Lingual Myoclonus Presenting As Neck Lump

N Agrawal, S D'Sa, J Addams-Williams

Citation

Abstract
Isolated lingual myoclonus is a very rare entity. A 40 year old woman presented to ENT outpatients with lingual myoclonus referred as a pulsatile neck lump. A vigilant history and examination is required to ascertain this rare diagnosis.

INTRODUCTION
Rhythmic lingual myoclonus is a rare disorder and is more often described in association with similar palatal, ocular, facial, diaphragmatic, or shoulder movements. Isolated continuous rhythmic lingual myoclonus is exceptional and has not been described previously in the ENT literature. We report the case of a 40 year old woman referred to ENT outpatients with a neck lump thought to be pulsatile in nature. Closer examination revealed it to be lingual myoclonus.

She was referred to the neurology department for EEG assessment to ascertain which tongue muscle was involved and MRI brain to ascertain any unusual brainstem activity. Her rhythmic activity of the tongue musculature stopped spontaneously after 4 weeks of activity.

DISCUSSION
Abnormal involuntary movements may appear in the tongue, such as tremor, fibrillations, myokimia, and dyskinesias. Episodic or paroxysmal rhythmic lingual movements have been reported in association with chronic epilepsy and after head trauma. Continuous rhythmic nonepileptic lingual movements may accompany myoclonus of the palate-pharyngeal muscles (rhythmic palatal myoclonus); however, isolated continuous rhythmic nonepileptic lingual myoclonus is a very uncommon entity and has been documented only in three patients and never previously in the ENT literature.

The differential diagnosis of neck lumps include cervical auricle, lumps in submandibular and parotid regions, branchial sinus, branchial cyst, carotid body tumour, pharyngeal pouch, cervical rib, thyroid lumps, cervical lymphadenopathy, malignant lymphoma, cystic hygroma.

Isolated lingual myoclonus is a very rare phenomenon and confirmation of the diagnosis involves the use of monopolar electrodes to record abnormal lingual muscle movements, electroencephalography and MRI brain. Previous literature locating the anatomical site of the lesion producing lingual myoclonus is not clear. Electrical stimulation of an area medial to the inferior olive and close to adjacent hypoglossal nucleus in cats produces rhythmic movements of the posterior part of the tongue. Olivary ‘hypertrophic degeneration’ (in the medulla oblongata), is the most frequent lesion associated with palatal myoclonus. However a ‘chemical’ denervation due to unknown transmitter changes could also be invoked, as in other movement disorders.

In 2 of the previous cases of isolated lingual myoclonus assumed aetiological agents included head trauma and possible encephalitis. In the third case without any obvious aetiological agent the myoclonus disappeared after 2 weeks of sodium valproate administration. The isolated nature of these cases makes it difficult to precisely locate the aetiology of this condition but the busy ENT practioner should be aware of myoclonus of the orofacial musculature in his differential diagnosis of neck lumps.

CORRESPONDENCE TO
Mr Namit Agrawal BSc MRCS
1A Blackroot Road
Sutton Coldfield
West Midlands
B74 2QH
UK
Tel 00 44 7718916214
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Author Information

Namit Agrawal, BSc MRCS
Department of Otorhinolaryngology, Southmead Hospital

Sapna D'Sa, FDSRCS
Department of Otorhinolaryngology, Southmead Hospital

Julia Addams-Williams, MRCS
Department of Otorhinolaryngology, Southmead Hospital