

GILLES DE LA TOURETTE'S SYNDROME- A CASE REPORT.

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ABSTRACT

This case report is on Gilles de la Tourette's syndrome, a disorder of movement and sound.

It is a very uncommon diagnosis that was made on a female 29 years old Nigerian undergraduate. The diagnosis was predicated on clinical features, like virtually all psychiatric diagnoses. Management was behavioural interventions and low dose haloperidol with resultant complete remission.

This case brings to the fore the need for appropriate referral and high clinical diagnostic acumen.

Key words: Gilles de la Tourette's syndrome, uncommon diagnosis, clinical features, haloperidol, behavioural interventions)

INTRODUCTION

Gilles de la Tourette's syndrome is a rare psycho-neurological tic disorder¹.

Tics are defined as rapid and repetitive muscle contractions resulting in movements and or vocalizations that are experienced as involuntary².

The syndrome was named by Jean Martin Charcot after the French neuropsychiatrist Georges Albert Edouard Brutus Gilles de la Tourette³. In 1884 Gilles described nine patients who were affected with tics characterized by echolalia, pallilalia, coprolalia, a want for touch and stuttering³. One of the nine patients was the Marquise de Dampierre. This

aristocratic lady lived as a recluse and ticked and blasphemed¹. Her disturbance began when she was seven years old and persisted until her death at the age of eighty years.

During the closing years of the 19th century to the mid 1900's the syndrome was well documented and extensively reported. It had seemed to disappear as interest was lost in it. In 1978 Shapiro and his colleagues published a comprehensive multidisciplinary monograph about the condition and thereafter Gilles de la Tourette's syndrome was accepted as a specific entity⁴.

Several historic figures might have been affected by this syndrome, including Prince Conde, a member of the French royal family, Wolfgang Amadeus Mozart and Dr. Samuel Johnson, the British diarist^{1,5}.

A diagnosis of the syndrome is made when there are multiple motor tics and at least one vocal tic, occurring many times a day for nearly everyday for more than one year⁶.

The life time prevalence of the syndrome is estimated to be 4 to 5 per 10,000; commoner in children and 3 times more in males^{2,7}. The motor tics usually precede the vocal tics².

Examples of motor tics that occur in this syndrome are eye blinking, facial twitching or grimacing, shoulder shrugging, neck jerking, grooming behaviour, smelling of objects,



jumping, touching behaviour, echopraxia, copropraxia; while the vocal tics include grunting, sighing, throat clearing, coughing, sniffing, snorting, barking, coprolalia, pallilalia and echolalia².

The aetiological factors are numerous and could be idiopathic. The factors include hereditary, psychogenic, post-traumatic, post-central nervous system infections, derangement of the biogenic amines, drugs like methylphenidate, levo dopa, long-term chlorpromazine therapy etc.⁸⁻¹⁷.

There was no local literature found on Gilles de la Tourette's syndrome on literature review, possibly because of its rarity.

CASE REPORT

A 29 year old female Nigerian undergraduate was referred from the neurology team of University of Benin Teaching Hospital to the psychiatric team of same hospital.

She was referred on account of abnormal movements and sounds of two years duration and a one year history of fleeting 2nd person auditory hallucination.

She was apparently well until 2 years prior to presentation when she suddenly began to observe abnormal movements. The eyes could suddenly close and she would be unable to open them, her head could turn to one side, or she could be grimacing, grinning, smiling or showing her teeth. The neck and the trunk sometimes twist. The head or any of the limbs could shake individually or in combination. One or both knees could suddenly bend. Some times her mouth becomes as if glued together and she would be unable to talk.

About six months later, she also began to make abnormal sounds. She could hiss, grunt, or at times actually spoke words or make sentences that no one or she could understand.

These abnormal movements and sounds could happen at the same time or separately. They have no functional significance and do not follow a particular pattern. They are sudden and rapid, with each episode lasting for about a minute. These symptoms have remained stable. She had no full control over them but was sometimes able to suppress them. The abnormal movements and sounds occur everyday, sometimes several times a day. She could predict their occurrence but unable to tell what brings them up. The episodes were self-limiting.

The patient maintained awareness of self, the event and able to recall the experience; communicates normally in the absence of vocal tics.

It has never happened in her sleep, or caused her road traffic or domestic accident, but has occurred occasionally in the class and church.

She refuted hearing strange voices, claiming they are her thoughts. She thinks of herself negatively because of the above symptoms.

No history of fever, loss of consciousness, headache, seizures, heat or cold intolerance, sore throat, carbon monoxide poisoning, substance use disorder, tremors, dancing movements or regularly prescribed medication was elicited. Also, no psycho-social stressors predated these symptoms and no family history of similar condition or mental illness generally.

In her mental state evaluation there were no thought disorders, compulsive acts or cognitive deficits. However several episodes of abnormal motor behaviours and vocalizations which consisted of stretching and turning the neck, shrugging the shoulders, facial grimacing, flexing elbow and shoulders; as well as hisses, grunts and sniffs were extravagantly



manifested during the evaluation. She pointed at the reviewing doctor, made incoherent speeches as if she were abusing the doctor. They were sudden, rapid, non-rhythmic, and stereotypic and appear to be involuntary. They lasted between 30 and 60 seconds. She was conscious and accessible during each episode.

Physical examination including central nervous system was normal.

A diagnosis of Gilles de la Tourette's syndrome was made.

She could not afford a cranial computed tomographic scan. This would have ruled out intracranial neoplasm. Serum calcium and phosphorus were normal. This was done because their derangement could cause abnormal movement. Urinalysis was also normal.

Individual psychotherapy and behavioural therapy were commenced. She was educated on the disorder. The prognosis was discussed with her and she was helped to gain insight into the disorder.

She was thought self monitoring, and how to control or modify the symptoms. She was reassured.

She was also put on Tablets Haloperidol 2.5 mg daily and trial of anticonvulsants. She did not comply with medications because of side effects.

Within ten weeks of treatment she had complete remission.

It is noteworthy that the emphasis of the treatment was on behavioural modification to which unarguably she responded.

DISCUSSION

Some authorities opine that most cases of tic disorder including Gilles de la Tourette's syndrome start as habit. This habit progresses to become involuntary and self perpetuating.

The fact that this patient who was not compliant with her medication was

able to achieve complete remission within ten weeks of treatment further buttresses this assertion.

The patient had these symptoms for two years during which she suffered psychologically, socially and academically.

The imperative of a high diagnostic acumen is emphasized.

Human movement involves an astonishing complex interaction of the brain, nervous system and muscles; even a simple action such as picking up a biro engages several different parts of the brain²⁰. The conscious thought areas of the brain trigger the motor area to send signals to the muscles of the arm, as the movement begins, sensors in the arm are activated, sending signals back into the different areas of the brain that interpret them and then send further messages to the motor area to fine tune power, speed, coordination and balance²⁰. With this level of complexity, it is not surprising movement disorders occur.

Other conditions in which movement disorder occur are stereotypic movement disorder²¹, Parkinson's disease, parkinsonism, Parkinson-plus syndromes, Huntington's disease, Wilson's disease, inherited ataxias, essential tremors, restless leg syndrome, dystonia, cerebrovascular accident, cerebral palsy, encephalopathies, intoxication, poisoning by carbon monoxide, cyanide, methanol or manganese²².

The diagnosis of Gilles de la Tourette's syndrome is a clinical one. Brain imaging techniques and other investigations are done to rule out other causes of movement and sound disorder.

Management of this syndrome includes behavioural interventions such as habit reversal techniques, massed (negative) practice, self



monitoring, incompatible response training, and relaxation techniques². Drug management includes high potency typical antipsychotics like haloperidol, the atypical antipsychotics like olanzapine, alpha-2 adrenergic agonists like clonidine; other drugs that are of use are physostigmine, clonazepam and fluoxetine^{2, 23-28}. Prognosis is guarded².

CONCLUSION

The benefits of early presentation and early appropriate referral are obvious. The role of behavioural intervention in the management of Gilles de la Tourette's syndrome may supersede pharmacotherapy. There is need for clinicians to document properly and publish uncommon disorders.

ACKNOWLEDGEMENT

I thank the almighty God. I dedicate this article to Professor O. Morakinyo from whom I got the inspiration to write it. I know I would have enjoyed the privilege of his superlative supervision while writing this article, if he was not on leave.

REFERENCE

1. Enersen OD. Georges Albert Édouard Brutus Gilles de la Tourette. <http://www.whonamedit.com>. Assessed 8th August, 2008.
2. Sadock BJ, Sadock VA. Tic disorder. In: Kaplan and Sadock's synopsis of psychiatry Behavioural Sciences/Clinical Psychiatry 9th edition. Cancro R, Edmonson J, Gabbard GO, Grebb JA et al (ed). Lippincott Williams & Wilkins 2003; 1246-1250.
3. Lees AJ: Georges Gilles de la Tourette: The Man and His Times Revue Neurologique Paris 1986; 142(11): 808-816.
4. Shapiro Ak, Shapiro E, Wayne HL, Clarkin J, Bruun RD. Tourette's syndrome, Summary of data on 34 patients. Psychosom Med. 1973; 35(5): 419-435.
5. McHenry LC: Samuel Johnson's tics and gesticulation. Journal of historical medicine and allied science, 1967; 22: 152-168.
6. World Health Organisation. International Statistical Classification of Diseases and Related Health Problems, 10th edition (ICD - 10) 1992.
7. Jagger J, Prusoff BA, Cohen DJ, Kidd KK, Carbonari CM, John K. The epidemiology of Tourette's syndrome: a pilot study. Schizophr Bull 1982; 8(2): 267-278.
8. Jerkins RL. Ashby HB. Gilles de la Tourette's syndrome in identical twins. Arch Neurol. 1983; 40(4): 249-251.
9. Comings DE, Comings BG, Devor EJ, Cloninger CR. Detection of major gene for Gilles de la Tourette's syndrome. Am J Hum Genet. 1984; 36(3):586-600.
10. Fahn S. A case of post-traumatic tic syndrome. Adv. Neurol. 1982; 35: 349-350.
11. Bleeker HE. Gilles de la Tourette's syndrome with direct evidence of organicity. Psychiatr Clin (Basel). 1978; 11(3): 147-154.



12. Pulst SM, Walshe TM, Romero JA. Carbon monoxide poisoning with features of Gilles de la Tourette's syndrome. *Arch. Neurol.* 1983; 40(7): 443-444.
13. Cohen DJ, Shaywitz BA, Young JG, Carbonari CM, Nathanson JA, Lieberman D, et al. Central biogenic amine metabolism in children with the syndrome of chronic multiple tics of Gilles de la Tourette: norepinephrine, serotonin and dopamine. *J Am Acad Child Psychiatry* 1979 18(2): 320-341.
14. Bremness AB, Sverd J. Methylphenidate-induced Tourette's syndrome: case Report *Am J Psychiatry.* 1979; 136(10): 1334-1335.
15. Sleator EK. Deleterious effects of drugs used for hyperactivity on patients with Gilles de la Tourette's syndrome. *Clin Pediatr (Phila).* 1980; 19(7):453-454.
- 16.16. Klempel K. Gilles de la Tourette's symptoms induced by L-dopa. *S Afr Med J.* 1974; 48(32):1379-1380.
- 17.17. Klawans HL, Falk DK, Nausieda PA, Weiner WJ. Gilles de la Tourette's syndrome after long-term chlorpromazine therapy. *Neurology.* 1978; 28(10):1064-1066.
18. Health communities. Movement disorders. <http://www.healthcommunities.com>. Assessed 8th August, 2008.
19. Golden GS. Tics and Tourette's: a continuum of symptoms? *Ann Neurol.* 1978; 4(2):145-148.
20. Macnair T. Movement disorders. <http://www.bbc.co.uk>. Assessed 8th August, 2008.
21. American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders. 4th ed. Washington, DC: American Psychiatric Association; 1994.
22. Robinson R. Movement disorders. *Movement disorders.mht.* Assessed 8th August, 2008.
23. Shapiro AK, Shapiro E, Wayne H. Treatment of Tourette's syndrome with haloperidol, review of 34 cases. *Arch Gen Psychiatry.* 1973; 28(1):92-97.
24. Rose MS, Moldofsky H. Comparison of pimozide with haloperidol in Gilles de la Tourette's syndrome. *Lancet.* 1977; 1(8002):103.
25. Cohen DJ, Detlor J, Young JG, Shaywitz BA. Clonidine ameliorates Gilles de la Tourette's syndrome. *Arch Gen Psychiatry.* 1980; 37(12):1350-1357.
26. van Woert MH, Rosenbaum D, Enna SJ. Overview of pharmacological approaches to therapy for Tourette syndrome. *Adv Neurol.* 1982; 35:369-375.
27. Stahl SM, Berger PA. Physostigmine in Tourette syndrome: evidence for cholinergic underactivity. *Am J*



Psychiatry. 1981; 138(2):240–242.

28. Gonce M, Barbeau A. Seven cases of Gilles de la Tourette's

syndrome: partial relief with clonazepam: a pilot study. Can J Neurol Sci. 1977; 4(4):279–283

