La Prensa Medica Argentina

Case Report

Tourette's Syndrome Following Severe Head Trauma Sustained during Adolescence: A Case Report

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Rec date: 21 July, 2014 **Acc date:** Nov 18, 2014 **Pub date:** Nov 22, 2014

Abstract:

Tourette's Syndrome (TS) is known to be a neuropsychiatric syndrome that manifests itself mainly in children and adolescentswith tics, and is frequently associated with behavioral problems.

Neurophysiological dysfunctions linked to neuro-developmental pathology of the basal ganglia and the associated neuronal network has been a focus of interest in the study of TS. It has been suggested that TS may result from a defect or damage in this area, caused by various risk factors on neurotransmission. Possible accused causal agents include: genetic factors, trauma at birth, concussion, stroke, infections and psychotropic drugs. Studies have shown that approximately one in three TS patients do not acquire this disorder genetically. In more severe cases of TS, reports have revealed incidents of trauma, dating back to the childhood of patients.

In this study, we present two cases of TS, one male and one female patients with no known previous medical history of TS. According to the medical history and to the medical records available, the tics and other symptoms have developed in the adolescence of both patients, following severe head trauma in severe road traffic accidents.

These two cases support the view of multi-factorial aetiology in TS. We have emphasized the impact of severe head trauma on neuro-psycho-development process in childhood and adolescence in TS.

Keywords: Tourette's syndrome; Severe head trauma; Tics; Adolescence

Introduction

TS is a childhood or adolescent onset neuropsychiatric developmental disorder, characterized by the presence of multiple motor tics and one (or more) vocal tic, which tend to fluctuate in intensity [1,2]. The prevalence of TS is estimated at 0.5-1,5 % in children and adolescents [3,4,5,6].

Motor tics occur in the form of grimacing, eye movements, blinking, jerking or shrugging. Vocal tics include recurrent coughing, throat clearing or noisily breathing in and out. The majority of patients develop mild to moderate tic disorders; however, even mild tics may lead to severe psychosocial problems in daily life [7]. Complex tics occur as coordinated movements of several muscle groups. Complex motor tics can also be combined with grimacing, head turns and shrugs. Other motor tics may actually appear to affect selective movements such as, hopping, jumping or body rotations. More complex vocal tics include coprolalia or echolalia. Tics are usually most severe in childhood and often significantly reduced during sleep -though are not completely suspended [8,9].

The precise neuropathology and the mechanisms of TS are not yet known. The majority of published papers support that TS is an inherited developmental disorder of synaptic neurotransmission, however, the precise genetic abnormality responsible for the phenotype is not clear. Studies have shown that approximately one in three TS patients do not acquire this disorder genetically. There are people in whom the symptoms of TS occur immediately after a traumatic event, such as an insult to the basal ganglia, severe traumatic brain injury, viral infections, certain drugs or toxins [7,8,9].

Current thinking about the likely mechanism of TS is being reshaped. TS is primarily biological in origin, with stress-diatheses interactions often playing a role in the course of the illness [10,11]. Environmental factors including severe head trauma are, however, implicated in the pathogenesis of TS. Strong evidence of this has been provided by functional neuro-imaging studies showing abnormalities within dopaminergic systems of the basal ganglia, the caudate nucleus and prefrontal cortex. Dysfunction within these circuits results in an inability to suppress unwanted movements, behaviors, or impulses [12,13,14].

"Secondary Touretism" has been named for certain neuro-pathological changes affecting the brain in childhood or in adolescence [15,16]. These usually emerge after damage to the basal ganglia, influencing the dopaminergic connections to the prefrontal cortex. Suggested causal agents include: head injury, encephalitis, sudden withdrawal from old neuroleptic drugs and streptococcal infections [17-20]. These varying paths to a similar Tourette's picture suggest that TS may represent a multi-factorial pattern of responses in the brain, including variations occurring in normal development, rather than a unique disorder [21- 25].

The majority of those affected could have other psychiatric co-morbidity. Attention deficit hyperactivity disorder and obsessive-compulsive disorder, learning disabilities, auto-aggression and sleep problems are the most common comorbid psychiatric disorders. These co-morbid disorders could badly affect the course of the disease, and the overall outcome may be most closely related to the course of the co-morbid disorders [26].

The prognosis for uncomplicated cases tends to be relatively good. There are few studies of clinical outcomes that encompass the life span of individuals with TS. Clinical impressions suggest that the severity and the waxing-and-waning course often diminish with each passing decade of adult life [27]. There is no effective treatment for every case of tics. Drug therapy is usually limited to those patients in whom

symptoms cause impairment in daily life [28, 29]. Recently, transcranial magnetic stimulation and deep brain stimulation have been recommended, as an alternative to drug treatment [30, 31].

Material and Methods

Two cases of Tourette's Syndrome are presented here, in patients with no known previous history of TS, following severe head trauma.

A diagnosis of TS was made according to DSM-V [32], in which the TS was described by the following symptoms:

Both multiple motor and one or more vocal tics are present at some time during the illness, although not necessarily concurrently. (A tic is a sudden, rapid, recurrent, non-rhythmic motor movement or vocalization).

The tics may wax and wane in frequency but have persisted for more than 1 year since first tic onset.

Duration of at least one year and the onset is before 18 years of age.

The disturbance is not due to the direct physiological effects of a substance (e.g. cocaine), or a general medical condition (e.g. Huntington's disease, or post-viral encephalitis.)

Case 1

History: HE, the 50-year-old woman suffered, at the age of 13, a severe head trauma and multiple injuries, after a serious road traffic accident in Germany in 1977. She was in a coma for several days in an intensive care unit at a local hospital. She received several in- and out-patient treatments for physical, as well as mental, complaints in the years that followed.

After the accident, certain mental and neurological complaints, such as obsessive-compulsive symptoms, as well as tics, began to develop, while no pre-existing mental or physical complains were identifiable according to her anamnesis and medical records.

Since 2005, she regularly attended our psychiatric community practice. She was treated for prevailing complaints, such as obsessive-compulsive symptoms, behavioral problems and tics.

She described suffering from distinctive obsessive thoughts and aggressive compulsive behaviors. On a number of occasions, she reported suffering from compulsive thoughts when holding sharp objects like knives, scissors or glass, with a desire to stab or harm somebody; therefore she had to avoid these types of objects, even touching them. At times, she suffered from suicidal thoughts. She often suffered an irresistible urge to touch her nose with her fingertips, and from time to time, demonstrated phonetic utterances and grimacing. She lacked confidence around other people and avoided situations that lead to an increase in tics and compulsions. Her restricted affect modulation, anxiety and depressed mood with sleep disorders and sometimes suicidal thoughts, dominated the clinical appearance. She expressed that these symptoms had developed gradually after the accident.

Having assessed the evidence based on reviewing the extensive medical reports and opinions, we think that there is a direct causal link between the road traffic accident and the development of TS.

Case 2

History: According to his statement, MS was born in 1959 in Turkey as the youngest of three children. At the age of 16, he was involved in a deadly road traffic accident while travelling with his local soccer team. Despite several deaths he survived from his injuries and head traumas.

He was treated in the local hospital for his coma and injuries. In the following years, he received various hospital treatments for his physical recovery but not for his mental health. In the next few years, he began to develop tics; especially a coprolalia. His tics and coprolalia worsened while he was in the military service. He injured a fellow soldier by stabbing him over a joke made in reference to his symptoms. Despite a referral to forensic medicine, establishing the correct diagnosis failed and he was, subsequently, made to serve 3 years in jail.

The symptoms continued to prevail over the years that followed. MS, at the age of 29, came to Germany, and received treatment for his mental complaints in various psychiatric practices and hospitals. He has regularly attended our community psychiatric practice, for the past 5 years. His complaints reduced and the mental symptoms improved significantly with medical treatment, as well as cognitive behavioral therapy.

MS suffered from typical echolalia, -when someone coughed he would also cough, and from coprolalia, -swearing as a reaction to people offering him a handshake. In addition he suffered from distinctive obsessive thoughts and aggressive compulsive behaviors. At times, even homicidal thoughts occurred when other people failed to appreciate his problems. He suffered from insecurity, fear and anxiety when in crowded environments, which could trigger an undesirable display of his symptoms. He lacked confidence and distrusted foreign people around him. He had, therefore, adopted a strategy for avoiding situations that led to an increase in tics and compulsions. He demonstrated restricted affect modulation with insecurity, anxious and depressed mood with avoidance behavior. He experienced fears constantly, leading to coprolalia.

MS expressed strongly that he suffered no neurological or mental health conditions before the aforementioned accident. Evidence based there showed no known pre-existing obvious neuropsychiatric disorders. MS has insisted on a causal link between his mental illness and the accident of 1975. Also, he expressed that he had received no psychiatric care until he was in Germany.

Discussion

We presented two patients with TS, whose symptoms both emerged after severe road traffic accidents. According to the medical history, both patients suffered severe head trauma in their adolescence. Both patients have pronounced no known previous history of TS.

These two cases support the idea that TS can be associated with environmental factors rather than purely genetic aetiology -which has been the claim of many authors over the last 100 years. We also wanted to encourage similar studies to come forward, to question the hundred-year old 'genetic inheritance' theory in the aetiology of TS [33,34]. The precise influence of the environment on this basically biological disorder is difficult to ascertain, particularly when TS is complicated by comorbidities [35].

It is not surprising that there are many ambiguities about TS. In reality, there are few agreed-upon facts. It is also unknown whether TS is one entity or a heterogeneous condition [36,37]. The diagnosis is mainly based on recognition of clinical appearances. There is no single test to prove its presence or to exclude it [1,2]. One should also exercise caution against making any generalizations about TS. Greater awareness of the disorder may be necessary in the identification of simpler and milder cases. Unfamiliarity of TS by physicians and falsely defining the tics may also contribute to its misdiagnosis [21,38]. These problems illustrate some of the difficulties facing professionals in the field and those attempting to understand the growing literature on the subject [39,40].

Our presentation has several limitations; firstly, given that the aforementioned accidents took place in the 1980s, our assessments and assumptions rely purely upon the medical history of our patients, (-as well as the existing medical notes.) However, no radiological reports or recordings were available to refer to. Secondly, the number of cases is too small to prove our views regarding the aetiology of TS. Our observations, however, encourage further studies to be carried out, in order to help clarify the role of trauma in the development of TS.

Conclusion

Although the exact causes are unknown, genetic and environmental factors were suggested to play a role in the etiology of TS. In view of similarities in clinical presentation, we assume that TS caused by genetic etiology and Tourette-Like-Syndrome (or secondary Touretism) resulted from environmental factors, are similar medical conditions, but with multiple etiology. Further study is, however, needed to confirm the exact relationship between these two conditions.

In addition, our observations and the findings support the need for testing the hypothesis of the environmental origin of TS.

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