Auricular Myoclonus

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ABSTRACT: We describe a young man with a two and a half year history of idiopathic irregular contractions of an antitragicus muscle in the absence of a more generalized movement disorder. These contractions persisted in sleep and could not be replicated voluntarily. Because proximal nerve block temporarily eliminated the movements and complex hand movements reduced their amplitude and frequency, we suspect a central generator. However, these movements were not associated with any known pathologic condition.

RÉSUMÉ: Myoclonus auriculaire Nous décrivons le cas d'un jeune homme qui présentait, depuis deux ans et demi, des contractions irrégulières idiopathiques du muscle de l'antitragus en l'absence d'un désordre du mouvement plus généralisé. Ces contractions persistaient pendant le sommeil et ne pouvaient pas être reproduites volontairement. Nous soupçonnons un site générateur central parce qu'un bloc nerveaux proximal a éliminé temporairement le myoclonus et que les mouvements manuels complexes en réduisaient l'amplitude et la fréquence. Cependant, ce myoclonus n'était pas associé à une condition pathologique connue.

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Movement disorders involving the external ear have not been commonly reported. We describe a case of irregular clonic movements of the antitragicus muscle, persisting during sleep.

Case Report

A 20-year-old left-handed university student presented with a two year history of involuntary movements of his right ear. Initially, these had been intermittent and he had been particularly aware of them while talking on the telephone with the receiver against his ear. After six months the movements became constant during all activities. He had difficulty falling asleep because of the ear movements and had been told that they persisted during sleep. There was no tinnitus (clicking or otherwise). He noted no hearing impairment and denied any other motor or sensory symptoms. He had never noted any abnormal movements of the other ear, or of the face, mouth, or throat. There were no previous neurologic symptoms. At age 7 he had suffered an uncomplicated otitis media but could not recall which ear had been affected. He was taking no medications and denied illicit drug use. The patient had no known toxin exposure. There was no family history of neurologic disorders. He had never been able to voluntarily wiggle his ears. There was no history of or evidence for a psychiatric disorder.

Findings of general physical and mental status examinations were normal. Cranial nerve examination revealed no abnormality apart from movement of the right ear. There were irregular clonic movements in which the antitragus and antihelix were approximated. These were oddly reminiscent of the openings and closings of a fish's gill and were compatible with isolated contractions of the antitragicus muscle, which is innervated by the facial nerve. No movements were seen in the scalp, face, contralateral ear, tongue, palate, pharynx, or elsewhere. Although irregular, the ear movements occurred consistently at an overall rate of 70-75 per minute. They were not synchronous with his pulse. Mental arithmetic and other cognitive tasks had no effect on these auricular movements. They persisted unchanged during talking, swallowing, and facial and head movements but could be reduced in amplitude and frequency by having him perform complex repetitive movements with either hand. Even so, the movements were never absent for more than five seconds. Although he felt that he could reduce the movement by intense concentration, we were unable to appreciate any change. Hearing was normal. Motor, sensory, and reflex examinations were normal as were coordination and gait. Plantar responses were flexor.

A surface electrode over the antitragus revealed irregular brief bursts of synchronous firing by several motor units simultaneous with the visible movements. Complex repetitive hand movements caused these bursts to be less frequent and of lower amplitude. MRI scan of head was normal. Serum ceruloplasmin, antinuclear antibody, rheumatoid factor, syphilis serology, blood count, and routine biochemical studies were normal.

A premastoid facial nerve block with 4 mL of 2% lidocaine resulted in complete cessation of the movements for the duration of the facial paralysis. A therapeutic trial of an oral medication or of a local therapy, such as botulinum toxin, might have proven effective. The patient, however, declined any pharmacologic intervention, explaining that he could tolerate the movements themselves, as long as they were not a manifestation of any more serious illness. When last seen in follow-up, two and a half years after the onset of the ear movements, he remained otherwise well.

Comment

Because facial nerve block eliminated these movements, we do not believe they arose from an abnormality of the distal nerve or muscle. Although we cannot discount the possibility that the movements originated in the proximal nerve, their diminution during complex hand movements suggests that they were subject to modulating influences within the central nervous system and arose there rather than in the periphery.

The intrinsic muscles of the external ear are not normally under voluntary control[1] although many people can wiggle their

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ears using extrinsic and scalp muscles.[2] Movement disorders involving solely the auricle have rarely been reported. Keshavan[3] described ten cases of ear tics but these differed from the present case in several respects. Keshavan's patients all had bilateral intermittent symptoms while awake and exhibited some degree of voluntary control over the movements. Their movements were attributable to contractions of extrinsic rather than intrinsic musculature and in most cases scalp movements were also present. His patients, unlike ours, often exhibited signs of psychiatric illness and improved with psychotherapy. Although tics may persist during sleep in Tourette's syndrome,[4] our patient had no other features to suggest this disorder. Tics often imitate coordinated voluntary acts and are often multiple rather than being isolated contractions of a vestigial ear muscle.[5]

Hemifacial spasm usually affects older patients and begins about the eye and mouth.[6] We are unaware of any reports of hemifacial spasm starting in the ear.

This patient's movement disorder is probably best characterized as myoclonus — an irregular, involuntary, shock-like movement.[7] Palatal myoclonus can involve areas distant from the palate.[8] Although facial muscles may be involved and Eustachian tinnitus may result from the palatal movements,[9] no reports mention spread to intrinsic auricular muscles. Although usually faster than the present case, palatal myoclonus may occur at rates of 40 - 600 per minute.[10, 11] Palatal myoclonus typically persists during sleep[12] and, even though classically described as inexorably persistent,[13] variability and intermittency have been described.[11, 14] Some cases of palatal myoclonus, unlike this case, abate during swallowing and phonation.[13] Palatal myoclonus may result from any of a variety of lesions within the Guillain-Mollaret triangle.[15] The present patient had no evidence for infarction,[16] tumor,[17] trauma,[18] or plaque[18] within this system. Olivary hypertrophy was not evident on MRI[19] and he had no other signs to support the possibility of a degenerative disease of the nervous system.[19] Most of these disorders could not be entirely ruled out.

Although the present patient's movement disorder may represent an unusual variant of any one of the disorders discussed above, it does not seem that intrinsic auricular movements like those described here have been previously noted. We suspect that this disorder represents focal myoclonus of brainstem origin although some classification systems[20, 21] do not explicitly recognize such an entity as distinct from palatal myoclonus. As no pathologic explanation for this patient's movements was evident, one can only speculate whether this represents a distinct clinical entity. Correlation with future cases of auricular movements in patients with and without abnormal movements elsewhere may establish the relationships between this and other disorders of movement.

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