

Letters

extradural haematoma, hypertension, uraemia or diabetes, with progression to dementia. Although incontinence occurred more often than with other forms of pre-senile dementia, attention to their anticonvulsant medication rarely influenced the frequency of enuresis or encopresis.

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Meige's syndrome associated with spasmodic dysphagia

Sir: In 1910, Meige described ten cases of a neurologic syndrome characterised by symmetrical dystonic spasms of the facial muscles, which he termed "spasm facial médian".¹ This syndrome was later called oral-facial dyskinesia,² Brueghel's syndrome,³ blepharospasm-oromandibular dystonia syndrome,³ Meige's syndrome or Meige's disease.⁴⁻¹⁵ Recently, we treated a patient with this syndrome who also had severe dysphagia. There are a few case reports of Meige's syndrome accompanied by dysphagia,^{2,3,11} but this is the first report studying the mechanism of dysphagia by electrophysiological methods.

In March 1982, a 69-year-old man presented with progressive difficulty in eye opening for 1 year and dysphagia for 6 months. Difficulty of eye opening was especially exacerbated by bright light and mental stress, and perioral spasmodic movement was also noted during voluntary efforts to open eyes. There was no history of drug intake, psychiatric illness, or similar illness in the family. Neurological examination revealed sustained spasms of orbicularis oculi and oris, and marked dysphagia. Bilateral blepharospasm was much worse when he tried to open his eyes, especially following maximum eye closure. Voluntary mouth opening or closure did not influence the orbicularis oculi. Dysphagia was characterised by simultaneous spasmodic contractions of sternocleidomastoid and pharyngeal muscles,

which occurred each time he attempted to swallow. The thyroid cartilage did not elevate in spite of an effort to swallow. Taking a bolus of either fluid or solid food required several deglutition movements, and it took a few hours to finish a meal. Movement of the soft palate was normal and symmetric. The gag reflex was present bilaterally. There was no atrophy or fasciculation in

the tongue. Protrusion and rapid movement of the tongue were normal. The jaw jerk was normal. Phonation and speech were normal. Examination of the neck, limbs and gait was normal. Electroencephalogram, skull radiographs and computed tomography of the brain were normal.

Surface electromyographic (EMG) study

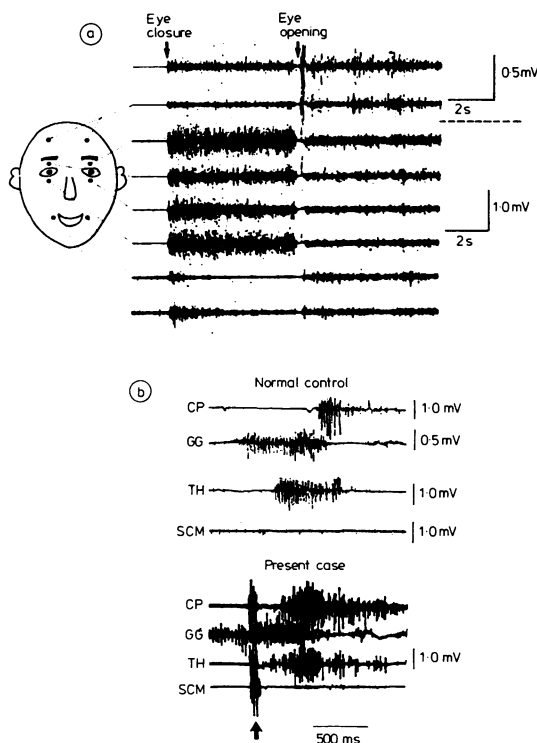


Fig (a) Polygraphic study of facial surface EMG. Surface electrodes were placed on frontal, orbicularis oculi (upper and lower eye lids) and orbicularis oris muscles bilaterally. Recording was performed by bipolar derivation. (b) Needle EMG study during swallowing in a normal person (upper tracings) and in the present patient (lower tracings). CP; cricopharyngeal, GG; genioglossus, TH; thyrohyoid and SCM; sternocleidomastoid muscle, respectively. In a normal person, GG, TH and CP contract in this order, while SCM remains silent during swallowing. In the patient, however, all these muscles including SCM show marked contractions simultaneously at moment of swallowing (solid arrow), followed by sustained irregular contractions in the CP, GG and TH.

showed that orbicularis oculi and oris were continuously active even at rest. When the patient opened his eyes, after forced eye closure, the frontalis strongly contracted and orbicularis oculi became silent, but only for a short period of time (approximately 400ms). After this, orbicularis oculi and oris exhibited continuous bilateral contractions in spite of the continuous effort to open his eyes as can be seen in the increased EMG discharge in the frontal muscles (fig 1A). Needle electrodes were inserted into the right palpebral levator and orbicularis oculi muscles. Orbicularis oculi showed continuous contraction even at rest. During a voluntary effort to open his eyes, orbicularis oculi showed sustained EMG discharge although to lesser degree as compared to the resting condition.

EMG studies of deglutition muscles was done with special hooked-wire electrodes inserted into the right cricopharyngeal (CP), genioglossus (GG), thyrohyoid (TH) and sternocleidomastoid (SCM) muscles (according to the methods of Shin¹⁶ and Hirose and Gay¹⁷). The mechanism of normal swallowing consists of three stages; the first stage (oral or voluntary stage), the second stage (pharyngeal, reflex or involuntary stage) and the third stage (esophageal or involuntary stage).¹⁶ In the normal person, each pharyngeal muscle contracts in a certain sequence of order to produce smooth swallowing and SCM is not involved (fig 1B). In the present case, the first stage (oral stage) of swallowing was normal. However, while the alimentary bolus passed through the upper pharyngeal portion, all pharyngeal and SCM muscles showed simultaneous strong contractions (solid arrow in the lower tracing of fig 1B), making swallowing impossible. Therefore, dysphagia in the present case is considered to be due to disorganised spasmodic contractions of deglutition muscles (spasmodic dysphagia). To our knowledge, there has been no previous report of similar case.

Haloperidol was found, by a double blind cross-over study, to be effective for both bilateral blepharospasm and spasmodic dysphagia. Haloperidol was administered with an initial dose of 2.25mg/day and was gradually increased. Its effectiveness appeared to be dose-dependent. The time spent for finishing a meal shortened from a few hours to 30 minutes. The good response to haloperidol (maintenance dose 3–4mg/day) in the present case has been observed for 9 months.

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