

Facio-mandibular myoclonus: a rare cause of nocturnal tongue biting

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ABSTRACT – Facio-mandibular myoclonus is a parasomnia characterized by forceful myoclonus of masticatory muscles in sleep. This condition typically presents with recurrent nocturnal tongue biting, which can be misdiagnosed for nocturnal seizures. The use of surface electromyographic channels over the facial muscles during video-EEG monitoring is helpful in confirming the diagnosis based on a typical burst pattern. This case report highlights difficulties in the diagnosis of facio-mandibular myoclonus and useful features which differentiate it from similar conditions.

Key words: epilepsy monitoring, myoclonus, parasomnias, sleep, tongue biting

Patients presenting with tongue biting in their sleep may be referred to epilepsy clinics to exclude nocturnal seizures. Another well recognized cause includes bruxism of which the diagnosis is supported by a collateral history of teeth grinding in sleep, revealed by a bed partner, with or without coexistent obstructive sleep apnoea. Facio-mandibular myoclonus (FMM) is a relatively less conspicuous condition which typically also causes nocturnal tongue biting. We report a case of FMM of which the diagnosis was delayed for two years.

sation of jaw closure with biting of the edge of his tongue resulting in pain and bleeding. These episodes were more frequent over the preceding six months occurring at least four days a week. No collateral history was available as he did not have a bed partner. He had a background history of type 2 diabetes, psoriasis and chronic back pain following a motor vehicle accident. There was no history of diurnal seizures or risk factors for epilepsy. He had been unsuccessfully treated with carbamazepine for an empirical diagnosis of nocturnal epilepsy.

The general examination did not reveal any masseter muscle hypertrophy or tenderness over the temporo-mandibular joints. His neurological examination was unremarkable. Magnetic resonance imaging of the brain was normal.

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Case study

A 58-year-old man was referred to the epilepsy clinic with a history of recurrent tongue biting in his sleep for two years. He would wake from sleep with a sudden “tapping” sen-

Routine electroencephalography (EEG) did not demonstrate any epileptiform abnormalities.

He underwent long-term video-EEG monitoring (VEM) with scalp electrodes placed according to the 10-20 international electrode system. In addition, electromyographic (EMG) signals were acquired through surface electrodes placed over masseter, orbicularis oculi, and tibialis anterior muscles.

A typical clinical event was captured at day three of monitoring in light sleep. The video demonstrated a myoclonic jerk of the mandible resulting in sudden closure of the jaw causing laceration of the tongue. This was accompanied by activation of masseter and orbicularis oculi muscles on the EMG channels along with associated artefact on EEG channels. There was no contraction of the tibialis anterior muscle (*figure 1*). No ictal or interictal epileptiform abnormalities were detected during the entire video-EEG.

Based on the typical history and classic VEM findings, a diagnosis of FMM was concluded. Treatment was initiated with 1 mg clonazepam per night. On review, three months later, he reported only one episode of nocturnal tongue biting indicating a good response to the drug.

Discussion

Facio-mandibular myoclonus resulting in nocturnal tongue biting was first reported in 1991 (Aguglia *et al.*, 1991). Since then, a Medline search (accessed on 15 May 2010) has yielded only five more reports

(Kato *et al.*, 1999; Vetrugno *et al.*, 2002; Loi *et al.*, 2007; Wehrle *et al.*, 2009; Dylgjeri *et al.*, 2009), suggesting this condition is either rare or under-recognized. In a cohort of 41 patients with a clinical diagnosis of sleep bruxism (SB), four were found to have FMM rather than SB, based on sleep laboratory data (Kato *et al.*, 1999).

FMM is characterized by sudden forceful myoclonus of masticatory muscles causing vertical jaw movements occurring in isolation or clusters (Kato *et al.*, 1999). Patients with FMM classically present with tongue biting during sleep (Vetrugno *et al.*, 2002; Loi *et al.*, 2007; Wehrle *et al.*, 2009; Dylgjeri *et al.*, 2009). This is seen most commonly in stage 1 and 2 sleep followed by rapid eye movement (REM) sleep and rarely in slow wave sleep (Kato *et al.*, 1999).

Sleep bruxism is characterized by rhythmic, involuntary contractions of masticatory muscles in sleep, most commonly in stage 1 and 2. It is clinically suspected by the presence of teeth grinding noises in sleep, abnormal tooth wear, morning stiffness or pain in jaw muscles and masseter muscle hypertrophy (Lavigne *et al.*, 1996).

In FMM, the myoclonic movement of the jaw produces a "tapping" or "clicking" sound of teeth as opposed to a "grinding" or "clenching" sound in SB (Vetrugno *et al.*, 2002; Loi *et al.*, 2007). In FMM, the EEG polygraphy demonstrates activation of masseter and temporalis muscles followed by orbicularis oris and oculi muscles 7-20 milliseconds later (Vetrugno *et al.*, 2002; Loi *et al.*, 2007). The EMG burst duration is significantly shorter (<0.25 seconds)

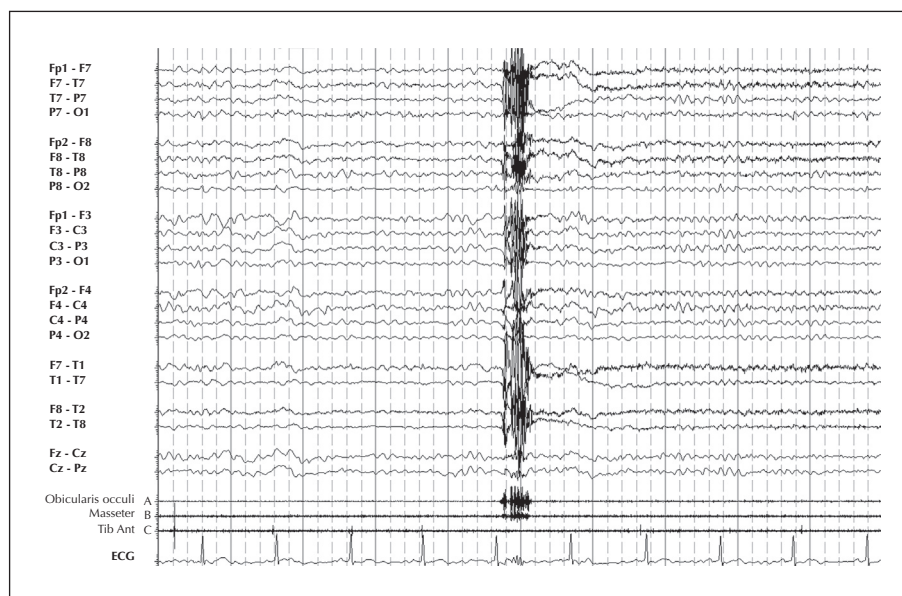


Figure 1. Video-electroencephalography with surface electromyography recording during an episode of facio-mandibular myoclonus. Note the activation of masseter and orbicularis oculi muscles.
A: orbicularis oculi; B: masseter; C: tibialis anterior.

compared to SB, although interburst intervals tend to be shorter in SB (Kato *et al.*, 1999). There is no associated activation of tibialis anterior muscle (Kato *et al.*, 1999). In SB the EMG activity could be phasic (three or more bursts in succession), tonic (sustained muscle contraction lasting >2 seconds) or mixed. The EMG bursts of FMM occur in clusters or isolation (Kato *et al.*, 1999). In SB the EEG usually shows “checker board pattern” of muscle artefact due to alternating jaw muscle contractions (Hirsch and Crispin, 1999).

The optimal treatment for this condition is unclear. Our patient demonstrated a good response to clonazepam. Clonazepam appears to be the most commonly used drug with case reports indicating a good (Wehrle *et al.*, 2009; Dylgjeri *et al.*, 2009) or partial (Vetrugno *et al.*, 2002) response, but also no response (Aguglia *et al.*, 1991).

FMM is considered to be a form of brainstem reticular myoclonus, involving a circuitry of fifth and seventh cranial nerve nuclei along polysynaptic pathways (Vetrugno *et al.*, 2002).

It is important for neurologists and epileptologists to be aware of FMM as such patients may be referred with a suspected diagnosis of epilepsy. Additional surface EMG channels should be included in VEM to confirm the diagnosis as illustrated in this case. □

Disclosure.

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