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NUMBNESS OF THE RIGHT HAND AS THE INITIAL SYMPTOM IN CREUTZFELDT-JAKOB DISEASE

(creutzfeldt-jakob disease/panencephalopathic type/sensory disturbance)

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A patient with Creutzfeldt-Jakob disease who showed numbness of the right hand as the initial symptom was reported. A 51-year-old man, a dairy farmer, complained of numbness of the right hand. All sensations, except for cortical senses, were reduced in the right hand. Nerve conduction velocities and cervical spine X-ray were normal. During the next three weeks, ataxic hemiparesis, cortical blindness and dementia appeared. He died eight months after the onset. Neuropathological findings revealed patchy and localized spongy states in the subcortical white matter in addition to severe neuronal loss with status spongiosus in the cerebral cortex (panencephalopathic type).

The major initial symptoms of Creutzfeldt-Jakob disease are psychiatric (more than 50%), visual and gait disturbances (1), and it is very rare for sensory symptoms to develop in this connection. We report here a case of the panencephalopathic type of Creutzfeldt-Jakob disease presenting numbness of the right hand as the chief complaint at the early stage.

CASE REPORT

A 51-year-old man, a dairy farmer, was well until the end of March of 1980, when numbness of the right hand occurred. The numbness was localized in the right hand distal to the wrist.

Two weeks after the appearance of the numbness, clumsiness of the right hand appeared, and after that he sometimes complained of mild blurred vision while reading the news paper. The clumsiness of the right hand worsened during the following two weeks, and he became unable to milk his cows. He visited the Shimane Medical University Hospital on April 25. On neurological examination, he was found to be alert and well-oriented. Sensory deficit for pinprick, touch and deep sensation was observed in the distal portion of the right hand, but graphesthesia was normal. The distribution of hypesthesia was not matched either with the peripheral innervation or the spinal segment. Mild motor weakness and incoordination were observed in the right hand. Deep tendon reflexes were normal, and no pathological reflexes were observed. An involuntary flexion of the right fingers occurred a few times per minute. He complained of blurred vision, but he could read letters of 5 × 5 mm in size at the distance of 30 cm with glasses and his visual field was normal by the confrontation test. Optic fundi, extraocular movement and pupils were normal. The motor and sensory conduction velocities of the median and ulnar nerves were normal, and needle electromyography revealed normal findings. The X-ray of the cervical spine and the brain CT scan were normal. General examinations revealed normal. The following week, gait disturbance and dysarthria appeared, and he was admitted to the hospital on May 9. After this, right sided ataxic hemiparesis occurred, blurred vision became worse, and he deteriorated rapidly. The sensory symptoms remained unchanged until this time. The electroencephalogram (EEG) upon admission revealed a diffuse high voltage delta wave without paroxysmal rhythm. Laboratory data of the blood, urine and cerebrospinal fluid were normal. His contact with the external environment diminished day by day. Complete urinary retention appeared, and cystometry revealed atonic bladder. On around May 19, vertical gaze palsy was noticed, but the oculocepharic reflex was normal. He became immobile, with severe flexion rigidity of all four extremities, during the next three weeks. Myoclonic movements of the extremities increased markedly. The EEG at this time showed diffuse typical periodic synchronized discharge (PSD). From the end of July, muscle atrophy of the extremities with fasciculation appeared and progressed rapidly, while myoclonus gradually disappeared. The EMG and muscle biopsy at this time revealed a neurogenic pattern.

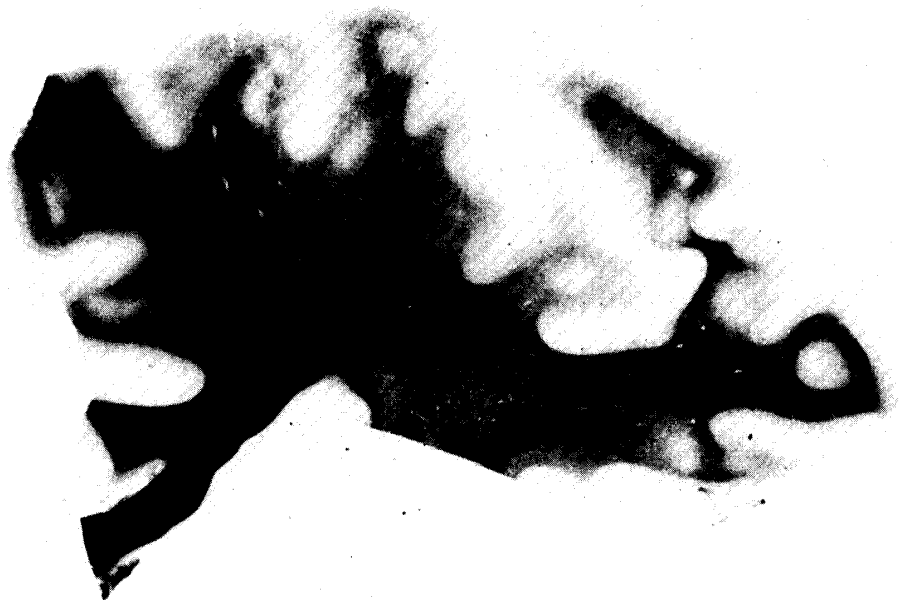

Fig.1. Klüver-Barrera's staining of the left parietal lobe shows marked cortical atrophy and patchy demarcated subcortical demyelinations.

The PSD decreased gradually and EEG became flat in October. The CT scan revealed marked diffuse cerebral cortical atrophy and ventricular dilatation. He died of respiratory complications on December 5.

Neuropathological Findings (Autopsy was permitted only for the brain)

Macroscopic Findings

The brain was severely atrophic with prefixed weight of 1120g. In a 1 cm coronary slice of the cerebral hemispheres, there was moderate enlargement of the ventricules and marked atrophy of the cerebral cortex, especially in the frontal and occipital lobes. The basal ganglia and thalamus were slightly atrophic. The cerebellum was also atrophic, but the brainstem was normal on section.

Microscopic Findings

In the cerebral cortex, abnormalities were most conspicuous in the frontal and occipital lobes, where there was marked status spongiosus and a moderate proliferation of fibrous astrocytes without microglial reaction. There was an almost complete fallout of the nerve cells. Patchy localized spongy states were also observed in the subcortical white matter with sparing of the U-fiber (Fig.1). The spongy state was also observed in globus

pallidus and corpus striatum. Moderate fallout of the neurons was observed in the thalamus. There were no kuru-like plaques. In the brainstem, the number of neurons was moderately decreased in the superior colliculus, and the medial lemniscus and pyramidal tract were slightly pale on myelin staining.
In the cerebellum, severe fallout of the Purkinje's cells associated with proliferation of the Bergmann's glia and a mild spongy state was observed in the vermis and hemispheres. Moderate fallout of the granule cells was also observed, but the subcortical white matter was spared.

DISCUSSION

Patchy and localized spongy stated in the subcortical white matter were conspicuous in this case. This white matter lesion may have been primary, so this case should be classified into the panencephalopathic type that has been reported in Japan (2). But the clinical picture of this case is not identical with the panencephalopathic type, which generally shows marked psychic symptoms as the initial sign. It is very rare that this disease begins with sensory symptoms. Heidenhein (3) has rported a case (his third case) presenting dysesthesia of the bilateral extremities, especially at night, as the initaial symptom. We could find only five cases in the Japanese literature showing sensory symptoms as the initial symptom (4). However, all of them complained of dysesthesia of the bilateral hands or feet, and there was no patient showing unilateral localized hypesthesia as this one did. Vanrossum (5) has reported that disturbance of sensation occurs more frequently in the legs than the arms, and this may be caused by a cortical or thalamic lesion. It is supposed that the sensory symptoms in this case is may be caused by the subcortical white matter lesion, because cortical sensation was spared and ataxic hemiparesis was observed. Ataxic hemiparesis from the lacune in the corona radiata has been reported as well as that from the pontine basis (6). The distribution of the senory disturbance is similar to that of a cortical localized lesion, but it can occur from a subcortical lesion. Therefore, it is possible to suppose that the disease developed from a localized frontoparietal subcortical lesion in this case.

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REFERENCES

1) May, W.W., (1968) Creutzfeldt-Jakob disease : 1)Survey of the literature and clinical diagnosis. Acta Neurol. Scand., 44, 1-32
2) Mizutani, T. (1981) Neuropathology of Creutzfeldt-Jakob disease in Japan with special reference to the panencephalopathic type. Acta Pathol. Jpn., 31, 903-922
3) Heidenheim, A. (1929) Klinische und anatomische Untersuchungen über eine eigenartige organische Erkrankung des Zentralennervensystems im Praesenium. Z. Gesm. Neurol. Pschiat., 118, 49-113
4) Akai, J., Kumagaya, N., Imai, S., Tsuchiya, Y., and Kato, Y. (1976) Ataxic form of Cretzfeldt-Jakob disease. A case report. Rinsho Shinkeigaku, 16, 124-131 (in Japanese)
5) Vanrossum, A. (1968) Spasatic pseudosclerosis. In: Handbook of Clinical Neurology. (Vinken P.J. and Bruyn G.W., eds.) Vol.6: pp.726-760, North-Holland, Amsterdam.
6) Iragui, V.J. and McCutchen, C.B. (1982) Capsular ataxic hemiparesis. Arch. Neurol., 39, 528-529