The Journal of Laryngology and Otology
August 1993, Vol. **107**, pp. 746–747

Palato-pharyngo-laryngeal myoclonus: an unusual cause of dysphagia and dysarthria

A. J. DRYSDALE, F.R.C.S., J. ANSELL, M.A., F.R.C.S., J. ADELEY, B.Sc., M.B., B.S. (London)

Abstract
We describe a case of palato-pharyngo-laryngeal myoclonus, an unusual variant of palatal myoclonus, which presented with dysphagia and dysarthria. The aetiology and presenting features of myoclonus are discussed. Various treatment options are considered.

Key words: Myoclonus; Palatal muscles; Deglutition disorders; Dysarthria

Introduction

Palatal myoclonus is a rare condition characterized by an involuntary, rhythmic and usually bilateral contraction of the palatal musculature. It usually presents with tinnitus which may be objective. It may be associated with myoclonus of other muscle groups including the pharyngeal, laryngeal, diaphragmatic, facial and ocular muscles.

We present a rare case of palato-pharyngo-laryngeal myoclonus presenting with dysarthria, dysphagia and aspiration, but no tinnitus. Videofluoroscopy was helpful in the diagnosis and in planning successful treatment using a technique of speech therapy.

Case report

A 78-year-old man presented with an 18-month history of difficulty in swallowing, for both solids and liquids, which started after a 'blackout'. He felt that food 'went down the wrong way' causing him to cough. He also noticed a voice change, with difficulty in articulating words, and he was aware of a twitching in his neck. He had suffered a cerebro-vascular accident (CVA) five years before, causing a mild residual right-sided weakness and mild dysarthria. At that time he was found to have hypertension, which had been adequately controlled since then.

On examination he had a bilateral, rhythmic, myoclonic movement of the soft palate and pharynx at a rate of 70/minute and an obvious rhythmic movement of the laryngeal cartilages visible externally. There was no objective tinnitus and, on further questioning, no subjective tinnitus. Neurological examination was otherwise normal apart from signs of the old CVA. There were no cerebellar signs.

A barium swallow showed free flow into the stomach, but a little difficulty in initiating swallowing and some aspiration into the trachea. On videofluoroscopy myoclonic movements of the palate, pharynx and larynx were clearly shown. There was aspiration of liquids into the trachea, even when taken in small quantities or thickened. With speech therapy he was able to learn a supraglottic swallowing technique which prevented him from aspirating liquids (Logemann, 1983). A repeat videoswallow after three months showed persistent myoclonus but no aspiration.

Discussion

Palatal myoclonus is a rare condition which was described by Politzer in his textbook over 100 years ago (Politzer, 1878). Spencer coined the term 'pharyngeal and laryngeal nystagmus' in 1886, because of the associated ocular movements, but the more appropriate description 'myoclonus' was popularized by Guillain (Guillain, 1938).

It is characterized by involuntary rhythmic contractions of the palatal musculature, usually bilaterally, at a rate varying between 40 and 240 per minute. These movements are persistent and unaffected by natural sleep, phonation, intravenous barbiturates, carotid sinus stimulation, or coma. It may be associated with myoclonus of other muscle groups including the pharyngeal, laryngeal, diaphragmatic, facial and ocular muscles (Hanson *et al.*, 1985). Most patients present with tinnitus, often described as a repetitive clicking, which is occasionally perceived by the examiner. When other muscle groups are involved symptoms include dysarthria, dysphagia, aspiration, irregular respirations and airway obstruction (Tolando *et al.*, 1984). Various aetiologies have been linked with the condition. The commonest precipitating event is a cerebrovascular accident. Others include trauma, multiple sclerosis, encephalitis, brain stem and cerebellar tumours, syphilis, malaria, vertebral artery aneurysms, and heredo-familial tremor (Tolando *et al.*, 1984). There is commonly a delay of weeks to months between the apparent cause and the development of myoclonus (Dubinsky and Hallett, 1988). A constant pathological finding is hypertrophic degeneration of the inferior olivary nucleus in the medulla oblongata, contralateral to the myoclonus in unilateral cases, but on either or both sides in bilateral cases. This degeneration may be secondary to primary lesions of pathways connecting the brain stem nuclei. Guillain and Mollaret proposed a triangular pathway connecting the dentate nucleus through the superior cerebellar peduncle to the contralateral red nucleus, from there to the contralateral inferior olivary nucleus via the central tegmental tract, then back to the dentate nucleus through the inferior cerebellar peduncle (Guillain and Mollaret, 1931). Lapresle (1986) supported Trelles' simpler pathway between the dentate nucleus and the contralateral inferior olivary nucleus via the red nucleus, since there was no evidence of a lesion associated with palatal myoclonus between the dentate nucleus and the inferior olivary nucleus along the inferior cerebellar peduncle.

There have been a number of suggestions as to how these extrapyramidal lesions can lead to myoclonus. It is possible that they result in loss of inhibition of lower motor neurones and cranial nerve motor nuclei, so causing rhythmic contractions of the muscles supplied by their neurones (Lapresle, 1986).

From the ENT Department, St George's Hospital, London SW17.
Accepted for publication: 9 March 1993.

There is no definitive treatment for palatal myoclonus. It is not usually influenced by drug treatment although a number have been tried including carbamazepine, 5-hydroxytryptophin, phenytoin, sedatives and antispasmodics, with occasional anecdotal reports of benefits (Castro-Caldas, 1981; Fitzgerald, 1984). A number of surgical approaches have been attempted to relieve the troublesome objective tinnitus but none has proved consistently helpful (Parnes, 1977; Hanson *et al.*, 1985). In myoclonus of other muscle groups speech therapy and physiotherapy may be usefully employed to avoid aspiration (Logemann, 1983). Palato-pharyngo-laryngeal myoclonus should be considered in patients presenting with dysphagia, dysarthria, or aspiration. The diagnosis is usually evident on clinical examination.

Acknowledgement

We would like to thank Mrs V. Moore-Gillon, F.R.C.S., consultant otolaryngologist, for permission to report on her patient and for her help and advice.

References

Dubinsky, R. M., Hallett, M. (1988) Palatal myoclonus and facial involvement in other types of myoclonus. In *Advances in Neurology*, vol. 49: *Facial dyskinesias*. (Jankovic, J., Tolosa, T., eds.), Raven Press, New York, p. 263–274.

Ferro, J. M., Castro-Caldas, A. (1981) Palatal myoclonus and carbamazepine. *Annals of Neurology* **10 (4):** 402.

Fitzgerald, D. C. (1984) Palatal myoclonus – case report. *Laryngoscope* **94:** 217–219.

Guillain, G. (1938) The syndrome of synchronous and rhythmic palato-pharyngo-laryngo-oculo-diaphragmatic myoclonus. *Proceedings of the Royal Society of Medicine* **31:** 1031–1038.

Guillain, G., Mollaret, P. (1931) Deux cas de myoclonies synchrones et rhythmees velo-pharyngo-laryngo-oculo-diaphragmatiques. Le probleme anatomique et physiopathologique de ce syndrome. *Review Neurologique* **2:** 545–566.

Hanson, B., Ficara, A., McQuade, M. (1985) Bilateral palatal myoclonus. *Oral Surgery, Oral Medicine, Oral Pathology* **59:** 479–481.

Lapresle, J. (1986) Palatal myoclonus. In *Advances in Neurology*, Vol. 43: *Myoclonus*. (Fahn, S., Marsden, C. D., Van Woert, M. H., eds.), Raven Press, New York, p. 265–272.

Logemann, J. A. (1983) Management of the patient with disordered oral feeding. In *Evaluation and Treatment of Swallowing Disorders*, College-Hill Press, Inc., San Diego, p. 138–139.

Parnes, S. M. (1977) Palatal myoclonus – a case study. *Transactions of the Pennsylvanian Academy of Ophthalmology and Otolaryngology* **30 (2):** 193–196.

Politzer, A. (1878) Translation 1926. In *Diseases of the Ear for Students and Practitioners*. 6th Edition. Bailliere, Tindall and Cox, London, p. 645–646.

Tolando, A. D., Porubsky, E. S., Coker, N. J., Adams, H. G. (1984) Velo-pharyngo-laryngeal myoclonus: evaluation of objective tinnitus and extrathoracic airway obstruction. *Laryngoscope* **94:** 691–695.

Address for correspondence:
Mr A. J. Drysdale, F.R.C.S.,
6 Kenwyn Road,
West Wimbledon,
London SW20 8TR.