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Tourette Syndrome; Is It an Annoying Disorder or an Inspiring Companion??!!!

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Abstract

Tourette syndrome (TS) is a hereditary neurobehavioral disorder which starts early during childhood and manifests with a group of motor and one or more vocal tics for a duration of a year at least. It tends to be a lifelong chronic disorder with many remissions and exacerbations but in general, it is not a degenerative disease and has no negative percussions on intelligence or life span. Cases with TS need proper professional evaluation to exclude any differential diagnoses and detect any comorbidities. Cognitive Behavior Therapy (CBT), medications, and supportive intervention are indicated for cases with significant functional impairment. Some sufferers consider TS a very annoying lifelong disorder while others think about it as an inspiring companion that makes them gifted, unique and special.

Keywords Tourette Syndrome (TS); Tic disorders; Obsessive Compulsive Disorder (OCD); Attention Deficit Hyperactivity Disorder (ADHD); Neuroleptics; Selective serotonin reuptake inhibitors; Cognitive Behavior Therapy (CPT)

Introduction

Tourette syndrome (TS) is a hereditary neurobehavioral disorder which starts early during childhood and manifests with a group of motor tics with one or more vocal tics for a duration of a year at least. It tends to be a lifelong chronic disorder with many remissions and exacerbations but in general, it is not a degenerative disease and has no negative percussions on intelligence or life span; nevertheless; learning disabilities may be encountered [1-5].

Epidemiology

Tourette syndrome; which is 4 folds more common in males than females, is encountered in about 0.4% to 3.8% of those aged between 5 and 18 years. So, it is no more considered a rare disorder but because most cases are mild and sufferers learn how to control their tics by time, it could be under or misdiagnosed [3-5]. The severity of the disorder is variable from mild to severe but generally it tends to be lesser as the affected child gets older [5-8].

Historical overview

Sprenger and Kraemer wrote a book late in the 15th century describing a priest suffering from tics that were believed to be due to possession by the devil. That description was considered as the first Tourette syndrome presentation. In 1825, Jean Itard, a French doctor, reported the first case of TS in a famous noble woman in his era. Later in the 19th century, Jean-Martin Charcot; a French neurologist asked his resident; Georges Gilles de la Tourette, to define an illness which is different from hysteria and chorea among some patients at Salpêtrière Hospital. In 1885, Gilles de la Tourette succeeded to report 9 cases with a peculiar convulsive tic disorder which was given the name, Gilles de la Tourette syndrome, by Jean-Martin Charcot after his resident [9,10].

What is behind the occurrence of Tourette syndrome?

Both nature (genetic and other biological factors) and nurture (environmental factors) play a role in the development of TS but the definitive etiology is unknown [11-13].

The majority of Tourette syndrome cases are shown to be inherited but the exact mode of inheritance is an issue of debate and no single gene has been claimed to be responsible for its development. Some TS cases were reported to act like those suffering from autosomal dominant disorders with 50% chance to transmit the mutant gene(s) to their offspring but whether the mutant gene(s) will be expressed or not and to what extent and form is greatly variable (variable expression and incomplete penetrance). In addition, it was found that gender plays a role in gene expression in cases of TS as males are more likely to manifest tics than females [14,15].

More recently, the inheritance of TS was suggested to be more complex; few genes might have substantial effects or many genes with smaller effects interact with environmental factors resulting in subsequent development of the disorder. On the other hand, many environmental factors as infectious agents or psychosocial factors have been implicated to influence the severity of TS but not the vulnerability for its occurrence [12,16]. Autoimmune brain injury related to certain infectious agents as group A beta-hemolytic streptococci has been linked controversially with the subsequent development of TS (Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptoccocal infections; PANDAS) [12,13,16].

Findings of extensive and advanced neuroimaging and neuropathology research on cases with TS or their donated postmortem brains refer to abnormalities in thalamus, basal ganglia, frontal cortex, and other regions of the brain whether in neuronal circuitry interconnecting cortical and subcortical regions or in

neurotransmitters concerned with neuronal communication in such regions; namely dopamine, serotonin, and norepinephrine [8].

Symptomatology of Tourette syndrome; an annoying disorder

Motor tics which are the core symptomatology of TS could be defined as sudden, repetitive, non-rhythmic movements. The most commonly recognized motor (physical) tics in cases with TS are eye blinking, movements of other facial muscles, shoulder shrugging, arm thrusting, jumping, and kicking. They are usually preceded by a prodromal urge (patients feel it is coming and they have to do it to relieve the tension they experience before it), and can be temporarily reduced [3,5,10,17,18].

On the other hand, vocal tics are involuntary sounds that result from the passage of moving air via the nose, mouth, and throat. The most commonly encountered vocal tics in TS are throat clearing, coughing, and sniffing. Also, TS can be associated in a minority of cases with coprolalia (screaming and yelling with obscene words or socially inappropriate and insulting remarks), echolalia (repetition of others' words), and palilalia (repetition of one's own words) [3,5,7].

Because of the prodromal or premonitory urge felt by TS cases before the expression of tics whether motor or vocal, such tics are rather considered unvoluntary or semi-voluntary than involuntary. This urge is usually in the form of feeling something in the eye or throat, discomfort in the shoulder, or an itch anywhere in the body that needs to be scratched. Accordingly, it is sometimes referred to as premonitory sensory phenomenon. For patients with TS, it might be considered the core manifestation of their disorder although not included in its diagnostic criteria [19,20].

Differential diagnoses, comorbidities, and complications

Tics of TS are different from other movement disorders as chorea, dystonia, dyskinesias, and myoclonus in being temporarily reducible, non-rhythmic, and could be anticipated by a prodromal urge [21]. Also, they differ from stereotypy of Autism Spectrum Disorder that starts earlier in life, is bilateral, symmetrical, and rhythmical, and involves mainly the extremities as hand flapping [22].

On the other hand, the most frequently encountered comorbidities in cases of TS are obsessive compulsive disorder (OCD), attention deficit hyperactivity disorder (ADHD), learning disabilities, and sleep disorders. Different studies reported comorbidities to range from 43-60% in cases of TS [23,24]. Meanwhile, complications of this chronic disorder include social discomfort and withdrawal, self-injuries, depression, and sleep problems [21].

Tourette syndrome as an inspiring companion

There are evidences that TS without a comorbidity might be a gift and not a disability. Neuropsychological testing showed that children with isolated TS are faster on motor coordination timed tests compared to their non TS counterparts. Furthermore, heightened awareness and noticeable attention to surroundings and details; which are of prime adaptive importance, are latent advantages observed in those with TS or genetically prone to develop it [25-27].

TS has been described in many famous figures in different aspects of life as education, medicine, music, art, literature, sports, and media. For instance, Samuel Johnson; a creative writer, poet, and critic of the 18th century, was most probably a sufferer of TS. He wrote a dictionary of the English language and was known as the English man of letters [28]. On the other hand, the famous American soccer hero, Tim Howard, attributed his heightened perception and extraordinary ability to hyper-focus to his neurological make up as one of the well-known TS patients [29,30]. Mozart has been assumed to have TS, nevertheless, no credible evidence has been provided to prove it. Scatological writings of Mozart that once attributed to tics of TS were wrongly interpreted as tics are not transferred to the written form [31,32].

Eventually, not everyone with TS will consider it as an annoying disorder; on the contrary, many of its sufferers may prefer not to get rid of their inspiring companion.

Evaluation of cases with TS

Any individual who suffers from multiple motor and non-concurrent at least one vocal tics for a year duration with an onset before 18 years of age and cannot be explained by another condition or substance as cocaine is diagnosed as TS according to DSM 5 criteria [3,4]. The diagnosis is based on observation of the symptoms of the suspected case preferably by many observers. Positive family history supports the diagnosis of TS taking in consideration that it is not encountered in all cases [33].

A battery of investigations may be resorted to, to exclude differential diagnoses and or comorbidities. EEG could be done to differentiate tics from seizures. MRI may be done to exclude structural brain abnormalities. Hypothyroidism that may cause tics can be ruled out by TSH assessment. In adolescents, urine drug screen for cocaine and stimulants may be needed. In cases of family history of hepatic disorders, measurement of serum copper and ceruloplasmin levels may be required to exclude Wilson's disease [33,34].

Evaluation of TS cases for comorbidities as OCD, ADHD, and learning disabilities is crucial as missed diagnosis of comorbidities could remarkably worsen the prognosis because they usually result in significant social, functional, and or academic impairment [3,34].

Treatment modalities for Tourette syndrome

Tourette syndrome is not a curable disease but because most cases are mild with no functional impairment, most of the patients do not need drugs to ameliorate their tics. For those with severe symptoms and significant functional impairment, the benefit of improving their symptoms and degree of function must be weighed against the risk of developing the adverse effects of the prescribed medications [35].

There are many medication classes that can be used in severe cases of TS including typical and atypical neuroleptics (e.g., haloperidol, risperidone, pimozide) and alpha adrenergic agonists (e.g., clonidine and guanfacine). For those with ADHD as a comorbid condition, psychostimulants (as methylphenidate and dextroamphetamine) can improve ADHD manifestations without worsening the tics but this issue is a point of debate. Instead, atomoxetine and tricyclic antidepressants could be tried for such cases. In cases with TS associated with OCD as a comorbidity, selective serotonin reuptake inhibitors as fluoxetine, sertraline, and fluvoxamine may be useful [5,34,35].

Apart from medications, behavioral techniques such as awareness training and competing response may be helpful for many cases to reduce their tics. Also, combined cognitive and behavioral therapy may be tried to train patients to voluntarily shift their reaction to the prodromal urge to control their tics (Habit reversal) and it is useful as

well in TS cases suffering from OCD as a comorbidity. Relaxation techniques (exercise, yoga, or meditation) and biofeedback may be helpful in some cases [36-38].

On the other hand, supportive interventions could be useful to help patients to cope with their disorder and its social and emotional percussions. Educating the patients, their families, school personnel, and counterparts about the disease and its nature is very useful for the patients to minimize their stress and to learn how to live with their condition [35].

Deep brain stimulation has been used in some adults with severe TS who show no response to other available therapeutic modalities but it is an experimental invasive technique that is contraindicated if OCD is a comorbidity and is unlikely to be widely adopted [39].

Prognosis and outcome

The condition in many cases with TS tends to worsen during adolescence and improve during maturation to adulthood. Symptoms might disappear in some cases for a few years and then return while in others it might not return at all. The presence of comorbidities or complications worsens the prognosis but in general TS is not a degenerative disease although it is a chronic and a lifelong condition [8,34].

Summary

Tourette syndrome (TS) is a hereditary neurobehavioral disorder which starts early during childhood and manifests with a group of motor and one or more vocal tics for a duration of a year at least. It tends to be a lifelong chronic disorder with many remissions and exacerbations but in general, it is not a degenerative disease and has no negative percussions on intelligence or life span. Cases with TS need proper professional evaluation to exclude any differential diagnoses and detect any comorbidities. Cognitive Behavior Therapy (CBT), medications, and supportive intervention are indicated for cases with significant functional impairment. Some sufferers consider TS a very annoying lifelong disorder while others think about it as an inspiring companion that makes them gifted, unique and special.

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