CASE REPORT

Tourette syndrome in an elderly patient

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Abstract
Background: Tourette syndrome (TS) is a neurodevelopmental motor disorder. The first diagnosis during adult life involves cases of pre-existing undiagnosed TS.
Case description: We present the case of an elderly patient with severe, non-remitted TS, misdiagnosed with "psychoneurosis". The patient was correctly labeled at the age of 82.
Conclusion: Despite delays in TS diagnosis, only a few patients remain undiagnosed by the age of 45. Both TS under- and misdiagnosis have an impact on patients' outcomes. HIPPOKRATIA 2019, 23(1): 47-48.

Keywords: Tourette syndrome, elderly, underdiagnosis, misdiagnosis

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Introduction

Tourette syndrome (TS), a neurodevelopmental motor disorder, is the most common cause of tics and the most severe tic disorder[1-3]. Although the motor and vocal tics are the hallmark of TS, around 80 % of patients, usually adolescents/adults, report associated bodily sensations/discomforts; these premonitory sensory urges are relieved by tic performance[4].

Clinically, TS is divided into i) simple TS, manifested predominantly by tics; ii) full-blown TS, expressed by complex phenomena, such as copropraxia/coprolalia, echopraxia/echolalia, and palilalia; iii) TS plus, i.e., TS comorbid with other psychopathology[5]. Although TS was once considered a rare condition, worldwide prevalence is currently estimated at around 1 %[5,6].

Case report

An 82-year-old Caucasian woman had been diagnosed 38 years before with "psychoneurosis". She visited the outpatients' department requesting an up-dated disability certificate to continue receiving her disability support pension. The patient repeatedly showed sudden/rapid/involuntary/purposeless movements of the head, trunk, upper, and lower limbs. She agreed to be admitted for diagnostic purposes.

During hospitalization, she showed a broad repertoire of involuntary movements manifesting in a variable sequence, such as eye blinking, lip-smacking, teeth-baring, mouth opening, throwing the head back, bending over, right-shoulder jerking, right-arm flexing, right-hand movement as in writing, knee-bending, foot-shaking and tapping (mainly on the right side). Abnormal movements were often accompanied by the sound "ha-ah" and frequent throat clearing. Also, she described a discomforting sense of inner tension, worsening dramatically when asked to stay still for a longer time.

Based on the patient's history, symptoms first appeared at the age of 7-8 years and were attributed to psychologically stressful events related to World War II. During that period, there was a lack of accessibility to health care facilities. She described herself as an otherwise healthy child. Over the years, her symptoms showed a pattern of partial remission-exacerbation. Since she had always attributed them to "psychological causes", she sought medical care much later. At the age of 50, she was diagnosed with "psychoneurosis", a label confirming the patient's beliefs about her condition. She received venlafaxine 75 mg and bromazepam 3 mg only for a short period because her symptoms did not improve. At last, she accepted the fact that she suffered from "psychological problems" related to an "incurable" psychiatric disorder characterized mainly by "nervousness".

Regarding psychiatric history, she did not fulfil the criteria for any major psychiatric disorder. She had never received neuroleptics. Medical history did not reveal any significant pre-/perinatal events, neurodevelopmental milestones, or systemic illness. Occasionally, she had visited different emergency departments due to diverse somatic symptoms; she had been examined by various clinicians and had received at some time atenolol and orphenadrine, which were taken intermittently. The patient had no family history of similar or other neurological/psychiatric disorders.

Her symptoms were recognized as tics. According to the Yale Global Tic Severity Scale (YGTSS)[7], motor tic severity was estimated at 17/25, vocal tic severity at 15/25, and impairment at 40/50 (total YGTSS score: 72/100). Neurological assessment was otherwise unremarkable. Based on the patient's history, drug-induced tremor, dystonia, and secondary tourettism were diagnostically excluded[8].

Complete blood cell count/comprehensive metabolic panel were all within normal levels. The normal thyroid function test excluded hyperthyroidism-related tics[2]. The electroencephalogram, suggested for the differential diagnosis between tics and epilepsy, was normal[2]. Brain magnetic resonance imaging revealed mild cortical atrophy, arterial vascular lesions in both hemispheres (basal ganglia, centrum

semiovale, and corona radiate), and periventricular leukoencephalopathy. A neurocognitive assessment revealed deficits in attention, memory, and executive functions.

Conclusively, based on her history, clinical manifestation, and diagnostic procedures excluding another medical condition, she fulfilled diagnostic criteria for TS and probable mild vascular neurocognitive disorder[1].

Pharmacological treatment was considered because of tic frequency/intensity and the patient's discomfort. Haloperidol at a dose of 1 mg/day was chosen due to its "A" level of evidence and favorable adverse effect profile in low doses[2,3]. The patient was re-evaluated one month after discharge. There was only a slight improvement in motor tics (YGTSS: 13/25), while vocal tics were practically unaltered (YGTSS: 14/25). The degree of impairment remained the same (total YGTSS score: 67/100). She was no longer motivated towards systematic monitoring and psychoeducation.

Discussion

Although TS was originally thought a lifelong disabling disorder, its prognosis proved better. In uncomplicated cases, there is a striking reduction in tic severity by the age of 19-20, especially in male patients[6]. Since most symptoms improve in half of TS patients, adults with substantial discomfort due to tics are unusual TS representatives[9,10]. Moreover, published cases of non-remitted/recurrent TS in elderly patients are scarce, as if the syndrome "vanishes" in late life. Usually, tic manifestation in older patients involves cases of drug-induced secondary tourettism due to neuroleptics, levodopa, and sympathomimetic agents, or secondary tourettism related to degenerative neurological disorders, stroke, head trauma, brain tumors, and infections[2,8]. To the best of our knowledge, this was the first reported case of severe, non-remitted TS in an elderly patient.

Typically, the mean age of TS onset is 6 to 7 years[6]. Diagnosis during adulthood usually involves cases of preexisting, unrecognized TS. It was shown that a significant proportion of patients receives the correct diagnosis with a delay of four years after onset[11]. Despite this delay, only 16 % of patients reached adulthood undiagnosed, and only 1.6 % was diagnosed after the age of 45[6]. The present report describes an 82-year-old patient with undiagnosed TS, despite previous examinations by several clinicians.

There are different explanations for TS underdiagnosis. Due to the lack of genetic and biologic markers, diagnosis is based on personal/family history and physical/neurological assessment. A series of examinations is only performed to exclude other disorders. The multidimensional phenotype, the waxing-waning course, the changing repertoire of symptoms, as well as the erroneous belief that coprolalia constitutes an essential diagnostic criterion hamper diagnosis[3,5,12,13].

Although tic severity was associated with functional impairment, it is often the comorbid psychiatric disorders that determine global outcome[4]. The only TS phenotype with a very low rate of behavioral problems is that of "tics only", affecting predominantly females[6,12]. Based on history, it is probable that the 82-year-old patient suffered from "tics only", an additional explanation for the fact that her condition did not attract medical attention.

Lack of clinical experience, together with the fact that tics may be voluntarily suppressed for short intervals, may instigate the misconception that symptoms are under volitional control. In these cases, complex motor tics may be mistaken for signs of nervousness[3,4,14]. In other cases, complex tics may not be differentiated from stereotypies, mannerisms, and parakinesias, while mild TS symptoms may be labeled as "psychogenic tremor"[13]. As a result, several TS patients may receive a false diagnosis of a mental disorder[3]. TS was once thought to be of "psychological origin", the result of inner emotional conflicts[12]. Accordingly, the 82-year-old patient was misdiagnosed with "psychoneurosis", an unspecific diagnosis, formerly used to describe a variety of psychiatric manifestations, such as anxiety, hypochondria, hysteria, and neurasthenia.

Altogether, this report underscores the remaining issue of TS under-/misdiagnosis, based on the unusual case of an elderly patient with misdiagnosed, non-remitted TS. Tics' phenomenology may not be well tolerated, especially in communities lacking educational background. Therefore, TS remains a potentially stigmatizing disorder. In the present case, the patient's embarrassment caused by TS symptoms led to social withdrawal. Misdiagnosis deprived her of the opportunity to receive appropriate treatment. Furthermore, she became so accustomed to her former "incurable" disorder that she rejected another diagnosis. As a result, she discontinued systematic clinical monitoring and, probably, medication as well.

Conflict of interest

Authors declare no conflict of interest.

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