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Objective Tinnitus Associated with Essential Palatal Myoclonus: Report in a Child

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Abstract: Palatal myoclonus is an uncommon, rhythmic, "shock-like" involuntary movement of the muscles of the soft palate, throat, and other structures derived from the branchial arcs. Objective tinnitus is frequently neglected in review articles about childhood tinnitus. Our aim was to present the case of a 7-year-old girl with bilateral objective tinnitus due to palatal myoclonus without hearing impairment (normal hearing thresholds between 250 Hz and 8 kHz) but with otherwise normal hearing thresholds (250 Hz–8 kHz) and no evidence of intracerebral or systemic disorders. No treatment was useful.

Key Words: objective tinnitus; palatal myoclonus

A definition of tinnitus is any abnormal sound sensation perceived by the patient and originating inside the head. Tinnitus can be subjective (perceived only by the patient) or objective (perceived also by a consultant). Forty cases of objective tinnitus were reported in the literature between 1900 and 1966 [1]. Objective tinnitus is rare and can be the consequence of vascular disorders (e.g., internal carotid disease, arteriovenous fistula, arteriovenous malformation). Contractions of muscles of the oropharyngeal region and palatal myoclonus can also produce objective tinnitus. Usually, palatal myoclonus is "symptomatic" of a neurological disorder (approximately 70%); in a few cases, the palatal myoclonus is considered as essential [2]. Approximately 200 cases of palatal myoclonus are reported in the literature [3], and more than 10 cases concern infants [4].

We describe here a case history of a 7-year-old girl who presented with palatal myoclonus that induced objective bilateral tinnitus. The child was without hearing impairment, however (normal hearing threshold, 250–8 kHz). Etiopathogenic hypotheses and a therapeutic approach are discussed.

CASE REPORT

A 7-year-old girl was referred to the department of otorhinolaryngology because of annoying sounds resembling a clock's ticking and originating inside her head. These abnormal sounds began when the girl was 4 years old, but no research was carried out until she was 7. Then, for the first time, the young girl asked her parents to listen to the sounds coming from her ears. The sounds were audible by others as a clock's ticking; they were immutable and disappeared during sleep. Over time, the patient developed the ability to control the release and cessation of these sounds, which were characterized by a frequency of 90–100 per minute. The sounds were perceived as coming from both ears, although mainly from the right side, and were increased in intensity during angina.

The girl's medical history reflected a fall from a bed at age 3 without loss of consciousness and an attention disorder until age 5. At the time of the examination, the psychomotor development of this patient was registered as normal.

Endobuccal examination revealed evidence of symmetrical palatal myoclonus that may have been released by the patient. This palatal myoclonus was not perceived by the patient and was synchronous with the sounds resembling a clock's ticking. Articulation, deglutition, and vocal fold motility were normal. Neurological examination confirmed the absence of abnormalities of the cranial nerves, of facial tic, of cerebellar syndrome, and of sensorimotor deficit. It also confirmed the symmetry and normality of reflexes. Otoscopy showed a normal tympanum (eardrum) and enabled the visualization of a tympanic oscillation whenever the clicking occurred.

Otherwise, the results of the girl's clinical examination were normal. Acoustic impedance measurements were characterized by a normal tympanogram on both

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the right and left sides. Stapedial reflexes were recorded after a stimulation intensity of 85 dB. The tone-decay test results revealed only oscillation synchronous with the clicking. Tonal and vocal audiometry results were normal. Auditory evoked potentials demonstrated reproducible and synchronous waves I, III, and V without any asymmetry of the latency and with no lengthening of the interwave intervals. Cerebral magnetic resonance imaging (MRI) with and without gadolinium injection was normal and confirmed the absence of pathology in the brainstem. Electroencephalography results were normal. Because the girl's activities of daily living were not compromised, no treatment was started. At more than 1 year after diagnosis, no change was reported by the child and her parents. Her academic results were good.

DISCUSSION

Myoclonus is a brief, rapid, shock-like, jerking movement of a muscle or group of muscles and may be secondary to various etiologies. Myoclonus can be physiological during sleep but can be secondary to various neurological diseases (e.g., Creutzfeldt-Jakob, hepatitic encephalopathy) or to various toxins (e.g., bismuth, bromide) and to some metabolic diseases. Palatal myoclonus is a quite uncommon condition, defined as a continuous rhythmic contraction of the palatal musculature; its etiology is not clear. Palatal myoclonus occurs in two forms: It is either essential or symptomatic of a neurological disease. It has been shown that palatal myoclonus may be caused by a lesion affecting the dentate nucleus, the red nucleus, the inferior olivary nucleus, and the central tegmental tract (called the Guillain-Mollaret triangle) [5]. Studies performed in nonhuman primates have demonstrated that electrical stimulation of the inferior olivary nucleus induced palatal myoclonus [6]. In humans, hypertrophic degeneration of the inferior olivary nucleus may induce symptomatic palatal myoclonus [7]. In the case reported here, MRI did not demonstrate any lesion in this region, arguing for the myoclonus's essential origin. A hypersensibility of the reflex, including the mandibular nerve, has been assumed [8]. This mechanism remains a possibility in our patient. However, owing to the normality of the MRI and the neurological examination, we have made the diagnosis of essential palatal myoclonus.

The relationship between palatal myoclonus and tinnitus is not clear. One study has suggested that the contraction of the levator veli palatini muscle may induce variation of the pressure into the eustachian tube and, consequently, may induce tympanic oscillations [9]. It has been reported that the Valsalva maneuver may transiently stop the tinnitus from increasing the pressure inside the eustachian tube [4]. Our patient was able to stop the tinnitus at will but failed to describe very clearly the method she used.

A generally accepted therapeutic approach for palatal myoclonus does not exist. Because her activities of daily living were not impaired, we decided not to treat our patient. In cases of important symptoms, various therapeutic modalities can be proposed. First, drug therapies have included sedatives and anticonvulsants: clonazepam, carbamazepine, valproic acid, antiarrhythmics, and trihexyphenidyl. Treatment with botulinum toxin type-A injection in the palatal muscles under electromyographic guidance has proved its efficacy [10–14]. The injections are performed in the posteromedial face of the maxillary tuberosity. Though few cases have been reported in the literature, palatal myoclonus appears to be benign in the majority of the cases, usually regressing spontaneously.

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