

# COMPLEX PHONIC TIC AND DISINHIBITION IN TOURETTE SYNDROME

## Case report

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**ABSTRACT** - Tourette syndrome (TS) is a neuropsychiatric disorder characterized by a combination of multiple motor tics and at least one phonic tic. TS patients often have associated behavioral abnormalities such as obsessive compulsive disorder, attention deficit and hyperactive disorder. Coprolalia, defined as emission of obscenities or swearing, is one type of complex vocal tic, present in 8% to 26% of patients. The pathophysiology of coprolalia and other complex phonic tics remains ill-defined. We report a patient whose complex phonic tic was characterized by repetitively saying "breast cancer" on seeing the son of aunt who suffered from this condition. The patient was unable to suppress the tic and did not meet criteria for obsessive compulsive disorder. The phenomenology herein described supports the theory that complex phonic tics result from disinhibition of the loop connecting the basal ganglia with the limbic cortex.

**KEY WORDS:** Tourette syndrome, coprolalia, complex phonic tic, obsessive compulsive disorder.

Tique fônico complexo e desinibição em síndrome de Tourette: relato de caso

**RESUMO** - Síndrome de Tourette (ST) é uma condição neuropsiquiátrica caracterizada pela combinação de múltiplos tiques motores e ao menos um tique fônico. Frequentemente tiques se associam a distúrbios de comportamento como transtorno obsessivo compulsivo e déficit de atenção e hiperatividade. Coprolalia, definida como emissão de obscenidades, é um tique fônico complexo presente em 8% a 26% dos pacientes com ST. A fisiopatologia de tiques complexos permanece mal compreendida. Nós descrevemos um paciente com tique fônico complexo caracterizado por dizer repetidamente "câncer de mama" ao encontrar primo cuja mãe sofria dessa doença. O paciente não conseguia suprimir o tique e não apresenta transtorno obsessivo compulsivo. A fenomenologia desse paciente sustenta a teoria de que tiques fônicos complexos resultam de desinibição da alça límbica dos núcleos da base.

**PALAVRAS-CHAVE:** síndrome de Tourette, coprolalia, tique fônico complexo, transtorno obsessivo compulsivo.

Tourette syndrome (TS) is a neuropsychiatric disorder with onset usually in the first two decades of life characterized by a combination of multiple motor tics and at least one phonic tic. Tics are often associated with behavioral abnormalities such as hyperactivity and attention deficit disorder, obsessive compulsive disorder and others. Characteristically there is a temporal fluctuation of the type and severity of symptoms and signs<sup>1</sup>. The findings of a study of a large kindred with tic disorder showing that 30% of patients with tics were not aware of their existence and that just 18.5% of patients sought medical care for this condition indicate that the majority of patients in the community are not disturbed by the

clinical features of TS<sup>2</sup>. However more severe forms of TS may cause profound emotional, social and professional distress.

Phonic tics may be simple sounds (simple phonic tics such as grunting or sniffing) or whole words (complex phonic tics). In the latter case they are called complex phonic tics whose most characteristic example is coprolalia (swearing or emission of obscenities), present in about 26% of patients with TS<sup>3</sup>. It must be emphasized that although possibly the most characteristic symptom of TS<sup>4</sup>, coprolalia is not mandatory for the diagnosis of this condition. The pathophysiology of coprolalia and other complex phonic tics remains ill-defined. Older studies suggested the

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possibility that these tics are produced by random contractions of pharynx and larynx muscles<sup>5</sup>. However more recent investigations indicate instead that they result from disinhibition of the neural loop connecting the basal ganglia with limbic areas of the cerebral cortex<sup>3,6,7</sup>. In this study we report a patient followed up at the Movement Disorders Clinic of the Federal University of Minas Gerais whose clinical phenomenology provides additional support to the latter hypothesis.

### CASE

At age 13 this 26 year old man developed multiple motor tics (excessive blinking, grimacing, tooth grinding and copropraxia). His parentes also noticed a decreased attention span as well as inability to remain still for very short periods of time and in the next year the patient started shouting obscenities repeatedly. Since that time he remains doing multiple simple and complex motor and phonic tics. There is a temporal fluctuation of the severity and type of tics presented by the patient but he never stopped having copropraxia and coprolalia. He also has had disabilities in his personal, educational and social life. Because of poor performance, he dropped out of school without finishing high school. Despite many attempts, the patient has failed to keep a job and he is currently financially supported by his parents. His pregnancy and delivery were uneventful and prior to the onset of the tic disorder the patient had not had any significant health problem. The family history is remarkable for the father, personally examined by one of us (FC), having a tic disorder which meets criteria for TS<sup>8</sup>. On a recent visit to the Movement Disorders Clinic of the Federal University of Minas Gerais, the most important findings on the neurologic examination were excessive blinking, facial grimacing, copropraxia and another complex motor tic characterized by jumping several times when sat on a chair or the examination table. The patient also shouted a gutural simple sound and had coprolalia. A semi-structured psychiatric interview failed to disclose obsessive compulsive symptoms but the patient met DSM-IV criteria for attention deficit and hyperactivity disorder and major depressive disorder<sup>8</sup>. The episode which led us to report on this patient happened two years ago. At that time one maternal aunt of his already known to have breast cancer was diagnosed with brain metastasis of this neoplasm. On the occasion, every time our patient met the son of this aunt, who happened to be his best friend, he repeatedly and loudly said "breast cancer". This situation persisted through the demise of the aunt a few months later. When asked to elaborate on the mental process during the performance of this complex phonic tic, the patient informed that, on seeing his cousin, the expression "breast cancer" immediately appeared in his mind. Despite all attempts to suppress its vocalization, the patient always failed to do so. This process was associated with severe anxiety that he attributed

to the fear of hurting the feelings of his friend. At that time the patient also did not meet criteria for obsessive compulsive disorder although remained with symptoms of hyperactivity and attention deficit disorder. Treatment with neuroleptic failed to suppress this complex phonic tic.

### DISCUSSION

In this study we describe a patient with a peculiar vocalization, breast cancer, triggered by meeting a cousin whose mother suffered from this condition. The patient was not able to suppress the emission of the expression whose performance was associated with severe anxiety.

The first point to be discussed regarding this case report is whether the vocalization represented a complex phonic tic or a compulsion. It must be emphasized that the lack of a diagnostic biologic marker for either conditions occasionally renders difficult the differentiation between the two of them<sup>9</sup>. The lack of obsessions and compulsions and the patient's failure to meet diagnostic criteria for obsessive compulsive disorder strongly suggest that the vocalization was indeed a complex phonic tic. The vocalization of our patient is similar to "non-obscene complex socially inappropriate behavior", found in 41% of patients with TS in one study<sup>6</sup>. Offensive vocalizations such as racial slurs and others were among the inappropriate behavior identified in this investigation. Interestingly and supporting the hypothesis that our patient's vocalization corresponds to a complex phonic tic, in this study there was no relationship between the described phenomenology and obsessive-compulsive disorder.

This case report also gives insight into the mechanism underlying complex phonic tics, including coprolalia. The description of the mental process experienced by the patient during its performance clearly indicates that the first phase consisted of an external stimulus triggering the appearance of the word (the vision of the cousin leading to association with the disease of the aunt). Subsequently the patient could not refrain himself from vocalizing the word despite all attempts to stop it. Finally, the realization of the failure to prevent the emission of the vocalization was associated with anxiety. Evidently the first stage of this process (association of the cousin with the breast cancer of his mother) corresponds to a phenomenon expected to happen in healthy human beings. The pathology, however, is related to the posterior inability to suppress the vocalization. This finding strongly suggests that the complex phonic tic of our patient is related to failure of

an inhibitory mental process. The result of one previous study of our group showing that the content of coprolalia corresponds to obscenities commonly pronounced by subjects without TS in a determined culture (ie, the normal subjects and TS patients speak the same obscenities but the latter do this more often and in an inappropriate context) further supports the hypothesis of coprolalia and other complex phonic tics being generated by a disturbance of inhibition<sup>3</sup>. As emotionally charged behaviors are related to the limbic system, one could speculate that the underlying anatomic basis of complex phonic tics, coprolalia in particular, would be a hyperactive limbic cortex-basal ganglia loop. Indeed, recent neuroimaging investigations support such hypothesis. In one study with functional magnetic resonance<sup>10</sup>, the authors concluded that patients with TS failed to suppress tics because of an impairment of the ability to suppress subcortical neuronal activity. More recently and with better technical conditions but similar methodology, Stern et al<sup>7</sup> found that one patient with severe coprolalia had hyperactivity of Broca's area and the frontal operculum (obviously related to production of speech), several temporal lobe areas (probably associated with hearing of the speech), areas involved in the motor control of the speech (thalamic and cerebellar structures) and others that may be relevant to emotional linguistic material: head of the caudate and posterior cingulate areas. The dorsolateral prefrontal cortex, also found to be hyperactive in this and other patients with others tics, may be important to understanding the external trigger of the vocalization in

our patient since this area is known to play a role in the generation of movements cued by sensations. Interestingly, clinical observations indicate that TS patients have a tendency to display stimulus-induced behaviors<sup>11</sup>.

In conclusion, we describe a patient with TS who presented with a complex phonic tic whose phenomenology supports the concept that this particular feature of the syndrome is related to disinhibition of the limbic loop of the basal ganglia.

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