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CASE REPORT

GILLES DE LA TOURETTE SYNDROME (GTS)- A RARELY DIAGNOSED AND UNDER REPORTED DISORDER IN NIGERIA

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ABSTRACT

Background: Tourette Syndrome (TS) is an uncommon neuropsychiatric disorder and less frequently diagnosed. The population prevalence estimate of TS in children was 0.3% to 0.9%. In Nigeria, this suggests that approximately 100,000 people are affected considering the estimated population of 200,000,000.

Objective: To present a case report of a teenager with a diagnosis of Gilles de la Tourette Syndrome.

Case Report: This is case report of a 15-year-old female secondary school student who presented to psychiatric department of a Nigerian Teaching Hospital with 3 years history of verbal and physical aggression towards parents and siblings. She was abusive and sometimes beats her siblings and parents for no obvious reason. She talks repeatedly to imaginary person, saying "leave me alone, I am not killing your child, you do not have respect" She sometimes hits the imaginary person. She blinks, hisses, grunts and shouts. These behaviours are repetitive, usually last for about 3 minutes.

Tourette syndrome diagnosis was made after comprehensive evaluation using the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) criteria.

Conclusion: The appropriate referral of this patient is of great benefit in the course of her illness. She achieved significant improvement to enable her continue education.

Keywords: Gilles de la Tourette Syndrome, Sokoto, Northwest Nigeria

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Introduction

Tourette Syndrome (TS) is a rare neuropsychiatric disorder and uncommonly diagnosed. Its prevalence in children is estimated at 0.3% to 0.9%.¹ In Nigeria, this would mean approximately 100,000 people are affected considering the estimated population of 200,000,000.

The condition was named 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 syndrome is characterized by multiple motor tics and at least one vocal (phonic) tic. A tic is a sudden, repetitive, nonrhythmic motor movement or vocalization involving discrete muscle groups.² Tics are classified as either motor or phonic, and simple or complex. Motor tics are movement-based tics affecting discrete muscle groups. Phonic/Vocal tics are involuntary sounds produced by moving air through the nose, mouth, or throat.³ Simple motor tics are typically sudden, brief, meaningless movements that usually involve only one group of muscles, such as eye blinking, head jerking, or shoulder shrugging.⁴ Motor tics can be of an endless variety and may include such movements as hand clapping, neck stretching, mouth movements, head, arm or leg jerks, and facial grimacing. Complex motor tics are typically more purposeful-appearing and of a longer nature. They may involve a cluster of movements and appear coordinated.⁵ Examples of complex motor tics are pulling at clothes, touching people, touching objects, echopraxia (repeating or imitating another person's actions) and copropraxia (involuntarily performing obscene or forbidden gestures). Complex phonic tics include echolalia (repeating words just spoken by someone else), palilalia (repeating one's own previously spoken words), lexilalia (repeating words after reading them), and coprolalia (the

spontaneous utterance of socially objectionable or taboo words or phrases). Coprolalia is a highly publicized symptom of TS; however, only about 10% of TS patients exhibit coprolalia.⁵

The fifth edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5, 2013), reclassified Tourette's and tic disorders as motor disorders listed in the neurodevelopmental disorder category, with persistence for more than a year.⁶ According to DSM-5, Tourette's may be diagnosed when a person exhibits both multiple motor and one or more vocal tics over the period of a year; the motor and vocal tics need not be concurrent. Tics may appear up to the age of eighteen, but the most typical age of onset is from five to seven.²

A study from the Yale Child Study Center showed that the ages of highest tic severity are eight to twelve (average ten), with tics steadily declining for most patients as they pass through adolescence.⁷ The exact cause of Tourette's is unknown, but it is well established that both genetic and environmental factors are involved.⁸ Genetic epidemiology study has shown that the overwhelming majority of cases of Tourette's are inherited, although the exact mode of inheritance is not yet known and no gene has been identified.⁷

Tourette syndrome is uncommonly diagnosed. Very few cases of this disorder have been reported in Nigeria. One of such cases was a 13 year old Nigerian boy with Gilles de la Tourette's syndrome.⁹ Another case of TS was a 9-year old school boy with co-existing Attention Deficit Hyperactive Disorder (ADHD).¹⁰

CASE REPORT

A 15-year-old female secondary school student was referred by a nurse to seek for treatment at the psychiatric clinic of the Usmanu Danfodiyo University Teaching Hospital (UDUTH), Sokoto, Northwest Nigeria.

She presented with 3 years history of verbal and physical

aggression towards parents and siblings. She was abusive and sometimes beats her siblings and parents for no obvious reason. She talks repeatedly to imaginary person, saying "leave me alone, I am not killing your child, you do not have respect" She sometimes hits the imaginary person. This usually last for about 3 minutes.

She blinks, hisses, grunts and shouts repeatedly. These behaviours are sudden and rapid. The parents have noticed these abnormal movements and sounds prior to when she started making meaningless sentences. These symptoms have remained stable, occur several times daily. She had no control over them. All these occur in clear consciousness at school, home, church and other places She kept herself clean in spite of the illness and communicates normally whenever she is not exhibiting abnormal facial movements, sounds and sentences. No history of hearing strange voices or other unusual experiences.

No history of fever, loss of consciousness, convulsions or substance use. She has been out of school for the past two years because of these complaints. No previous history of mental illness or chronic medical illness like Sick Cell Disease. Both parents are alive. Patient is the first born and has 3 siblings. There was similar history of abnormal movements like repeated blinking of eyes in her 3 younger siblings.

She was a product of full-term pregnancy, delivery was uneventful. Her developmental milestone was normal. Patient academic performance was within average in both primary and secondary school, until when her illness became pronounced to interfere with her school.

Mental state evaluation revealed a teenager with several episodes of abnormal motor behaviours and vocalizations like facial grimacing, hissing, grunting, repeatedly and intermittently saying the same statement "leave me alone, I am not killing your child".

She pointed at the reviewing doctor, made incoherent speeches as if she was abusing imaginary object. They were sudden, rapid and appear to be involuntary. They lasted for about 2-3 minutes. She was conscious and responded to call during each episode.

She also paced repeatedly during assessment. These abnormal movements and sounds have no functional significance. There were no thought disorders or compulsive acts. Physical examination including central nervous system was normal.

A diagnosis of Gilles de la Tourette's syndrome (TS) was made and she was admitted. Relevant laboratory and radiological investigations were conducted with no positive findings. The prognosis was discussed with her parents and they were reassured. She was commenced on Tablets Haloperidol 5 mg tds and trial of anticonvulsants. She did not comply with medications because of side effects.

The Haloperidol was discontinued and replaced with Risperidone 2mg tds. Within six weeks of treatment, she had significant improvement. The vocal tic was no longer spontaneous but occurs only on provocation and was limited to just saying "leave me alone".

Behavioural modification could not be commenced because rapport was difficult to establish. Patient was still irritable and uncooperative. She attends follow up clinic regularly. There was sustained improvement and the possibility of going back to school seems to be bright according to the parents. The typical rage attack of parents and siblings has subsided since on medications.

The parents were advised to come with the other 3 siblings for evaluation, but they were reluctant for no obvious reasons.

Discussion

Tourette syndrome diagnosis was made after comprehensive evaluation using the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) criteria.¹¹ The diagnosis of TS should be in doubt in the absence of simple tics. Vocal tics can be helpful in ruling out other diagnoses because they are rare in other neurological conditions. Exceptions include Huntington's disease and Sydenham's chorea (SC).¹² A number of conditions produce symptoms resembling the tics of TS, including myoclonus, tremors, chorea, athetosis, dystonias, akathisia movements, paroxysmal dyskinesias, and ballistic movements.¹³ The differential diagnosis of TS includes genetic conditions such as Huntington's chorea, metabolic diseases such as Wilson's disease, structural diseases as in hemiballismus associated with insult to the subthalamic nucleus, post infectious autoimmune processes such as Sydenham's chorea (SC), neuroacanthocytosis, and side effects of antipsychotic medications such as the dystonia and akathisia. In appearance, complex tics may also be indistinguishable from some compulsive rituals, but they can be distinguished based on the antecedent presence of either premonitory urges or obsessional thoughts.¹⁴

Approximately six weeks of being on medication she had significant improvement. The vocal tic was mainly observable on provocation, no longer spontaneous and was limited to a phrase "leave me alone".

Behavioural modification could not be commenced because rapport was difficult to establish. Patient has been very irritable and uncooperative. There is increasing evidence supporting the use of habit reversal (HRT) in the treatment of tics.¹⁵ Relaxation techniques, such as exercise, yoga or meditation, may be useful in relieving the stress that may aggravate tics, but the majority of behavioural interventions such as relaxation training, with the exception of habit reversal have not been systematically evaluated and are not empirically supported therapies for Tourette's.¹⁶ Habit reversal has been found effective for the treatment of individual tics. A study used habit reversal to treat three subjects with the multiple tics of Tourette Syndrome.¹⁴ Tic frequency was reduced over a 6–8-month period by 93–95% in the clinic setting and by 64–99% in the home setting with a concurrent decrease in severity. The procedure holds promise as a treatment for Tourette Syndrome.¹⁴

This patient has been out of school for 2 years. The major distressing symptom is coprolalia. Among the most disruptive behaviors in some children with TS are recurrent episodes of explosive anger or aggression (commonly termed rage attacks). Approximately 25–70% of patients with TS in clinical settings report experiencing episodic behavioral outbursts and anger control problems.^{17,14} When present, episodic rage is often identified by families as the most impairing symptom in their children with TS.¹⁸

A study of 46 subjects at 19 years of age found that the symptoms of 80% had minimum to mild impact on their overall functioning, and that the other 20% experienced at least a moderate impact on their overall functioning.²

Electro Encephalography, CT Scan, Thyroid Function Test (TFTs) were requested by the neurology team of the hospital, but due to financial constraint, some of these investigations

could not be carried out. Brain imaging studies are not usually warranted.¹⁹ The treatment of Tourette's focuses on identifying and helping the individual manage the most troubling or impairing symptoms.¹⁵

The gene or genes associated with Tourette's have not been identified, and there is no potential "cure".⁴ The classes of medication with the most proven efficacy in treating tics are typical and atypical neuroleptics including risperidone, ziprasidone, haloperidol, pimozide and fluphenazine.¹⁹

Deep brain stimulation has been used to treat adults with severe Tourette's that does not respond to conventional treatment, but it is regarded as an invasive, experimental procedure that is unlikely to become widespread.²⁰

Although she was out of school for 2 years, parents felt there was possibility of her to go back to school due to improvement in her condition. In a follow-up study of thirty-one adults with Tourette's, all patients completed high school, 52% finished at least two years of college, and 71% were full-time employed or were pursuing higher education.²¹

This patient was maintained on Risperidone and has been responding to treatment. After discharge, she was attending the follow up clinic regularly with sustained improvement. However, a few months later, she stopped coming for follow up.

Conclusion and Recommendation

The appropriate referral of this patient is of great benefit in the course of her illness. She achieved significant improvement to enable her continue education.

There is need to strength the referral system among clinicians at the primary care for expert evaluation of uncommon disorders.

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