Case Report

Supranuclear Paralysis Preventing Lid Closure in Amyotrophic Lateral Sclerosis

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We report a case of amyotrophic lateral sclerosis (ALS) in which the ability to close the eyes on command or voluntarily, was lost in spite of retention of reflex activity. A electrophysiological study of the blink reflex revealed a prominent R1 component with normal latency, which confirmed that the blink reflex was exactly preserved and also suggested a hemispherical lesion. Postmortem examination disclosed prominent cortical and subcortical lesions of the precentral areas on both sides. These lesions seem to be very closely related to the inability to initiate lid closing.

Key Words: Lid apraxia, Blink reflex, Pyramidal sign, Motor impersistence

The inability to initiate eye closure on command or voluntarily in spite of the preservation of reflex activity, such as the corneal reflex, blinking or eye closure to threat, is a rare symptom. It has previously been reported in a few patients with pseudobulbar palsy (1-5), encephalitis periaxialis diffusa (6), chorea minor (7), Creutzfeldt-Jakob's disease (8) and amyotrophic lateral sclerosis (5, 9, 10). Of these cases, only a few were subjected to postmortem examination (4, 6, 8). The lesions in these autopsied cases, however, were not permitting a precise localisation.

In this study, detailed electrophysiological and postmortem examinations were carried out on a case of ALS who could not perform voluntary eyelid closure. The pathogenesis of the symptom was discussed.

CASE REPORT

A 42-year-old man with progressive muscular weakness and wasting, dysphagia, dysarthria and dyspnea was admitted to hospital in October 1985. His parents were not related and there was no family history of neurological disorders. In October 1984, he had muscle cramps of the legs and abdomen. Four months later he noticed progressive muscular weakness in all extremities, particularly on the left side. In August 1985, dysphonia and swallowing disturbance appeared, followed by dyspnea. He became unable to walk in October. On admission, physical examination disclosed no abnormality. Neurologically, his intellect was normal. There was slight limitation of abduction of eye movements. The eyes showed a saccadic pattern on smooth pursuit. Bell’s phenomenon was not seen. There were bilateral facial paresis, dysarthria, swallowing disturbance and slight muscle wasting of the tongue with mild fasciculation. Movement of the tongue was poor. He could open and close his mouth. There was diffuse muscular atrophy with fasciculation in all limbs, predominantly in the distal parts. There was marked spasticity in the legs and marked rigidospasticity in the arms, predominantly on the left side. The jaw, gag and deep tendon reflexes were markedly exaggerated but ankle jerks were decreased. There were no pathological reflexes. Ataxia, dys-
diadochokinesis, involuntary movement, sensory disturbance or sphincter disturbance was not seen. Serum CK levels were mildly elevated. Needle EMG of the tongue, face and limb muscles disclosed a marked denervation pattern at rest and long-duration high-amplitude Neuromuscular units during voluntary contraction. The results of other investigations, including lysosomal enzyme analysis of leukocytes, were all normal.

During hospitalization, his legs became flaccid and paralytic. The rigidospasticity and hyperreflexia in the arms remained unchanged. In January 1986, the jaw jerks became more hyperactive and jaw clonus appeared. At the same time, he showed an inability to close his eyelids voluntarily or on command although he showed blinking. This symptom persisted throughout his hospitalization. He showed corneal reflexes, spontaneous blinking, rapid eyelid closure to threat and a positive Myerson’s sign. He could not fix his gaze in any direction, and slight limitation of abduction was seen. The doll’s vestibulo-ocular reflex was intact. There were hyperactive orbicularis oculi reflexes. He slept with his eyes closed. He could open and close his mouth, and keep it opened or closed although the movement was slow due to weakness. He could not protrude his tongue from his mouth. The snout or palpebral reflex was not seen. In June he could not eat due to worsening of the swallowing disturbance. After then he had cyanotic spells because of weakness of the respiratory muscles. He died of respiratory failure and broncopneumonia on September 14, 1986.

**ELECTROPHYSIOLOGICAL EXAMINATION**

Examinations were performed in April 1986. The upper lid movements and surface electromyograms of the orbicularis oculi muscles on both sides, during blinking in response to repetitive gentle taps on the glabella with a small hammer and electrical stimuli to the supraorbital nerve at a frequency of one/sec, respectively, were recorded by the method previously described (11).

Persistent blinking, that is, a positive Myerson’s sign, in response to glabella taping was observed, and only a prominent R1 of the orbicularis oculi reflex was detected on both sides. The latency of R1 was 14.0 mseconds (normal; 14.4 ± 0.9 mseconds, n = 35). The upper lid also showed persistent blinking on electrical stimulation of the facial nerve, and only a prominent R1 of the blink reflex was seen on the stimulated side (Fig. 1). The latency was 10.8 mseconds on the right side and 10.7 mseconds on the left side (normal; 10.8 ± 0.6 mseconds, n = 34).

**POSTMORTEM EXAMINATION**

An autopsy was performed 10 hours after death. The lungs showed purulent bronchitis and pulmonary edema. The brain weighed 1,660 g. Macroscopically, there were no abnormalities in the cerebrum and cerebellum. The brainstem and spinal cord were reduced in bulk. Microscopically, the most prominent changes were observed in the cortex and subcortex of the precentral areas on both sides. Extensive loss of cortical neurons, including Betz cells, and marked gliosis were seen in the motor cortex (Fig. 2). In the subcortical areas under the motor cortex, there was marked gliosis. Mild diffuse gliosis was observed in the subcortical white matter of the frontal, except for the motor areas, parietal, occipital and superior temporal lobes on both sides. Loss of nerve fibers with gliosis was seen in the pyramidal tract, including the internal capsule, cerebral peduncle, and lateral and anterior pyramidal tracts of the entire spinal cord. Neuronal loss in the oculomotor, abducent, trochlear, facial and
Fig. 2. Pathological findings in the deep layer the motor cortex, Holzer, × 285 (A) and the subcortical area, Klüver-Barrera, × 285 (B). Extensive loss of cortical neurons, including Betz cells, and marked gliosis were present in the motor cortex. In the subcortical areas there was a markedly reduced number of myelinated fibers.

hypoglossal nuclei was not evident, and there was no increase in the number of astrocytes in the vicinity of these nuclei. The rostral interstitial nuclei of the medial longitudinal fasciculus and the parapontine reticular formation showed no abnormality. Foci of mildly increased glia were detected in the putamen, globus pallidus, lateral part of the thalamus and corpus callosum. The other basal ganglia, including the corpus Luysii, cerebellar nuclei, substantia nigra, inferior olivary nucleus, red nucleus and superior collicus, were intact. There was no abnormality in the Mynert nucleus. Neurofibrillary tangles, senile plaques or Lewy bodies were not seen. Motor neuron cells in the anterior column of the spinal cord were markedly decreased in number. There were some motor neurons containing a Bunina body. Axonal swelling was observed in the ventral root fobers. There was no abnormality in the posterior columns, spinocerebellar tracts or posterior roots. Brain maps showing the distribution and degree of the involvement are presented in Fig. 3.

Fig. 3. Brain maps showing the distribution and degree of the lesions.
DISCUSSION

The electrophysiological examinations of the blink reflex revealed only Rj component with normal latency. This supports that the oligosynaptic reflex activity as to eyelid closure was intact, electrophysiologically in addition to clinically. Our patient showed a prominent R1 and an absent R2 for the blink reflex. Rushworth (12) and Kimura (13) and Kimura et al. (14) reported that the first component was increased in amplitude and R2 was small or absent on the side of a hemispheric lesion. Kimura et al. (14) also reported that patients with abnormal blink reflexes had lesions in the inferior Rolandic area. The findings as to the blink reflex in our patient may reflect hemispheric lesions, particularly in the inferior Rolandic area.

The symptom of an inability to close the eyes, with retention of reflex activity, was reported in patients with pseudobulbar palsy about 100 years ago (1, 2). Postmortem examination, however, has been carried out on only a few cases showing the symptom. Schilder’s case of encephalitis periaxialis diffusa showed extensive demyelination in both hemispheres, but more prominently on the left side (6). Schilder (6) regarded the symptom as form of apraxia due to damage to the frontal regions and corpus callosum. Alajouanine and Thurel (4) summarized their own and previously described cases showing the symptom, and reported that bilateral cortical infarcts of the Rolandic operculum were the common pathological finding. Russel (8) described three patients with Creutzfeldt-Jakob’s disease. Pathological examination revealed extensive neuronal loss, and a spongy change with a glial reaction of the whole cortex, extensive pyramidal tract degeneration and myelin loss. He attached much importance to lesions in the cortex, rather than those in the descending motor tracts, for the pathogenesis of the symptom. In our case, the most prominent lesions were in the cortex and subcortex of the precentral areas on both sides, although mild involvement was observed in the subcortical white matter, corpus callosum and a part of the basal ganglia. Lesions of the precentral region are considered to play an important role in production of the symptom.

On the other hand, since the Lewandrowsky’s report, it has been well-known that patients with acute hemispheric lesions, such as after a stroke, are unable to keep their eyes closed (15-17). These patients also frequently had the following symptoms: inability to keep the mouth open and the tongue protruded, and to maintain a gaze, which were observed in our patient. Lewandrowsky (15) used the term “lid apraxia” for the lid symptom, and Fischer (16) used the term “motor impersistence” for all the symptoms. Some authors (18, 19) considered these symptoms to be a compulsive reaction. Unilateral hemispheric damage, especially on the right side, is thought to cause these symptoms. However, there have been no detailed pathological investigations. Even if symptoms or signs of only unilateral hemispheric damage are observed, clinically, bilateral hemispheric involvement cannot be ruled out, pathologically (4). In many patients, these symptoms were recognized in the acute phase of a unilateral hemispheric disorder (17), in which the damage is spread widely over the unilateral hemisphere. De Renzi et al. (17) reported that three of 19 patients with right-sided damage were also completely unable to initiate lid closure. Fischer (16) considered that the two symptoms, a complete inability to initiate lid closure and an inability to keep the lids closed, were different degrees of the same phenomenon. It is uncertain whether these symptoms are different or not. To clarify this problem, however, electrophysiological and pathological investigations on such patients are needed.

In addition to supranuclear palsy preventing lid closure, rigidospasticity, spasmodic fixation and limitation of the ocular movements were observed in our patient. Esteban et al. (10) reported two cases of ALS showing paralysis preventing voluntary eyelid occlusion, who also showed impaired ocular movements and spasmodic fixation, and one of whom showed cervical and limb rigidity. These symptoms seem to result from lesions close to those which cause supranuclear palsy preventing lid closure, because they frequently occur together, although they are a rare manifestation in ALS.

REFERENCES

Lid Closure Inability in ALS