

Essential Palatal Myoclonus Accompanied with Psychiatric Symptoms

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• Abstract

Palatal myoclonus is a rare and unique neurological disorder which can be primary or secondary to lesions of the central tegmental tract in the brain stem. This is a case report of a patient who complained of hearing a continuous disturbing noise for many years. Later an affective disorder superimposed on his previous complaints and confounded his auditory complaints. The co-morbidity resulted in attributing the complaint of hearing noise to his psychotic states and as a hallucinatory phenomenon. Thorough clinical and paraclinical examinations revealed that the patient had primary palatal myoclonus and the misattribution of the auditory complaints had resulted in judicious prescription of antipsychotics. Administration of appropriate therapy resulted in improvement of both problems.

Keywords • Myoclonus • palatal myoclonus • bipolar mood disorder

Introduction

Palatal myoclonus is a rare disorder presenting as unilateral or bilateral rhythmic involuntary movements of the soft palate. This condition which is also called "palatal tremor" or "palatal nystagmus" is a type of segmental myoclonus^{1,2} which manifest with a frequency of about 1.5-3 Hz.^{3,4}

Patients with palatal myoclonus may complain of hearing an annoying rhythmic click, which is related to the opening and closing of the Eustachian tube due to pharyngeal contractions. This click may be perceived by the examiner by placing a stethoscope over the patient's ear.³ Palatal myoclonus may also be accompanied with myoclonic contractions of other parts of the body such as extraocular, facial, pharyngeal, laryngeal or diaphragmatic muscles and muscles of the neck, trunk and even the extremities.³

Case Report

A 46-year-old, married, unemployed man from Azarbaijan, north-west of Iran, was hospitalized in Roozbeh Hospital in March 1999 for diagnostic evaluation. The patient's psychiatric problems started eight years prior to admission following his father's death and observation of his autopsied corpse. He had desperate thoughts about death, suicidal ideation, delusion of poverty and unusual behavior such as begging and collecting of worn out clothes from time to time. After three years, exacerbation of symptoms resulted in hospitalization in a psychiatric hospital and he received antipsychotic medication. However, the patient's condition was refractory to drug treatment. Six sessions of electroconvulsive therapy (ECT) were also unsuccessful in controlling the patient's condition. He had experienced a remarkable functional decline in the past four years and therefore he was hospitalized for further diagnostic evaluation. He displayed symptoms such as elation of mood and affect and uninhibited sexual behavior.

The remarkable point in this patient's history was the hearing an annoying noise in both ears for 20

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years. The noise was heard continuously by the patient and resembled the clicking of a clock. He believed that others would be able to hear the sound if they came close enough to him. No accompanying neurologic symptoms, such as hearing loss or vertigo were reported. The patient believed that his problem had remained unchanged during preceding years. His relatives stated that he was less intelligent than his peers since childhood.

On observing the patient's general appearance, isotropia of both eyes was evident. The patient had euphoric mood during the interview and seemed to have a subnormal IQ. Results of systemic physical examination were unremarkable. Ocular movement was normal on neurological examination. A continuous rhythmic movement of the soft palate was observed with a frequency of about 80/min. This was accompanied by an audible click which could be heard by the examiner when a stethoscope was placed on the patient's ear. No other abnormal or involuntary movement was observed in the other parts of the body. In addition, soft neurologic signs such as primitive reflexes (palmomental and mild sucking reflex) and mild dysdiadochokinesia were observed without any significant cerebellar signs.

Psychometric assessment revealed borderline mental retardation, dependent, inactive and immature personality with affective symptoms accompanied by transient episodes of psychotic symptoms.

Laboratory studies included complete blood count (CBC), erythrocyte sedimentation rate (ESR), liver function tests (LFT), thyroid function tests (TFT), electrolytes, blood urea nitrogen, creatinine, triglyceride, cholesterol, fasting blood sugar, urinalysis, VDRL and Wright test and were all normal. The results of audiometric assessment revealed no evidence of hearing loss.

A diagnosis of an affective disorder and palatal myoclonus in a subnormal patient was made and the presence of underlying disorders was evaluated. Palatal electromyography confirmed the presence of palatal myoclonus but the involved muscles were not defined. On ophthalmologic consultation pseudoisotropia due to negative Kappa angle was reported. Electroencephalo-graphy was normal and magnetic resonance imaging (MRI) of the brain revealed mild brain atrophy and mild ventricular dilatation.

Clonazepam (2 mg, qhs) along with carbamazepine (200 mg, bid) was prescribed. The affective symptoms were controlled and palatal myoclonus was virtually stopped with this treatment.

Discussion

Several pathophysiological mechanisms have been proposed to explain this phenomenon.^{1,4,5} Basically, all the lesions which interrupt the hypothetical circuit (lateral superior cerebellar peduncle, brachium conjunctivum, and dentate nucleus) will result in palatal myoclonus. This circuit is called the triangle of Guillain and Mollaret.

If the cause of palatal myoclonus is identified, the condition is called secondary palatal myoclonus. Otherwise, palatal myoclonus is considered to be primary or essential.

The diagnosis of essential palatal myoclonus is confirmed by the presence of the above mentioned signs and symptoms and paraclinical studies such as brain MRI, electromyography (EMG), somatosensory evoked potential (SSEP), electroencephalography (EEG) and biochemical studies.

In this patient, considering rhythmic myoclonic contractions of the soft palate and the presence of an annoying click audible by the examiner, symptomatologically a palatal myoclonus was diagnosed. Early onset of the disease (26 years old), absence of contractions click during sleep (confirmed by patient's spouse), absence of cerebellar signs and involuntary movements of other muscles favors the diagnosis of primary (essential) palatal myoclonus.

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In order to exclude any underlying disease and due to the fact that the patient had suffered various psychiatric problems, including full-blown affective states, a thorough paraclinical assessment was mandatory. The collected body of evidence suggested that the patient was affected with a subnormal constitution of unknown origin (probably a genetic disorder, birth trauma, ...) confirmed by brain atrophy at MRI. The stress experienced at the death of the patient's father and viewing his corpse probably triggered the appearance of an affective disorder, which initially manifested itself as depression and then as euphoria. According to the DSM-IV criteria, diagnosis of bipolar I disorder was made.⁶

To date, there has been no known etiological correlation between these two conditions. The rarity of palatal myoclonus and its coexistence with bipolar mood disorder with psychotic features, resulted in misdiagnosis of the sound which was induced by myoclonic jerks as an auditory hallucination. This incorrect interpretation of objective tinnitus as an auditory hallucination resulted in prolonged treatment of the patient with antipsychotic medication.

The treatment of choice for essential palatal myoclonus is the administration of medications, including clonazepam, sodium valproate, tetrabenazine, haloperidol, trihexyphenidyl and carbamazepine. Among these, clonazepam, 0.25-0.5 mg/day increasing gradually to 3.0-6.0 mg/day, and sodium valproate (250 mg/day, increasing to 1000 mg/day) have suppressed the movement in some cases. Surgical treatment for essential palatal myoclonus has not been favorable. Our patient responded to clonazepam and carbamazepine.

This case emphasizes the importance of detailed neurological examination in psychiatric patients. Attributing unusual complaints of mentally disordered individuals to their psychiatric illness is unwarranted and even rare and unusual signs and symptoms merit further scrutiny.

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