CASE REPORT

A Challenging Case of Recurrent Nocturnal Tongue Biting in an Infant

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Abstract

Sleep disorders are common in children and adolescents, with a major impact on their quality of life. Although considered a likely sign of an underlying sleep or neurological disorder, nocturnal awakenings are usually of psychogenic or behavioral nature. Clinical history, physical examination, brain imaging, and polysomnographic studies can be necessary to establish an appropriate differential diagnosis, particularly in the presence of alarm signs. Tongue biting is considered to be highly suggestive of an epileptic seizure, justifying the exclusion of this diagnosis, although it may occur in psychogenic events and sleep-related movement disorders as well, such as geniospasm or facial myoclonus. This study presents a curious and challenging case of recurrent tongue biting during sleep of unknown etiology.

Keywords: Bites, Human/etiology; Diagnosis, Differential; Infant; Sleep Initiation and Maintenance Disorders/diseases; Tongue/injuries

Keypoints

What is known:
- Nocturnal events can be challenging to assess and tongue biting is widely recognized as a clinical feature of epileptic seizures.
- A careful clinical history and recording of events on video electroencephalogram polysomnography usually enable an accurate diagnosis.

What is added:
- Hereditary geniospasm is a benign and self-limited condition with onset in early infancy and good clinical response to clonazepam.
- Psychiatric comorbidities and sleep disturbances must be evaluated and treated since they can impact the outcome of the disease.

Introduction

Sleep disorders are common in children and adolescents with major impact on their quality of life.[1] They may arise in isolation or concomitantly present with a range of neurological diseases.[1] Nocturnal awakenings are generally of psychogenic or behavioral nature, although they may be a telling sign of an underlying sleep disease, namely parasomnias and rhythmic movement disorders, or a neurological disorder, most frequently epilepsy.[2] Clinical history, physical examination, brain imaging and polysomnographic studies are essential to establish an appropriate differential diagnosis, particularly in the presence of red flags.[2] Tongue biting, specifically lateralized tongue biting, is considered highly suggestive of an epileptic seizure, which should prompt exclusion of the diagnosis.[3] However, it may occur in sleep-related myoclonus, sleep bruxism, geniospasm, dystonia and other sleep-related movement disorders.[4] Most of these are underrecognized entities that share a good clinical response to clonazepam and other benzodiazepines.[5] We describe our experience with a challenging case of recurrent tongue biting during sleep of unknown etiology.

Case Report

A 12-month-old infant was referred for a sleep medicine consultation due to a four- month history of multiple episodes of nocturnal awakenings preceded by tongue biting events, in which he woke up at night crying and agitated with swelling and bleeding lacerations in the anterior dorsum of his tongue. Most nights he fell

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asleep while breastfeeding and he needed the same association to fall asleep again after the incident. In addition to daily sleep deprivation, he had begun having trouble swallowing solid foods, which made a significant impact on his quality of life. There were no other signs suggestive of a seizure. No significant past medical and family history was reported, particularly regarding neurological diseases. His physical examination, including neurological examination, was normal except for the tongue lesion (Fig. 1). Brain magnetic resonance imaging and video electroencephalogram (EEG) monitoring were also unremarkable. Laboratory investigation revealed only the presence of microcytic, hypochromic anemia (hemoglobin 10.6 g/dL, mean corpuscular volume 71.4 fL, mean corpuscular hemoglobin 22.8 pg) with low ferritin (18 ng/mL) and iron reserves. Video polysomnography with supplementary EEG channels and surface electromyography (EMG) revealed 65% sleep efficiency, with most sleep on non-rapid eye movement (NREM) stage N2, without periodic leg movements and events of apnea-hypopnea, with normal awake and sleep EEG and no epileptiform or focal slowing abnormalities. The patient exhibited hypertonia of the mentum followed by quick suction movements on the tongue and brief arousal with awakening and crying in several stereotyped events captured during sleep. Throughout the day, recurrent episodes of chin trembling were detected, which the mother and the maternal grandmother also reportedly shared. After evaluation by pediatric neurology and maxillofacial surgery, his treatment plan included the implementation of a sleep hygiene routine, behavioral strategies, supplementary iron, and a trial of a low dose of oral clonazepam. At follow-up, his tongue biting events gradually diminished, his tongue showed good signs of healing, and his sleep thoroughly improved, enabling the possibility of tapering the dose with the same outcome.

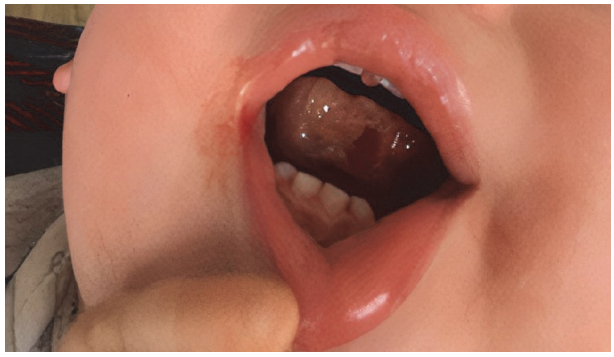

Figure 1. Tongue biting lesions.

Discussion

Sleep-related paroxysmal motor phenomena have a wide spectrum of etiologies, including epilepsy, sleep bruxism, sleep disorders, sleep-related movement disorders (like dystonia), and psychogenic disorders.[1] However, non-specific, nocturnal tongue biting, and agitation are uncommon and worrisome events in children that should prompt investigation. In particular, the role of nocturnal tongue biting in epilepsy has been thoroughly evaluated, justifying the exclusion of this diagnosis.[3]

Among the epileptic syndromes, nocturnal frontal lobe epilepsy (NFLE) is the most frequently associated with sleep, due to it being a focal epilepsy that is almost exclusively characterized by nocturnal episodes.[6] Ictal crisis can occur in any stage of sleep and is typically manifested in stereotypical, bizarre, and violent bursts of anomalous movements and postures.[6] A normal electroencephalogram, the absence of abnormal movements, and the spread of the events throughout the entire night are indicative of an alternative diagnosis. Hypnic myoclonus and rhythmic movement disorders occur frequently before sleep onset and do not repeat throughout the night.[7] Likewise, parasomnias such as non-rapid eye movement parasomnias occur almost exclusively during non-rapid eye movement slow-wave sleep, last several minutes and the person actively resists comforting, which does not apply to our case.[2] Sleep-related faciomandibular myoclonus, on the other hand, is a nocturnal motor phenomenon characterized by sudden, forceful, myoclonic jerks, involving muscles innervated by the fifth and seventh cranial nerves.[4] The disorder is more frequent in middle-aged adult males with onset after 50 years of age, and usually presents with normal electroencephalogram and neuroimaging.[4] The age of onset and the absence of muscle activity in the surface electromyography makes this hypothesis very unlikely. Sleep bruxism is an involuntary muscle activity associated with abnormal tooth wear and masseter muscle hypertrophy, resulting in constant teeth grinding, and clenching of teeth. It presents with a characteristic electroencephalogram and electromyography patterns, which were absent in the case described.[4]

The stereotypical movements observed during video polysomnography as well as tongue suction and chin trembling while awake evocated the diagnosis of geniospasm. This association has been described in previous studies.[5,8,9] Otherwise known as hereditary chin trembling, this is a rare, autosomal dominant condition, with paroxysmal and rhythmic chin and lower

lip movements accompanied by involuntary (continuous or intermittent) contractions of the mentalis muscle.[8] Geniospasm starts in early infancy (reaching peak at 10-18 months), is regularly exacerbated by stress and emotions, and typically improves with age.[8] Although optimal treatment is unclear, tongue biting shows a good clinical response to clonazepam, inducing remission in most cases, although it lacks significant effect on the chin trembling.[5,8-10] Therefore, further insight into the pathophysiology of this condition is needed. In our case, our hypothesis is that the impaired muscle control and slight immaturity of oral functions are typical of the patient age and that the age-related tongue-suckling and chin trembling may facilitate the event. Given that certain characteristics of this condition are comparable to those observed in parasomnia events, they may share common mechanisms.[8] Even though genetic testing may establish an accurate diagnosis, the favorable outcome achieved and the high cost for potential benefit may dismiss the immediate need for its execution. A similar pattern to parasomnia events supports waiting until the child is older to perform such testing, if necessary. Iron deficiency, particularly brain iron deficiency, is commonly implicated in the physiopathology of restless leg syndrome and periodic limb movements disorder.[11] Current guidelines recommend the assessment of iron status in sleep disturbance disorders, especially if the patient presents with uncontrolled limb movements.[11] In our case, we hypothesize that this is more likely a comorbidity (possibly due to insufficient intake of iron-rich foods) than a cause or consequence of a specific disease.

Nocturnal tongue biting should always prompt the investigation of epilepsy. However, rare causes such as faciomandibular myoclonus, parasomnias, and geniospasm should also be considered in the differential diagnosis of sleep-related seizure disorders, especially in the presence of normal electroencephalogram and frequent tongue biting incidents that do not respond to conventional antiepileptic drugs. Nocturnal video polysomnography is essential to reach an accurate diagnosis. Geniospasm or hereditary chin trembling is a benign and self-limited condition with onset in early infancy and good clinical response to clonazepam.

Author Contribuitions

JR and NM participated in the study conception or design. JR, JTC and NM participated in acquisition of data. JR, RLA and NM participated in the analysis or interpretation of data. JR, JTC, RLA and NM participated in the drafting of the manuscript. JR, JTC, RLA and NM participated in the critical revision of the manuscript. All authors approved the final manuscript and are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this study.

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Consent for publication

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Um Caso Desafiante de Mordeduras Noturnas Recorrentes na Língua em Bebé

Resumo
As perturbações do sono são comuns em idade pediátrica, podendo ter um impacto significativo na qualidade de vida da criança e do adolescente. Os despertares noturnos são habitualmente de natureza comportamental ou psicogénica. Contudo, podem ser um primeiro indicador de doença subjacente. Para um diagnóstico diferencial adequado, é frequentemente necessário conjugar história clínica, exame objetivo, neuroimagem e estudo poligráfico de sono, particularmente na presença de sinais de alarme. Embora também presente em diversos distúrbios rítmicos do sono, a ocorrência de mordedura da língua justifica a exclusão do diagnóstico de epilepsia. Os autores descrevem um caso curioso e desafiante de episódios recorrentes de mordedura de língua durante o sono.

Palavras-Chave: Diagnóstico Diferencial; Distúrbios do Início e da Manutenção do Sono/diagnóstico; Lactente; Língua/lesões; Mordeduras Humanas/etiologia