Palato-pharyngo-laryngeal myoclonus ... an unusual cause of dysphagia

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Abstract

Dysphagia is a common problem in the elderly patient. Palato-pharyngo-laryngeal myoclonus, however, is a rare cause of this. We report a case of a 78-year-old man with dysphagia due to palato-pharyngo-laryngeal myoclonus that was ultimately managed conservatively with a good functional outcome.

Keywords: dysphagia, palato-pharyngo-laryngeal myoclonus, older people

Case report

A 78-year-old man required hospitalisation for features of congestive cardiac failure. He also gave a 3-year history of poor balance and 2 years of difficulty swallowing, with occasional coughing spells during eating. In addition, he noted 6 months of speech difficulty. He commented that ‘chewing’ all his food, including liquids, alleviated some of the swallowing and coughing symptoms.

His past medical history was positive for coronary artery disease, diabetes, hypertension, atrial fibrillation, hypercholesterolaemia, vertigo and cataracts.

Prior to admission, he was independent of basic activities of daily living, and managed his own finances and medications. He had some meal and housekeeping assistance. He was independently mobile with a walker or cane. Cognitively, he functioned well, earning his BA in history at the age of 62.

Relevant clinical features on examination were an obvious involuntary myoclonus of the right chin, and palatal myoclonus on the same side, but his lips and tongue were not involved. He had slight dysarthria. He denied an audible click. Nystagmus was absent. His myoclonus continued while asleep [{Supplementary data available in Age and Ageing online, Videos S1 and S2}]. The MMSE score was 27/30.

A modified barium swallow showed premature spillage to the valleculae and pyriform sinuses, visible penetration of all substances apart from puree, and aspiration with thin and level 1 liquids. He was unable to protect his airway despite an intact cough response.

Computerised tomography (CT) head scan showed calcification and tortuosity of the intracranial vasculature, mild dolichoectasia (dilation and tortuosity) of mid and superior basilar arteries, cavum septum pellucidum and cavum vergae. There were mild microangiopathic changes, but no cortical infarcts or intracranial haemorrhage.

Magnetic resonance imaging (MRI) of the brain confirmed the CT findings and showed enlargement and hyperintense signal within the olivary nuclei consistent with hypertrophic olivary degeneration. Figure 1 shows his MRI axial T2 weighted image through the medulla. There was no evidence of a neurodegenerative disease.

Treatment for the myoclonus was attempted with clonazepam, but he became quite sedated and confused, with no apparent improvement in the myoclonic features.

Consideration of botulinum toxin therapy prompted an ENT consultation. Laryngoscopy showed myoclonus of larynx and vocal cords, pharyngeal constrictors and lateral pharyngeal wall. Voluntary control of the vocal cords was maintained. As can be seen in the Supplementary data available in Age and Ageing online, Video S2, the soft palate was also involved. Given the diffuse oropharyngeal and laryngeal spasm, focal or localised treatment was not indicated due to the increased risk of aspiration. Botulinum toxin injection was therefore not attempted.

As he managed well with the prescribed modified diet alone, a gabapentin trial was not considered.

Discussion

Dysphagia is relatively common in Geriatric medicine, with a reported prevalence of 15–50% in those over 60 years of age.
The patient can be unaware of, or minimise (as in this case) their swallowing problems, until they present with another medical issue. This puts them at an increased risk of aspiration pneumonia. Dysphagia can be due to abnormalities in the oropharynx, oesophagus, or may be functional. We present a rare cause of dysphagia: palato-pharyngo-laryngeal myoclonus [2]. In 80% of cases of palatal myoclonus, the presentation is due to the underlying neurological disease, again making this case even more unusual [3].

Myoclonus can be classified as cortical or subcortical [4] (see Table 1). Subcortical is further divided into non-segmental or segmental. Palatal myoclonus is a type of segmental, subcortical, myoclonus. Rhythmic contractions of the soft palate result from a dysfunction [essential palatal myoclonus (EPM)] or lesion [symptomatic palatal myoclonus (SPM)] in the anatomical Guillain–Mollaret triangle (GMT). The GMT comprises connections between the dentate nucleus, red nucleus and inferior olivary nucleus. Essential palatal myoclonus (EPM) causes repetitive opening and closing of the Eustachian tube, which the patient notes as a rhythmic, audible click (due to rhythmic contractions of the tensor veli palatini muscle arising from the lateral wall of the Eustachian tube, innervated by the motor division of the trigeminal) [7]. EPM disappears while sleeping. In contrast, SPM is not associated with the audible click as it involves the levator veli palatini muscle (innervated by the facial or ambiguus nucleus). Thus, it is felt that the two conditions are related to two different cranial nerves [5, 6, 8, 9]. The movement persists during sleep in SPM, as in this case. SPM is more common than EPM and can be associated with vascular lesions, multiple sclerosis, brainstem tumours and progressive ataxia palatal syndrome. It is also more likely to involve the larynx than EPM. Isolated laryngeal myoclonus is very rare [3].

Pathologically, in SPM [6] there is involvement of the cerebellar dentate nuclei, the central segmental tract and inferior olivary nucleus on the opposite side. The inferior olivary nucleus may show marked hypertrophy [10, 11] within 1–6 months, but the palatal tremor may appear within days or up to 49 months after the acute vascular lesion. It is known that when a partial lesion or denervation of the brainstem or spinal grey matter occurs, abnormal firing of some remaining neurons occurs. The loss of inhibitory output from the inferior olive produces rhythmic or irregular firing causing the segmental myoclonus [12, 13]. This loss of inhibition is responsible for the olivary hypertrophy seen on MRI. The olivary hypertrophy may resolve 3–4 years after the acute event [14].

In this case, there was also involvement of the larynx. There was no underlying neurodegenerative disease, and no neuroimaging evidence of a discrete lesion in the GMT.

Hence, this case is classified as subcortical SPM.

**Table 1. Differential diagnostic features of palatal myoclonus (developed from references [5, 6])**

<table>
<thead>
<tr>
<th>Features</th>
<th>Essential Palatal Myoclonus (EPM)</th>
<th>Symptomatic Palatal Myoclonus (SPM)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscle involved</td>
<td>Tensor veli palatini</td>
<td>Levator veli palatini</td>
</tr>
<tr>
<td>Prevalence</td>
<td>Less common</td>
<td>More common</td>
</tr>
<tr>
<td>Audible click</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Myoclonus during sleep</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Laryngeal involvement</td>
<td>Much less common</td>
<td>Sometimes</td>
</tr>
<tr>
<td>Ocular involvement</td>
<td>Never</td>
<td>Frequently</td>
</tr>
<tr>
<td>Extremity involvement</td>
<td>Never</td>
<td>Rarely</td>
</tr>
<tr>
<td>Aetiology</td>
<td>Unknown</td>
<td>Vascular lesions, MS, degenerative disease, trauma, encephalitis</td>
</tr>
<tr>
<td>MRI</td>
<td>Usually no lesion</td>
<td>Medullary lesion</td>
</tr>
</tbody>
</table>

**Figure 1. MRI of the brain showing enlargement and hyperintense signal within the olivary nuclei (see arrows).**

**Progress**

With education about a modified oral diet, he had no further coughing or choking episodes. Following mobility and balance rehabilitation, he was discharged back to independent community living.
This case highlights the importance of comprehensively assessing dysphagia in geriatric patients, and how a simple dietary modification can maintain function and independence, even in a complex case.

**Key points**
- Dysphagia is common and should always be evaluated.
- Identifying the underlying cause is important.
- Dietary manipulation alone may be adequate.

**Conflicts of interest**
None declared.

**Supplementary data**
Supplementary data mentioned in the text is available to subscribers in *Age and Ageing* online.

**References**

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