Tourette's syndrome

A disorder characterized by random, repeated, and stereotyped motor tic or vocal tics for over > 1 year, [1] usually in several “bouts” per day. Onset is before age 18 years (mean age: 5 years). Male: female ratio is 4:1. The tics may be socially inappropriate, as such, are disabling. TS is often associated with OCD, ADHD & other personality disorders.

The eponym was bestowed by Jean-Martin Charcot (1825–1893) on behalf of his resident, Georges Albert Édouard Brutus Gilles de la Tourette (1857–1904), a French physician and neurologist, who published an account of nine patients with Tourette's in 1885.

Tourette's was once considered a rare and bizarre syndrome, most often associated with the exclamation of obscene words or socially inappropriate and derogatory remarks (coprolalia), but this symptom is present in only a small minority of people with Tourette's.

Tourette's is no longer considered a rare condition, but it is not always correctly identified because most cases are mild and the severity of tics decreases for most children as they pass through adolescence. Between 0.4% and 3.8% of children ages 5 to 18 may have Tourette's; the prevalence of other tic disorders in school-age children is higher, with the more common tics of eye blinking, coughing, throat clearing, sniffing, and facial movements. Extreme Tourette's in adulthood is a rarity, and Tourette's does not adversely affect intelligence or life expectancy.

Genetic and environmental factors play a role in the etiology of Tourette's, but the exact causes are unknown. In most cases, medication is unnecessary. There is no effective treatment for every case of tics, but certain medications and therapies can help when their use is warranted. Education is an important part of any treatment plan, and explanation and reassurance alone are often sufficient treatment.

Comorbid conditions (co-occurring diagnoses other than Tourette's) such as attention-deficit hyperactivity disorder (ADHD) and obsessive–compulsive disorder (OCD) are present in many patients seen in tertiary specialty clinics. These other conditions often cause more functional impairment to the individual than the tics that are the hallmark of Tourette's; hence, it is important to correctly identify comorbid conditions and treat them.

Associations

Cavum septum pellucidum may also indicate disruption of neurodevelopment and has been associated with neurodevelopmental and psychiatric conditions including bipolar disorder, Tourette’s syndrome, obsessive-compulsive disorder, and schizophrenia, among others [2]

Clinical features

Tourette syndrome (also called Tourette's syndrome, Tourette's disorder, Gilles de la Tourette syndrome, GTS or, more commonly, simply Tourette's or TS) is an inherited neuropsychiatric disorder with onset in childhood, characterized by multiple physical (motor) tics and at least one vocal (phonic) tic. These tics characteristically wax and wane, can be suppressed temporarily, and are preceded by a premonitory urge. Tourette's is defined as part of a spectrum of tic disorders, which includes provisional, transient and persistent (chronic) tics.

Characterized by motor and vocal tics, which is often associated with psychiatric comorbidities. Dysfunction of basal ganglia pathways might account for the wide spectrum of symptoms in TS patients. Although psychiatric symptoms may be related to limbic networks, the specific contribution of different limbic structures remains unclear.

Temiz et al. used tractography to investigate cortical connectivity with the striatal area (caudate, putamen, core and shell of the nucleus accumbens), the subthalamic nucleus (STN), and the adjacent medial subthalamic region (MSR) in 58 TS patients and 35 healthy volunteers. 82% of TS patients showed psychiatric comorbidities, with significantly higher levels of anxiety and impulsivity compared to controls. Tractography analysis revealed significantly increased limbic cortical connectivity of the left MSR with the entorhinal cortex (BA34), insular cortex (BA48), and temporal cortex (BA38) in TS patients compared to controls. Furthermore, they found that left insular-STN connectivity was positively correlated with impulsivity scores for all subjects and with anxiety scores for all subjects, particularly for TS. The study highlights a heterogenous modification of limbic structure connectivity in TS, with specific abnormalities found for the subthalamic area. Abnormal connectivity with the insular cortex might underpin the higher level of impulsivity and anxiety observed in Tourette syndrome [3)].

Treatment

see Tourette's syndrome treatment.

Case series

Tourette's syndrome case series.

Case reports

A 7-year-old girl presenting involuntary motor and vocal tics, intellectual disability, childhood hypotonia, and dysmorphic craniofacial appearances, as well as comorbidities including attention deficit-hyperactivity disorder (ADHD), obsessive-compulsive disorder (OCD), and self-injurious behavior (SIB). The patient's CNV-seq testing revealed a de novo 320-kb deletion in the 9q34.3 region encompassing the EHMT1 gene.

This is the first case reporting Tourette-like syndrome secondary to KS1 with a de novo microdeletion in the EHMT1 gene. Our case suggests TS with ID and facial anomalies indicate a genetic cause and

broadens the phenotypic and genotypic spectrum of both TS and KS1 [4].

2018

Richieri et al., report the first case of a patient with severe, intractable Tourette Syndrome (TS) with comorbid Obsessive-Compulsive disorder (OCD), who recovered from both disorders with gamma knife stereotactic radiosurgery following deep brain stimulation (DBS). This case highlights the possible role of the internal capsule within the neural circuitries underlying both TS and OCD, and suggests that in cases of treatment-refractory TS and comorbid OCD, bilateral anterior capsulotomy using stereotactic radiosurgery may be a viable treatment option [5].

1)
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