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Gilles de la Tourett's Disease
a Single case study
A Discussion on Aetiology and Treatment

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Gilles de la Tourett's disease is a chronic and relatively rare condition characterized by multiple motor tics, compulsive utterance and echolalia. It was first described by Itard in 1825. Many years later, at the Salpetriere, George Cilles de la Tourette (1899) called the disease "Maladie des tics compulsifs". He distinguished it from the large number of conditions which previously had been grouped under the heading of chorea and he defined it as *a nervous afflication characterized by motor incoordinationaccompaniedby echolalia.*

The rarity of the syndrome is exemplified by the fact that in the Mayo clinic, this diagnosis has only been made in seven patients from a total of approximately one and a half million patients admitted from 1935 to 1965.

Further evidence for the rarity is Fermando's review of literatures in 1966. He reported only sixty five cases of this syndrome. This author also reported four cases in 1967.

This paper will deal with one case of this syndrome which is of interest from two points of view.

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1. This patient is a woman and it has been reported in the literatures that this syndrome is very rare in the female sex.
2. The etiology of this syndrome is unknown up to the present time, and various authors have suggested different responsible factors none, of which have been completely approved. This variability of etiological factors is due to the importance of one factor and the loss or lack of importance of other factors in reported cases. But the interesting point of this case is concomittance of different etiological in the one patient. So, this case history may throw light on the etiology and pathology of this syndrome.

Mrs. A.M. age 36 yrs was admitted to Roozbeh Hospital Tehran, on the 10th Sept. 1975 on account of trembling of some parts of her body together with clapping of her hands and jerking of her shoulders. At the same time she was groaning and calling the names of her husband's brother and others, such as Ali and Hassan, at the rate of 50-60 times per minute.

This disordered behavior first began when she was 8 years old. (28 years ago). The onset of this kind of behavior was preceeded by an attack of ringworm of the scalp.

Additional symptoms such as tantrums, irritability and fatigue began at the same time and still persist. She has only been free of these symptoms for 2 or 3 days at a time during the whole 28 years.

Over the years she has been under the care of numerous specialists and has been given psychotherapy, tranquillisers, insulin therapy, E.C.T., etc. but without success, except for a 6 month remission, about 15 years ago.

At the present time her symptoms have become more exaggerated, and in addition over the last 5 years when she hears cats or dogs, she imitates them. And when she hears the number 8 she repeats it several times. The abnormal movements of her jaw and shoulders and the clapping of her hands which appeared at the beginning of the illness are almost always present and become more prominent when she is upset or angry and occurs when she is alone or in the presence of her immediate relatives. In the presence of strangers she can control herself,

unless she is irritated or becomes angry. However, if the presence of strangers become prolonged she is unable to maintain control and the symptoms appear. When she has some companies, she appears altogether normal. She screams, at times, when sleeping. When she first falls asleep she has some jerking of her legs. All her symptoms are more prominent at the time of her menstruation.

Family History

Father. 9 years age he died of a cerebral tumor, at the age of 45 years. He was a bad tempered man, rigid, strict and a perfectionist towards his daughter (the patient). By profession he was a civil engineer and employed by a company. He had no history of addition and other abnormalities.

Mother. Her mother is 55 yrs of age, a housewife and with a quick tempered personality. For the last few years she has been under treatment for a duodenal ulcer. She had two spontaneous abortions one before the birth of the patient and other after her sixth off-spring. The parents of the patient are first cousins.

Siblings: The patient is the second child of the family. The first child died of measles at the age of one year, before the birth of the patient. The patient has got two healthy younger brothers and three sisters. Briefly, except for the perfectionism of her father and the quick temperedness of her mother, and sibling rivalary with her younger brother there is nothing noteworthy regarding her family relationships.

Medical Notions Concerning Other Relatives

The cousin of the patient's mother is mentally sick and probably suffers from a degree of mental subnormality. The patient's maternal grandmother used to have attacks of weakness and convulsions at times of domestic upset. The patient's cousin's son (maternal side). Suffers from epilepsy.

Personal History

Her mother, whilst carrying the patient, at 4 months gestation suffered Malaria and was treated with quinine. The patient was subsequently born by normal delivery, and had an uneventful childhood. She was breast-fed. When she was born she was small and pale, and for the first month of life had only mother's milk. From the second month onwards she was given a syrup containing opium. At the age of six months she had both diarrhea and malaria. Growth and Development have been without any abnormality.

Childhood Events

Until the age of 8 which coincides with the onset of the illness, there is nothing noticeable.

During her childhood she used to have many bad dreams and nightmares. She began biting her nails at this time and still does. She also complained of pains in her legs and this still persists.

She started school at the age of 7 but was a poor scholar and had failed examinations 3 times. When she had ringworm she was absent from school for two years. She completed 8 grades and finally left school at the age of 19. She had no interest except she liked swimming and table-tennis. For 4 years she attended piano classes and enjoyed it.

Sexual and Marital History

Her Menstruation began at 13 years of age after that she has premenstrual back-ache and some abdominal pain, sometimes she felt pain at the end of menstruation.

She first heard a few things about sex and relationships with men, from her classmates, she had a vague fear of sexual relationships, but at fifteen years of age she began to stimulate herself, and this continued until a few years after her marriage. Her feelings about being a woman, as she says, "At times I wish I were a man, because if I were a man I would be less restricted and do those things that I want to."

She is the same age as her husband, they are cousins. They were engaged for 8 months but before they had had some correspondence for 5 years. At the beginning of their marriage they did not understand each other, as the patient says "My husband had a pessimistic attitude towards me and he always criticised me for my behavior. For example "Why did you look there? " "Why did you smile at that person?" and so on. But as a whole we had a good life together." Her husband says "I loved her but I didn't know anything about her sickness." (obviously the husband's tolerance has greatly lessoned.)

From her sexual life point of view she can be classified as frigid, she has got two healthy children, a 12 years old girl and 10 years old boy.

The Patient's Personality

Her husband believes she is completely careless, nothing is important to her, but she is extremely sensitive and very quickly upset. These characteristics however, are not constant, up to the point that sometimes, she does show some interest in her household activities, but then again she becomes inattentive to them. Her feeling and affection to the other members of her family are similarly fleeting.

She is very jealous, up to the point that her husband does not dare to mention the name of another woman. When she feels good she is obedient, but when angry takes no notice. She is selfish and egoistic, and pays no attention to anyone's advise, suggestions or comments, but at the same time she lacks selfconfidence, and likes to be backed-up and supported by others.

Present mental state

At the first contact she is a well-dressed young woman, who gives intelligent replies to questions. At the beginning of the interview she appears apprehensive but gradually she overcomes it.

Altogether, in this interview, there is nothing wrong except for the following negative factors.

a. To some extent she is sad and her mood is constantly

changing.

b. Her general knowledge is rather weak and she is not particularly good at calculations.

c. She is obsessive about cleanliness.

d. She states that she is tired quickly after some slight activities. She declares she has trembling in her body and shaking of her knees. The only abnormality evident during her interview, is that she claps her hands, and that she makes some inspiratory and expiratory grunts. The grunts are associated with some abnormal movements of the body. Speech is normal.

e. She had bad dream and interrupted sleep.

Physical Examination

On physical examination, apart, from the abnormal movements mentioned above, and symmetrical exaggeration of tendon reflexes there are no abnormalities in general.

EEG Rythm 10-11/sec, frequency normal, symmetrical pattern and during rapid respiration some theta waves appear and under intermittant light stimulation occasional pointed waves in the occipital areas can be seen. On the whole the EEG is within normal limits.

Psychological Testing

Her intelligence is below the normal range, her personality is immature with hysterical characteristics and apprensive reactions.

Discussion

The Gilles de la Tourette syndrome is a puzzling disorder, and no specific neurological or psychological aetiology has been found for this condition.

Many theories concerning the aetiology of Tourette's disease exist. Some authors believe an organic disturbance of the brain underlies the symptoms. However neurological evidence is lacking, at least in most of the cases. Others (Chalas) believe that psychological factors,

perhaps acting on an organic substratum are responsible for the symptoms.

Psychodynamically, the condition seems to develop under conditions encouraging repression, the adjustment appears to progress from repressed tension with psychoneurotic overflow into motor tics, then involuntary inarticulate vocal protest develops which gradualy evolves into involuntary coprolalic tics. These coprolalic expressions may then be used on voluntary level in a conscious nostile maneuver to needle be used on a voluntary level in a conscious hostile maneuver to needle or, in a words of one patient, to "goose" a resented parent. The progression is from the intra-punitive to the extra-punitive and from psychoneurotic-like to psychopathic-like symptoms and behavior. In other words as Fernado has hypothesized the onset of coprolalia in patients with ersistant childhood tics indicates a disturbance patiens with persistant childhood tics indicates a disturbance of normal balance between a need for tension relief by swearing and capacity to control such vocal activity.

As opposed to these psychodynamic interpretations some autors have recently investigated a hereditary pre-disposition and organic factors influencing the etiology of Tourett's disease. Partick B. Friel in a recent study (June 1973) has reported three cases of first familial occurence of this rare condition and has concluded that Gilles de la Tourette's disease might be hereditary. Recent advances in the field of brain chemistry suggest the condition may be due to hyer-activity of the dopaminergic systems (the catecholamines, norepine-phine and dopamine) in the corpora striata. This theory would explain similarity of the tics to those observed as a side effects in parkinsonian patients treated with L-dopa.

From a treatment point of view, the unique efficacy of Hallo-preidol, a potent central dopaminergic blocking agent, in controlling the symptoms is another reason for suggesting a subcortical involvement.

This patient's illness is of particular interest because the cause of her complaint is mainly of social origin. In the family history there is

one person with mental deficiency and another with epilepsy. It seems that the whole family have neurotic personalities and hence an inherited factor may be involved. The patient's mother had malaria and was treated with quinine whilst she was carrying the patient. When the patient was 6 months old, she also had malaria causing convulsions. The brain damage is even now evident on electro-encephalography which together with her present level of intelligence, both indicate CNS damage.

It is obvious that, from the point of view of psychology, she grew up and developed in an unhealthy and unsuitable family environment.

Apart from these points, there are some provocative emotional factors to be considered. When she was 8 years old she saw a fire, and around this time had a bout of tinea capitis and the resulting ugliness was psychologically traumatic.

In spite of the combination of all these causative factors which are found in this patient, and the similarity of them to those found by other workers, it is unsufficient evidence to explain the pathogenesis of this illness, but careful study of this patient confirms the CNS damage causing the illness. Most probably the inherited factors and the aquired organic disorders cause impairment of CNS function as regards intelligence, learning and adaptation. In addition, the constitutional disorders within the patient's family and the combination of all the above mentioned factors in the patient, have a reciprocal effect upon each other and aggravate the whole situation. If Tourette's syndrome is to be considered as a sort of reaction against adaptation to the unhealthy environment, this reaction and its continuity can be attributed to permanent CNS damage, most probably in the area of the corpora striata, beginning in childhood.

Whatever the cause, we have to find some means by which to relieve the signs and symptoms and alleviate the situation. To this effect some treatment procedures and various measures have been taken. For instance, psychoanalysis, and behavior therapy, have in a few patients given a satisfactory result. However, direct suggestion,

electro-shock and various types of chemothraphy have been continued for a relatively long period and it can be assumed that the improvement is part of the illness and that none of the above mentioned forms of therapy have any specific effect upon the cause of the illness," Ascher and his co-workers have concluded that the tics, as a whole do not have a clear prognosis and those patients having treatment resulting in some improvement are no greater than those showing spontaneous improvement. Here the obsessive background, especially oral tics (Tourette's syndrome) carries a poorer prognosis.

Recently a few reports indicate that Haloperidole may be effective in this condition. The dose prescribed in 2 to 5 mg three times a day, and as long as the patient takes the drug their symptoms are controlled.

Summary

A case of Gille de la Tourett's syndrome is reported and discussed in the light of conflicting views on the aetiology of the condition. It is hypothesized that if Tourette's syndrome is to be considered as a sort of reaction against adaption to an unhealthy environment, this reaction and its continuity can be attributed to permanent CNS damage (Probably in the area of corpora striata) beginning in childhood. Treatment with haloperidol is suggested as a most effective method of symptomatic treatment.

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