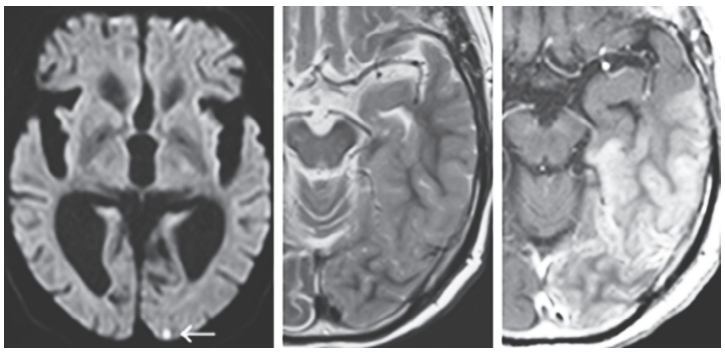
DSS-1

A left parietooccipital lesion of 73-year-old male with right hemianopsia and sensory aphasia

Manabu Hattori[1], Akio Akagi[2], Mari Yoshida[2]

[1] Department of Neurology, Daido Hospital, Nagoya, Japan,
[2] Institute for Medical Science of Aging, Aichi Medical University

Clinical History:
A 73-year-old right-handed male, who had been prescribed donepezil hydrochloride for treating dementia for the past four years, presented because of difficulty in seeing the right visual field.
Consciousness was clear, and there was no problem in verbal understanding and speech. Visual field test showed right hemianopsia. There was no paralysis of the limbs, but walking was unstable. All tendon reflexes disappeared. Orthostatic hypotension was present.
Magnetic resonance imaging (MRI) at the time of admission showed a small high-signal lesion in the diffusion-weighted image in the left occipital lobe, and antiplatelet drug was started as cerebral infarction.
Although sensory aphasia appeared on the fourth day of hospitalization, no corresponding infarct lesions were observed in MRI. Cerebrospinal fluid test showed protein elevation (85 mg/dL). Cerebral blood flow SPECT showed increased blood flow in the left temporal lobe, parietal lobe, and occipital lobe. MRI on the 18th day showed gadolinium enhancement on cerebral cortex and low-intensity signal in T2-weighted image in the cerebral white matter of the matching site.
Brain biopsy was performed from left parietal lobe on the 25th day of hospitalization for diagnosis.
Aphasia improved without treatment, but acute pancreatitis occurred on day 64. He died, and an autopsy was conducted.



MRI (Day 1) MRI (Day 18)
DWI T2WI Gd-enhanced

Presented samples: Hematoxylin-eosin staining of the left parietooccipital lobe lesion.

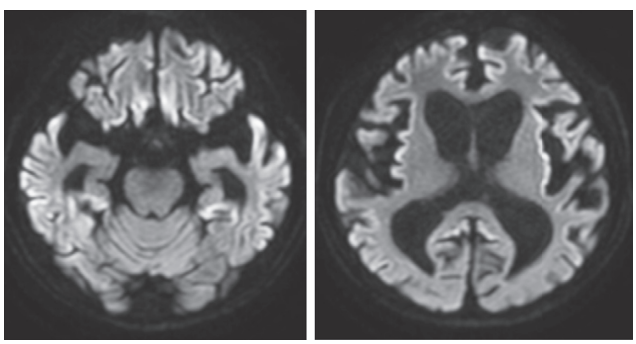
DSS-2

An 85-year-old female with advanced dementia progressing over the course of 5 years

Kanako Komatsu[1], Saori Onuma[2], Tadashi Tsukamoto[3], Kazuko Hasegawa[2], Hidehiro Mizusawa[3], Shigeo Murayama[4], Yuko Saito[1]

[1] Department of Pathology and Laboratory Medicine, National Center Hospital, National Center of Neurology ang Psychiatry,
[2] Department of Neurology, National Hospital Organization Sagamihara National Hospital,
[3] Department of Neurology, National Center Hospital, National Center of Neurology ang Psychiatry,
[4] Brain Bank for Aging Research, Department of Neuropathology, Department of Neurology Tokyo Metropolitan Geriatric Hospital and Institute of Gerontology

Case: 85-year-old female
Chief complaint: Memory loss
Present illness:
At 81 years of age, the patient presented with mild forgetfulness and numbness in the lower right side of the body. She had difficulty with language comprehension and expression. Her scores on the Mini Mental State Examination (MMSE) and the Frontal Assessment Battery (FAB) were 23/30 and 6/18, respectively. Diffusion-weighted magnetic resonance imaging (DW-MRI) revealed high intensity areas on both sides of the cerebral cortex.
Two months later, the numbness had spread to the right upper limb, and 6 months later, she started to wander around occasionally. By 82 years of age, the high intensity area identified on DWI had gradually expanded, and progression of cerebral atrophy was also observed. The same year, she experienced a left femoral head fracture and began to use a wheelchair. At 83 years of age, she was hospitalized. The conversation was not possible, and the MMSE could not be administered due to the progression of cognitive dysfunction. There were rare spontaneous movement. Episodic laughter stood out. The patient required the help of a caregiver at all times. Although spontaneous movement or speech were rare, but meal intake was orally possible with assistance. CSF examination showed that Tau protein was negative (460 pg/ml). Electroencephalography showed diffuse θ waves. At 84 years of age, limb myoclonus and startle reflex were observed, and the patient remained asleep. She continued meal intake, but often choked on her food.
She died at 85 years of age. Autopsy was performed 23 hours and 56 minutes after death.
Necropsy findings: The weight of the brain was 1186 g.
Virtual slides: Hematoxylin–eosin and immunohistochemically stained section of hippocampus
MRI (84 years of age): DWI



DSS-3

An autopsy case of 62-year-old male with FTLD – MND and parkinsonism

Hidetomo Tanaka, Ryoko Koike, Hitoshi Takahashi

Department of Pathology, Brain Research Institute, Niigata University

Clinical History:
A 58-year-old Japanese man developed stiffness in the right leg and this symptom progressed gradually. He also developed tremor in the bilateral arms and right leg. After 4 months, he visited a hospital. On examination, he showed spasticity in the right arm, and mild muscle weakness, with positive Babinski sign, in the right leg. Increased deep tendon reflexes were also present in the upper and lower extremities. About 6 months after onset, he showed mild muscle weakness in the bilateral upper and lower extremities. Electromyography (EMG) showed neurogenic changes in the tongue, sternocleidomastoid, femoral rectus, and anterior tibial muscles. Brain and spinal cord MRI revealed no apparent abnormalities. A tentative clinical diagnosis of motor neuron disease (MND) was made.
At the age of 60, he became unable to walk. At the age of 61, dysphagia appeared and progressed, necessitating the introduction of tube feeding. He developed dementia with a HDS-R* score of 13 (*the revised version of the Hasegawa dementia scale; a mental examination method commonly used in Japan that utilizes a scale of 0 to 30, where <21 represents dementia). Atrophy and fasciculation were evident in the tongue, and rigospasticity was present in the neck, with retrocollis. Vertical (both upper and lower) eye movement was mildly limited. He was treated with L-dopa without noticeable effect. At this point, an alternative clinical diagnosis of progressive supranuclear palsy (PSP) was considered. At the age of 62, he exhibited severe dementia with a HDS-R score of 4, and brain MRI revealed frontotemporal cerebral atrophy. The patient died of pneumonia, about 4 years after onset of the disease. There was no family history of neurological diseases.

Autopsy findings:
The brain weight was 1,090 g (fresh). There was atrophy of the frontotemporal lobes, which were more accentuated in the precentral gyrus

Material submitted:
Two H&E slides of motor cortex and spinal cord

Points for discussion:
1. Differential diagnosis
2. Useful immunohistochemical stains

DSS-4

A 67-year-old woman presented with progressive asymmetric weakness and numbness in all four extremities

Ryota Sato, Takashi Kanda

Department of Neurology and Clinical Neuroscience, Yamaguchi University Graduate School of Medicine

【Present illness】
A 67-year-old woman presented with a three-month history of progressive asymmetric weakness and numbness in her limbs.
【Blood exam】
WBC 21840 /μl, RBC 352x10^4 /μl, Hb 12.0 g/dl, Ht 34.9 %, PLT 10.3x10^4/μl
CRP 3.26 mg/dl, ESR1h 108 mm, HbA1c 5.7%, sIL-2R 906 U/ml
ANA (+), anti ds-DNA antibody (-), anti RNP antibody (+), anti SS-A antibody (+)
anti SS-B antibody (-), PR3-ANCA (-), MPO-ANCA (-)
【Cerebrospinal fluid】
normal
【Nerve conduction study】
asymmetric demyelinating sensory-motor polyneuropathy.
【Radiological findings】
Para-aortic nodes were enlarged, but any tumors didn't be found in contrast enhanced CT. There were no enlarged nerve roots or abnormal signal of brain and spinal cord in contrast enhanced MRI.

【Points of discussion】
1. Diagnosis
2. Mechanism of neuropathy

DSS-5

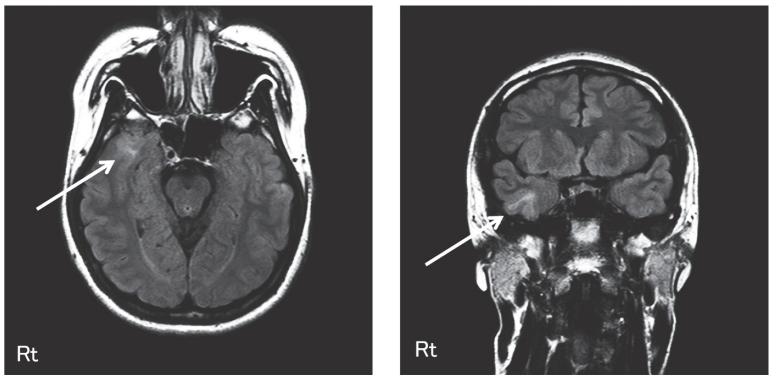
A right temporal lobe lesion of 18-year-old male with intractable epilepsy

Shoko Sadashima, Satoshi O. Suzuki, Toru Iwaki

Department of Neuropathology, Graduate School of Medical Sciences, Kyushu University

Case: 18 year old, male
Chief complaint: Complex partial seizure
Past medical history: None
Family history: No history of epilepsy
Present illness:
The case is an 18-year-old male with normal growth and development without history of febrile convulsion. At the age of 15, the patient developed a symptom of staring and behavioral arrest followed by thrashing the limbs. Electroencephalogram showed epileptogenic waves in the right frontotemporal region, and MRI revealed a lesion in the right temporal lobe. Since the epilepsy was drug-resistant, right lateral temporal lobectomy and amygdalohippocampectomy were performed.
Physical examinations: Height 169.0cm, Weight 99.0kg, BMI 34.7
BP 129/93mmHg, PR 78/min, RR 16/min, SpO_2 98%, room air
Neurological examinations: Consciousness, clear No neurological deficits
(Neuropsychological test: WAIS-III VIQ 58, PIQ 62, FIQ 57)

MRI: FLAIR (axial, coronal)



Presented samples: Hematoxylin-eosin staining of the right temporal lobe lesion.

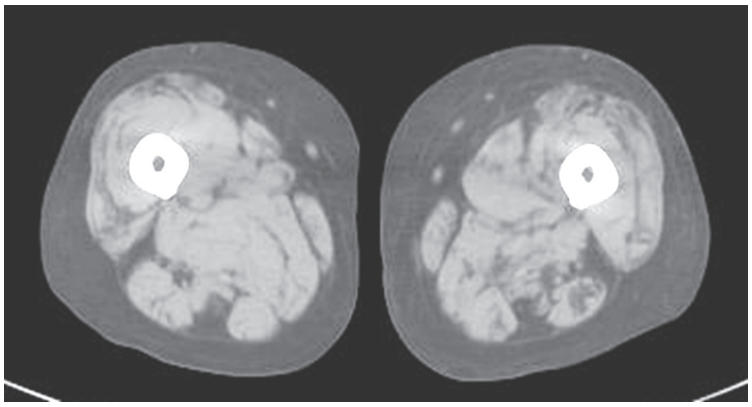
DSS-6

A 64-year -old man with a 17-year history of weakness in proximal limbs

Akira Arakawa[1], Masato Kadoya[2], Atsushi Unuma[1], Naohiro Uchio[1], Akatsuki Kubota[1], Jun Shimizu[1], Tatsushi Toda[1]

[1] Department of Neurology, Graduate School of Medicine, The University of Tokyo,
[2] Division of Neurology, Department of Internal Medicine, National Defense Medical College

Case: 64-year-old man
Chief complaint: weakness in proximal limbs
Past medical history: Taking simvastatin 5mg from 57 year old for dyslipidemia
Family history: No history of collagen disease
Present illness:
This patient had no muscle symptoms until he was found to have asymptomatic elevated serum CK (5000IU/L) at his age of 47. He was diagnosed as having polymyositis after the examinations including muscle biopsy. He was treated with oral prednisolone (5mg) for 10 years without subjective muscle symptoms, but with elevation of serum CK levels. At his age of 57, he gradually showed arthralgia and weakness in proximal muscles. His serum CK was 5651 IU/L. His second muscle biopsy showed mild myopathic changes with a small amount of inflammatory collections. He was treated with three courses of high dose intravenous methylprednisolone therapy followed by oral prednisolone (52mg), which were effective. At his ages of 59 and 60, he showed exacerbation of weakness associated with serum CK elevation, which was partially responded to immunosuppressive treatment. At his ages of 64, he showed gradual worsening of proximal muscle power with elevation of serum CK again (417 IU/L). He was hospitalized for re-examination of his muscle disease. He had symmetrical weakness in proximal limbs (4/5). He had no skin rash.



Muscle CT of proximal lower limbs (axial)

Presented samples: Hematoxylin-eosin staining of left deltoid muscle at his age of 64.