understanding the central neuropathological changes in a patient with MND. Characteristic metachromatic substances might be found in the cytoplasma. Neurons in the brain of a patient with MND might undergo a process of degeneration.

#### Keywords

Ultra-structural observation, Stereotactic brain biopsy, Motor neuron disease, Diusion tensor imaging, Metachromatic substances



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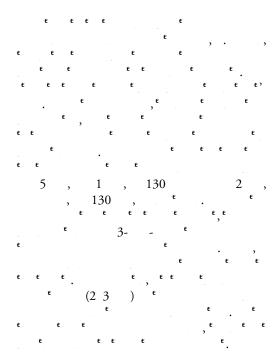
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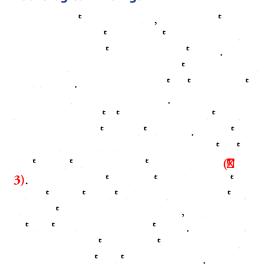
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**D**). **2C** 



#### **Pathological Findings**



#### Discussion

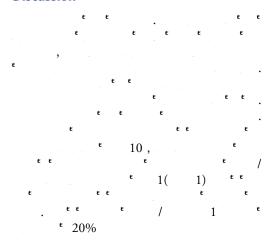


Figure 2: Light microscope examination of the internal capsule and the thalamus.

Extensive extra-cellular edema was observed in the internal capsule and the thalamus (A and C). Neuron necrosis and degenerations were also observed in the brain. The neurons were deformed and the Niss body disappeared. The nuclear structures of the neurons were ambiguous. Vacuoles as well as infiltrated neurogliocytes were observed in the cytoplasma (B and D).

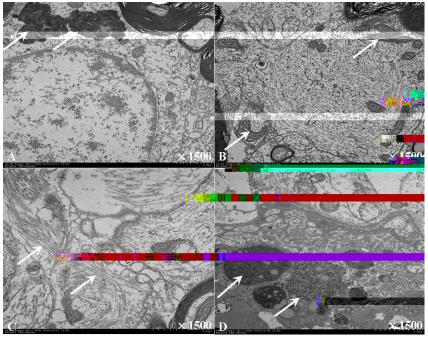


Figure 3: Electronic microscope examination of the internal capsule and the thalamus.

The electronic microscope demonstrated that the metachromatic substances deposited in the cytoplasma were characterized by electronic dense granules (A and D), arranged in flat plate layers (B) and in flat lines(C). Disorganized and degenerating mitochondria were observed in astrocytes and neurons.

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#### References

- Beghi E, Logroscino G, Chio A, et al.
   The epidemiology of als and the role of population-based registries. Biochim. Biophys. Acta 1762(1), 1150-1157 (2006).
- Chio A, Calvo A, Moglia C, et al. Phenotypic heterogeneity of amyotrophic lateral sclerosis: A population based study. J. Neurol. Neurosurg. Psychiatry 82 (1), 740-746 (2011).
- 3. Shen D, Cui L, Cui B, et al. A systematic review and meta-analysis of the functional mri investigation of motor neuron disease. Front. Neurol. 6 (1), 246 (2015).
- Fuchino Y, Nagao M, Katura T, et al. High cognitive function of an als patient in the totally locked-in state. Neurosci. Lett 435 (1), 85-89 (2008).
- Thorpe JW, Moseley IF, Hawkes CH, et al. Brain and spinal cord mri in motor neuron disease. J Neurol. Neurosurg. Psychiatry 61(1), 314-317 (1996).
- Garbuzova-Davis S, Haller E, Saporta S, et al. Ultrastructure of blood-brain barrier and blood-spinal cord barrier in sod1 mice modeling als. Brain. Res 1157(1), 126-137 (2007).
- 7. Konagaya M, Sakai M, Iida M, et al. [an autopsied case of dominantly a ecting upper motor neuron with atrophy of the

- frontal and temporal lobes--with special reference to primary lateral sclerosis]. *Rinsho. Shinkeigaku* 35(1), 384-390 (1995).
- Wu G, Wang L, Hong Z, et al. E ects of minimally invasive techniques for evacuation of hematoma in basal ganglia on cortical spinal tract from patients with spontaneous hemorrhage: Observed by di usion tensor imaging. Neurol. Res 32(1), 1103-1109 (2010).
- Wu G, Wang L, Liu J, et al. Minimally invasive procedures reduced the damages to motor function in patients with thalamic hematoma: Observed by motor evoked potential and di usion tensor imaging. J Stroke. Cerebrovasc. Dis. 22 (1), 232-240 (2013).
- Blumen SC, Inzelberg R, Nisipeanu P, et al. Aggressive familial als with unusual brain mri and a sod1 gene mutation. Amyotroph. Lateral. Scler 11(1), 228-231 (2010).
- Calvo A, Ilardi A, Moglia C, et al. An als case with a novel d90n-sod1 heterozygous missense mutation. Amyotroph. Lateral. Scler 13(1), 393-395(2012).
- Kim MJ, Bae JH, Kim JM, et al. Rapid progression of sporadic als in a patient carrying sod1 p.Gly13arg mutation. Exp. Neurobiol 25(1), 347-350 (2016).
- 13. Corcia P, Petiot P, Stevic Z, et al. Respiratory onset in an als family with I144f sod1

- mutation. J. Neurol. Neurosurg. Psychiatry 82(1), 747-749 (2011)
- 14. Shi YH, Zhao JH, Song JL, et al. [analysis on clinical features and functional mri of mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes]. Zhonghua. Yi. Xue. Za. Zhi 96(1), 2969-2972 (2016).
- Abe K, Yoshimura H, Tanaka H, et al.
   Comparison of conventional and di usion-weighted mri and proton mr spectroscopy in patients with mitochondrial encephalomyopathy, lactic acidosis, and stroke-like events. Neuroradiology 46(1), 113-117 (2004).
- Milo R, Miller A. Revised diagnostic criteria of multiple sclerosis. Autoimmun. Rev 13(1), 518-524 (2014).
- Kaunzner UW, Gauthier SA. Mri in the assessment and monitoring of multiple sclerosis: An update on best practice. *Ther.* Adv. Neurol. Disord 10(1), 247-261 (2017).
- Paskavitz JF, Anderson CA, Filley CM, et al. Acute arcuate fiber demyelinating encephalopathy following epstein-barr virus infection. Ann. Neurol 38(1), 127-131 (1995).
- 19. Friedman M. Acute di use demyelinating encephalopathy: Report of two cases. *Am. J. Pathol* 21(1) 519-526 (1945).