ground of the elaborate and athetoid phenomenology alone.

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The arm-mouth reflex in a patient with amyotrophic lateral sclerosis

Sir: Numerous pathological or primitive reflexes may appear in a variety of diseases of the central nervous system including amyotrophic lateral sclerosis. We have observed reflex opening of the mouth upon passive flexion of the forearm in a patient with an advanced state of amyotrophic lateral sclerosis. To our knowledge, this reflex, which we have called arm-mouth reflex, has never been described previously.

A 57-year-old Japanese man noticed wasting and weakness of both hands in April 1981. During the next nine months progressive weakness of lower limbs and difficulty in swallowing and speaking developed. The patient's history was otherwise normal. He was admitted to hospital on 11 January 1982. Findings on general physical examination were normal as was his mental status. Bilateral weakness was present mildly in facial muscles and moderately in bulbar muscles with dysphagia and dysphonia. Atrophy and fasciculations in the tongue were observed. Weakness was also present mildly in neck flexors, moderately in all limb girdle and proximal limb muscles and severely in bilateral hand muscles. Deep tendon reflexes were symmetrically hyperactive in association with Babinski's reflex. Jaw jerk was exaggerated and superficial abdominal reflexes were absent. Sensation and coordination were normal. Cerebrospinal fluid examination and radiographs of the skull and spine were normal. Electromyographic examination of the right biceps brachii, first dorsal interossei, and rectus femoris showed fibrillation potentials, positive sharp waves, an increase in potential amplitudes and polyphasic potentials with a decrease in the maximum interference pattern. Pulmonary function tests showed that his vital capacity was 49%. A diagnosis of amyotrophic lateral sclerosis was made. The patient became almost aphagic and apnoeic by June 1982 and required the use of a feeding tube and a respirator. Muscle weakness also progressed rapidly. Neurological examination in April 1983 showed that the patient was alert and oriented but was totally aphagic, aphonie, dysphagie and quadriplegie in extension. Supranuclear ophthalmoplegia and facial diplegia were noted. Forced crying was frequently seen. Deep tendon reflexes were symmetrically depressed but the sustained jaw clonus and head-retraction reflex were elicited. Tactile stimulation of the tongue resulted in reflex masticatory movement. At this clinical state, the arm-mouth reflex was observed and consisted of a wide opening of the mouth upon passive flexion of the forearm and could only be elicited by flexion of either forearm. The response movement was restricted to the perioral musculature and was constantly elicited by repetitive stimulation. The appearance, latency and duration of the response were absolutely dependent upon the strength of the stimulus (fig): faster and greater flexion of the forearm resulted in more prolonged and wider opening of the mouth at a shorter latency, whereas slower flexion of the forearm elicited the response less often. Flexions and extensions of other joints and sensory stimuli including pain and tendon tap of the triceps brachii did not elicit opening of the mouth.

The reflex opening of the mouth by the passive flexion of the forearm described here can be classified as a primitive brainstem reflex for the following reasons. Firstly, passive flexion of the forearm was the only stimulus that resulted in a stereotyped response of the perioral musculature at distinct latent periods following the stimulus. Secondly, the appearance, latency and duration of the response were dependent upon the strength of the stimulus. These physiological properties are in accordance with those of primitive brainstem reflexes. It has been demons-
trated electrophysiologically that corneomandibular and palmental reflexes are stimulus-strength-dependent. To our knowledge, the arm-mouth reflex has never been described previously, the most similar reflex being the hand-mouth reflex of Babkin. The hand-mouth reflex consists of opening of the mouth, flexion of the neck, and sometimes closing of the eyes and flexion of the forearm produced by pressure on the palm of the hand. In the present case, pressure on bilateral palms resulted in slight opening of the mouth but no other associated movements. The difference in the mode of stimulation and response between the two reflexes indicated that they are clinically distinguishable, although the physiological mechanisms of the two reflexes seem to be shared. The findings of prolonged opening of the mouth produced by sustained flexion of the forearm suggests that the afferent impulse of the arm-mouth reflex might originate from one of the slowly adapting mechanoreceptors. It has been reported that primitive reflexes have little clinical value because of their presence in a significant proportion of the healthy population. To date, however, we have not found the arm-mouth reflex in any normal adult. The appearance of the arm-mouth reflex in a patient with an advanced state of amyotrophic lateral sclerosis suggests that the reflex was related to the disease process and resulted from severe destruction of the upper motor neuron system innervating the perioral musculature. Accordingly, we consider that the arm-mouth reflex is a clinically valuable sign of severe destruction of the corticobulbar tract.

Fig. EMG recordings of arm-mouth reflex. Surface electrodes were placed on the chin and the right forearm (FA). Faster flexions (A and B) of the FA elicited reflex opening of the mouth (indicated by solid lines), whereas very slow flexion (C) of the FA did not result the response.

References

Letters

Triphasic spike-wave stupor in portal-systemic encephalopathy: a case report.

Sir: Lennox originally described prolonged stupor associated with bilaterally synchronous spike-wave discharges in the electroencephalogram (EEG). Though many authors described this condition as "petit mal status", Niedermeyer and Khalifeh proposed the term "ictal stupor", or "spike-wave stupor". Most of the cases of spike-wave stupor have been observed in epilepsy. A few cases have been reported as spike-wave stupor in non-epileptic patients. We report a case observed in portal-systemic encephalopathy.

A 59-year-old woman was admitted to Kagawa Central Hospital on 13 April 1983, because of bizarre abnormal behaviour. That morning she had been disoriented and mentally dull. Prior to this episode, a similar episode which lasted 3 hours occurred in January 1983. During examination she was cooperative, but her attention span was short. She could give her name, the time and the place, her husband’s name, but not calculate "100 - 7".

There was amnesia about that morning’s events. She appeared to be in a slightly stuporous state. Neurological examination revealed hyperreflexia of extremities and flapping tremor. On admission, EEG examination was performed immediately and showed continuous bilaterally synchronous spike-wave discharges (fig) which took the wave form of blunt spike-wave complexes. This was not altered by photic stimulation, nor by opening and closing the eyes. Her plasma ammonia level was 188 µg/dl (normal; 30-86 µg/dl). During her admission, lactulose and kanamycin were orally administered. By the next day, her spike-wave stupor and flapping tremor had disappeared, although the ammonia level was still high (149 µg/dl). These high levels of ammonia remained high during admission. Her EEG recording in a non-stuporous state showed slow alpha activities mixed with sporadic theta waves.


Accepted 18 September 1984
The arm-mouth reflex in a patient with amyotrophic lateral sclerosis.
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J Neurol Neurosurg Psychiatry 1985 48: 385-386
doi: 10.1136/jnnp.48.4.385