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CASE REPORT

Gilles de la Tourette syndrome co-existing with Attention Deficit Hyperactivity Disorder in a 9-Year old Nigerian boy

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Abstract

Tourette disorder is an uncommon neuropsychiatric disorder characterised by involuntary tics and behavioural disturbances. The disorder is most prevalent in childhood. Only a few cases of Gilles disorder had been reported in Nigeria. The disorder shares some characteristics and association with many neuropsychiatric disorders hence, the diagnosis can be easily missed. This is a report of a 9-year old school boy with Tourette disorder with co-existing Attention Deficit Hyperactive Disorder (ADHD). The diagnosis in this child was missed on initial assessment, hence this report highlights the need for high index of suspicion when children present with involuntary tics.

Keywords: Gilles de la Tourette, ADHD, Psychosis, Misdiagnosis

Introduction

Tourette syndrome or Tourette disorder has been described as a neurological disorder characterised by repetitive, stereotyped involuntary movements and vocalisations (usually called tics).[1, 2] Behavioural disorders are strongly associated with this disease,[3] named after Dr Gilles de la Tourette, a French neuropsychiatrist, who carried out extensive research on patients with the disorder in 1885.[1, 2] The onset of symptoms is in childhood, beginning from the age of 3years, although other reports put the age of onset at about 7 years of age but generally, before the 18 years of age.[1-4] Tics have a higher prevalence among non-Hispanic whites. [3] Epidemiological surveys in the western world suggested that the disease is commoner among males compared to the females in the ratio 3-5:1. [3] The overall lifetime prevalence is estimated to be 4-5/10000 and approximately between 5 and 30 out of every 10,000 children will have this disorder. [1]

Till date, the precise aetiological basis of Tourette disorder remains largely unknown. However, recent research has suggested abnormalities in the frontal lobe of the brain, the brain stem and the motor cortex. [1] There is also a strongly associated genetic link. [5]

The symptoms usually begin suddenly and can be grouped into the motor and vocal tics. Verbal or motor tics can be re-classified as either simple or complex. [1,2,4,6] Simple motor tics involve muscles around the head, face and neck, such as eye blinking, facial grimacing and head or shoulder jerking. This can progress to more complex tics involving multiple groups of muscles. Similarly, vocal tics can also be simple when it produces sounds such as grunting, barking or sniffing and can also progress to more complex vocal tics such as repetitive words, phrases and outbursts of obscenities or irrelevant speeches in about 10-15%

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of cases. [3] Tics are exacerbated by anxiety states.

In Nigeria, the prevalence of the disorder is unknown as epidemiological surveys are nonexistent. Tourette disorder is often thought to be rare, although, some cases have been reported which highlighted the need to have a high index of suspicion when evaluating patients with movement disorders as the disease can be readily misdiagnosed. [7-8]

The case of a 9-year old Nigerian boy with a history of involuntary tics is reported to illustrate the need for clinicians to have a high index of suspicion in order to avoid misdiagnosis of Gilles de la Tourette syndrome when attending to children with behavioural and motor tics.

Case Description

AS was a 9-year old primary four pupil of a public primary school in Lagos, Nigeria. The boy presented with a year history of repetitive shaking of the head and shrugging of shoulders which were noticed at school and at home. Initially, the parents thought he was forming a habit and had beaten him on occasions in attempts to discourage the act but the abnormal jerky movements of the head and shoulder persisted. These abnormal movements only abated during sleep. The jerks and twist seemed to make him restless and he was always on the move. There was no history of fever. Due to the increased jerks, the parents sought medical attention at a clinic six months into the illness. Presumptive diagnoses of severe malaria and enteric fever were made.

Within the first week of treatment with intravenous penicillin and combination antimalarial drugs, the jerks persisted, prompting the change of diagnosis to seizure disorder. Therefore, the medications were changed to oral Carbamazepine 200mg daily. In the second week of treatment, the boy developed propulsive grunts and occasional shouts which were brief and unpredictable. The vocal tics became more complex with sudden and brief outbursts of irrelevant speeches. The head and shoulders jerks also continued in intermittent bursts. At that point, the boy was referred for psychiatric evaluation, with a diagnosis of psychosis (childhood schizophrenia) made by the referring physician in the fourth week of the illness.

The boy was the third child in a family of six; the father was a trader while the mother was a civil servant. The birth history was uneventful and he was delivered per vaginam at term. He was described by his parents and teachers as a very active child, occasionally aggressive and had problems paying attention in class and at home. His academic performance was described as average. There was no past history of chronic medical illness, and no history of emotional, sexual or social abuse. There was no known family history of mental illness or movement disorders.

The mental state examination, of the boy, revealed a well-nourished, uncooperative, inattentive, active (was unable to sit down for 10 minutes at a stretch) with sudden repeated intermittent jerks in the face and shoulder shrugs usually lasting between 30 and 40 seconds. The speech was occasionally inconsistent and there were no abnormalities in perception. Physical examination did not reveal any abnormality in the central nervous system or other body systems. The Full Blood Count (FBC) parameters were within the normal limits and the Computed Tomography (CT) Scan of the Brain revealed no structural abnormalities. The Electroencephalogram (EEG) revealed paroxysmal spikes mostly in the frontal region.

A diagnosis of Gilles de la Tourette syndrome and Attention Deficit Hyperactivity Disorder was made by the Psychiatrist. The patient was treated with oral Risperidone 2mg daily and Carbamazepine was increased to 400mg daily in two divided doses. The parents were subsequently educated on the nature of the disorder, and the course of the disease. The symptoms resolved significantly after three weeks of treatment with the aforementioned medications. The boy resumed schooling and was reportedly calmer and more attentive in class. Presently, AS is being followed up at the medical out-patient clinic and the mental state was adjudged to be stable at the last evaluation.

Discussion

Gilles de la Tourette syndrome is a neuropsychiatric disorder that can mimic other neuropsychiatric conditions as our case showed. This is due partly to the fact that the disorder can also co-exist with other neuropsychiatric conditions such as seizure disorders and Attention Deficit Hyperactivity Disorder (ADHD). In about 50 -60% of cases, Tourette disorder is associated with ADHD while other researchers have reported as high as 90% association in children with Tourette disorder. [1, 3, 9] This strong association may be due to the similar pathophysiology around the basal ganglia and motor cortex as earlier suggested. [2, 10] The occasional brief motor tics in the present case, were initially misdiagnosed as infective diseases (such as malaria and enteric fever), seizure disorders and childhood psychosis. This highlights the need to be highly circumspect when motor tics and vocal tics are observed in children aged three years and above.

Motor tics, in Tourette disorder, are characterised by brief, repetitive, movements of muscle groups. Consciousness is usually preserved and the tics may not significantly affect their functionality unlike the situation in seizure disorders. On the other hand, the brief intermittent vocal tics may lead to a mistaken diagnosis of childhood psychosis. Indeed, motor tics typically precede vocal tics in Tourette disorder. Additionally, the nature and history of progression from simple to complex tics may help with differentiating Tourette disorder from childhood psychosis. [11] Tics are also different from myoclonus and choreiform movements which may be rhythmic or symmetric. The latter forms of abnormal movements are not associated with premonitory urge or subjective relief following the action. They are not predictable, reproducible or suppressible and do not usually present as complex movements. In addition, they also do not occur in bouts and coprolalia is absent. [11]

Although intellect is affected in Tourette disorder, many cases can reasonably do well in school, particularly with a supportive environment. [12] Unfortunately, we did not conduct an IQ test to ascertain objectively the level of intelligence of our patient due to operational constraints. However, our case illustrated the importance of parent-teacher cooperation in the management of children with Tourette disorder. It is highly important for clinicians to seek information regarding school performance and teacher's observations during the clinical evaluation of suspected cases. ADHD, as co-morbidity in this patient, may have preceded the onset of Tourette disorder. It is auspicious to search for associated co-morbidities among children with features of Tourette disorder.

A multi-pronged treatment approach was adopted for the index case. Antipsychotics are recommended in the treatment of Tourette disorder. [1-3] We chose the atypical antipsychotic, Risperidone, due to the low propensity to cause movement disorders (Extrapyramidal side-effects) and sedation relatively to typical antipsychotic drugs. We also maintained Carbamazepine therapy as an adjunctive treatment due to the non-availability of methylphenidate in our center. Methylphenidate would have been the ideal agent to treat the co-existing ADHD. In addition, it is also known that seizure disorders may also be associated with Tourette disorder. [11]

The diagnosis of Tourette disorder is clinical hence, laboratory investigations are not required. In addition, EEG abnormalities have no consistent pattern in Tourette disorder but can be a useful tool in excluding seizure disorders. [13] The clinical course of Tourette disorder is such that the disorder remits with increasing age, particularly in post-pubertal age. [2-3] In very rare situations, the disorder may persist into adulthood. [3-5] As shown in the case of AS, tics may also abate during sleep or focused activity or be exacerbated in anxiety states. [1] Parent education is essential for the continuation of medical therapy and to alleviate psychological stress on the child and parents. Frustrations encountered by parents with this presumably "errant behaviour" can lead to unnecessary beatings and punishments which only increase anxiety states and serve to increase the tics. Treatment must be holistic and encompassing the psychological adjustment difficulties in the environment of the patient.

Consent: The consent to use the particulars of the patient was given by the parents.

Authors' contributions: ATO participated in the clinical management of the patient and drafted the manuscript while OOO participated in the literature search and drafting of the manuscript.

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