

Tourette's syndrome

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Tourette's syndrome (TS) is named after the French neurologist, Georges Gilles de la Tourette. It is a neurobehavioral disorder of childhood onset characterized by multiple motor tics plus one or more vocal (phonic) tics.¹⁻⁴ Motor tics are sudden, brief, intermittent, involuntary or semi voluntary movements, whereas vocal tics are involuntary sounds produced by air movement through nose, mouth, or throat.^{4,5} The tics of TS are variable, not only in anatomic location but also in frequency and severity. This inherent variability is referred as waxing and waning.^{3,6} Besides motor and vocal tics, symptoms of comorbid neurobehavioral disorders are also frequently present including Attention Deficit Hyperactivity Disorder (ADHD) that is reported in 50-75% of cases and Obsessive-Compulsive Disorder (OCD) found in 20-60% of cases. Episodic aggression or rage attacks and learning difficulties are also frequent.⁷ The treatment of TS varies and should be multidisciplinary because of the combination of behavioral, emotional, physical and learning problems that affects performance at school, home and workplace.^{3,8} Many TS children must deal with the daily onslaught of teasing and ridicule from their peers. Parents, educators and physicians must work as partners in advocating the best possible school environment for children with TS.⁴ Medications are considered when the symptoms begin to interfere with peer relationships, social interactions, academic or job performance, activities of daily living and are not remediable to non-drug interventions.^{4,9,10}

Originally, TS was thought to be greatly rare, the prevalence was 0.5 per 1000, but there has been an

increase over the last 12 years. Tourette's syndrome is more common in children, especially in those with special educational needs.³ A recent study at a regular school found 3% of the students had TS^{7,8} whereas other study found 12% of children in special education classes had TS.^{3,6,8} Tourette's syndrome is found in all cultures, countries and racial groups³ and three times more common in males.^{3,4,8}

The aim of this report was to show that most behavioral disorders need multidisciplinary management by behaviorist, neurologist, psychiatrist, psychologist and pedagogue, and to give more information about Tourette's syndrome so that it can be detected earlier. Therefore, children who suffer from it can live a better life and develop their ability.

Report of the case

An 11-year-old boy was brought to the Growth and Development Clinic of Hasan Sadikin Hospital on April 23, 2003 with rage outburst as the chief complaint.

Since 2 years before admission, he had easily got angry and hit his mother especially when his wishes

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could not be accomplished. Besides that, he had always opposed his parents, uttered obscenities, and touched his brother's head every time he saw him, which caused them to quarrel. These complaints started with eye blinking that made his parents bring him to the Cicendo Eye Hospital. The blinking could only be reduced by replacing it with other movements, such as mouth opening, head jerking, shoulder shrugging, abdominal twitching and throat clearing, which happened many times daily. The parents said that when one movement seemed to reduce, then other movements would develop. These symptoms also reduced at school and he never uttered obscenities there, but when he came home, the symptoms worsened. There were neither history of hyperactivity, obsessive-compulsive behavior (OCB), imitating someone's words/actions nor history of tics or OCB in the family.

The family felt irritated, which caused them to be angry with him. At school, his friend often insulted him so he often took days off. According to his teacher, he had difficulty in learning. He did not pass the 2nd grade of elementary school and had always been at the lowest rank in his class.

He was born at term from an uneventful pregnancy of a multiparous mother and delivered normally by a midwife. There was no abnormal natal or post-natal history. His developmental milestones looked normal. There was no history of head trauma or seizure with/without fever. There was a history of high fever accompanied by cough, cold, and vomiting for 10 days when he was 9 years old.

Nine months before admission, he had been diagnosed at the Neurology Outpatient Clinic as having tic habits and given haloperidol. Six months after the diagnosis, the dose was increased because there had been no improvement. Three months after that, he complained of having headache after taking the drug, so the dose was reduced and another medicine, trihexyfenidyl (THF), was added. Then, he was referred to the Psychiatric Outpatient Clinic, diagnosed as having aggressive disorder, and referred to the Growth and Development Team of Hasan Sadikin General Hospital.

On physical examination, besides eyes, head and abdominal motoric tics, everything was within normal limit. Meanwhile, he was still taking the medicine from the Neurology Outpatient Clinic, which

were haloperidol 0.75 mg twice daily and THF 1 mg twice daily. The Growth and Development Team agreed to give him additional medicine i.e., clonidine 0.1 mg once daily, educate his family and perform an IQ test. A week later, the parents said that the tics seemed to reduce and the headache had recovered. Unfortunately, two weeks after that, the head and eye tics seemed to increase while uttering obscenities and abdominal twitching still happened sometimes.

The IQ test result using Weschler Intelegentia Scale for Children (WISC) was 59. It means that he was mentally retarded but still educable. He and his family were given educative counseling for his oppositional behavior and exercise therapy to control his tics. There was also dose adjustment for medicines given i.e., haloperidol 1 mg once daily, THF 1 mg three times daily and clonidine 0.015 mg twice daily.

Discussion

The diagnosis of TS is made based on history taking and observation of tics, sometimes accompanied by behavioral disorder like ADHD or OCB, and history of tics in the family. The Diagnostic and Statistical Manual of Mental Disorder-4th (DSM-IV) criteria includes,¹¹ both multiple motor and one or more vocal tics have been present at some time during the illness although not necessarily concurrent, the tics occur many times a day (usually in bouts) nearly everyday or intermittently throughout a period of more than 1 year and during this period there is never a tic-free period of more than 3 consecutive months, the disturbance causes marked distress or significant impairment in social, occupational, or other important areas of functioning, the onset is before 18 year-old and the disturbance is not due to the direct physiological effects of a substance (e.g., stimulants) or a general medical condition (e.g., Huntington's disease or post viral encephalitis).

In this case, the patient was diagnosed as having Tourette's syndrome based on the findings of repeated involuntary head jerking, eye blinking, and mouth opening (simple motoric tics) and throat clearing (simple vocal tic) that had happened many times daily since he was 9 years old. Meanwhile, other movements developed, from the shoulder to the hands, even in

the abdominal muscle. He always touched his brother's head (complex motor tic) and uttered obscenities (complex vocal tic). These tics were not caused by other neurological conditions or the use of neuroleptic drugs. This condition made him insulted by his friends, so he preferred to stay at home. All of those findings fulfilled the DSM-IV criteria.

Based on the IQ test, we found that he was mentally retarded, but still educable (59 WISC). Children with mental retardation are easier to have behavioral disorders than the non-retarded ones. There are four major causes of severe, challenging behavior in children with mental retardation: adaptive dysfunction, psychiatric disorders, medication side effects and organic causes. The most common psychiatric disorders are mood disorders, such as depression and bipolar disease. Less commonly seen are anxiety disorders, psychosis, Tourette's syndrome, ADHD, and OCB.²

The etiology of TS is mostly genetic, but the precise inheritance pattern is still unclear.³ A multi-site study that involved 76 affected families with a total of 110 pairs of siblings showed two suggestive regions (multipoint maximum-likelihood scores >2.0) i.e., 4q and 8p.^{4,12} Besides inherited genetic vulnerability, it has also been suggested that perinatal insults (e.g., birth injuries) and, a more recent (and somewhat speculative) hypothesis, infections with streptococci (Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections = PANDAS) or viruses were suspected to have a role in the development of TS.³

The etiology of TS in this patient was unclear. There was neither history of tic/OCB in the family nor abnormal perinatal conditions. Nevertheless, he had a history of high-grade fever for 10 days before he started developing tics.

One of behavioral disorders seen in TS is poor impulse control/inability to control anger, as a result of which, some patients may have outburst of temper, episodic attacks of rage, etc.^{4,6} Some investigators have suggested that it correlates with coprolalia, copropraxia, echophenomena, and OCB.⁶ Other behavioral disorders include antisocial or oppositional behavior, anxiety disorders, depression, and self-injurious behavior (SIB).^{4,6} This patient had already showed behavioral disorder. He had an outburst of temper, particularly to his mother, and always opposed his parents.

The majority of TS patients can be managed well without drug therapy.¹⁰ The first step of treatment is a proper education to the patient, family members, teachers, and others who interact with the patient.^{4,10} For all children, self esteem stems from mastering developmental challenges and taking new initiatives, and this requires great effort. However, children with TS must contend with both internal and external forces beyond the normal developmental challenges, not only in suppressing their tics in front of others, but also in developing a positive self-image in the face of negative feedback from their environment. The family can cope with this situation by helping the child to fight against those negative forces, that could undermine his stability.⁹

Chemotherapy is, at present, the mainstay of treatment for motor and vocal tics, as well as some of the associated behavior,³ that do not seem amenable to behavior modification and probably will be more effectively treated by pharmacological management in conjunction with psychotherapy or counseling.⁶ The philosophy of pharmacological management is to treat only if the tics are bothersome to the patient and to accept the fact that they will not completely disappear with treatment. Therefore, the target is to decrease the severity of the tics with the least amount of medication possible.⁸

Therapy for TS should be individualized and tailored specifically to the needs of the patient and the most troublesome symptom should become the first target.⁴ Dosage should be titrated slowly in order to achieve the lowest satisfactory one. In the long run, the goal is to use as little medication as possible.¹⁰

This patient was given haloperidol, a dopamine antagonist. Many studies reported that haloperidol is effective for TS, but it needs close monitoring for its unacceptable side effects. To reduce haloperidol side effect, he was given THF. Other medication given was clonidine. Other study found that clonidine is as good as haloperidol but has fewer side effects and can improve tics, ADHD symptoms and behavior problems.³ A special education was needed for his mental retardation, but he strongly refused it. Actually, an educable mental-retarded person could still follow general school, but only until 5th or 6th grade of elementary school. If his adaptive ability is good, he could live independently as a normal adult.¹³

As the prognosis, in 70% of TS children, the severity and frequency of tics will diminish significantly during the late teen years. A study estimated that 30-40% will experience a total remission by late adolescence, 30% or more will experience significant improvement in tic symptoms, and the rest will continue to experience the same severity of symptoms. It appears that there is no correlation between tic severity in childhood and long-term prognosis. Despite their difficulties, 99% of children with TS had graduated from high school and 90% were either employed full-time or enrolled in graduate programs.⁶ Unfortunately, this patient was mentally retarded, but with good and appropriate training, there is a chance for him to live independently as an adult.

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