GILLES DE LA TOURETTE'S DISEASE

By

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IN its classical form this peculiar disease is characterized by an irresistible compulsion to swear, together with motor tics. First recorded by Itard in 1825, it was later fully described (1885) and followed up (1899) by Gilles de la Tourette, whose name has been given to the condition. Associated with the compulsive utterances are multiple motor tics, hence Convulsive Tic is a synonym for the malady. The motor movements are more severe and widespread than those met with in simple tic, and they may involve the whole body in a violent spasm. Facial grimaces, violent jerks of the head, limbs and trunk may occur, often combined with an explosive utterance. This exclamation may consist of an unorganized sound such as a bark or a groan or a grunt. Or it may be a word—frequently of an obscene nature (coprolalia) which greatly embarrasses the patient. Other features of the condition are echolalia —the impulse to repeat sounds or words which the patient hears, and echopraxia—the compulsion to copy the actions of others.

The onset of the disease usually occurs in childhood with multiple motor tics. Later compulsive sounds occur, leading on to obscene utterances, and features of an obsessive-compulsive neurosis may be present. All writers give a uniformly bad prognosis for the condition. Kinnier Wilson (1955) states that the only case he tended personally ended in insanity. This was also the experience of Gilles de la Tourette. Of 15 cases reported by Ascher (1949), Heusher (1953), and Mahler, Luke and Daltroff (1955), four developed schizophrenic psychoses.

The condition is rare, and until 1957 there had been no recent recorded cases in the English literature. However, Michael (1957) gave a good description of a case recently treated in the Maudsley Hospital. This patient was a man of 28 who developed tics at the age of 7. At first labelled chorea, it was later diagnosed as habit spasm at Great Ormond Street Hospital for Children. At 9 the patient made involuntary noises, and at 13 explosive swearing commenced. He was symptom free for two years (18–20) while in the Army. Then his symptoms gradually returned, and finally he was unable to control his violent swearing. He became a recluse, unable to work or to leave his home. At the Maudsley Hospital, psychoanalytically orientated psychotherapy was ineffective, but CO_2 inhalation therapy was very helpful. He had four treatments per week, totalling 30 treatments. The coprolalia and motor tics disappeared, and only a residual vocal tic like a hiccough remained. When reported the patient had been back at work for 15 months.

In view of its extreme rarity in England I am prompted to report the following two cases, recently seen, which conform to the description of Gilles de la Tourette's disease.

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Case History: 1

The patient is a man of 65 who complains of compulsive shouting, together with violent body jerks. His first symptom occurred at the age of 5 when he started school. It consisted of facial tics, mainly grimacing, and it was soon followed by jerking movements of the arms. The onset of the symptoms, according to the patient, occurred after he was forced to use his right hand at school, though he was naturally left-handed. He is now ambidextrous and left-footed. He never stammered. His sister's child is left-handed. At 6 he attended Great Ormond Street where a diagnosis of chorea was made. At 12 he came under the care of the National Hospital, Queen Square, where a diagnosis of tic was made. He was treated under Gordon Holmes and Kinnier Wilson with "complete isolation, cold douches down the spine, and faradism". Unable to attend school from 6 to 12 owing to his violent tics, he nevertheless learned to read and write and it was clear that he was above average in intelligence. At about 8 he developed a vocal tic—"just a little pip sound". This gradually increased in intensity over the next 4–5 years, but at 14 he improved considerably and he started work. At 18 the tics had subsided sufficiently for him to take up the violin. At 19 he was called up into the Army (1916), but he was soon discharged because of his vocal and motor tics. He became a master electrician and worked for 32 years with the Eastern Electricity Board, with whom he reached the responsible position of electrician foreman. During most of this period he managed to keep his symptoms under control. With an effort he would keep his vocal tics down to a minimum, though occasionally a loud "bark" would occur.

His illness showed remissions and exacerbations over the years. Symptoms were marked in 1940, and again were severe in 1951. At this time he attended St. Thomas's Hospital where motor tic was diagnosed and myanesin was prescribed, but with no effect on his symptoms. Early in 1958 there was a severe exacerbation of the malady and he again attended the National Hospital. His vocal tic had now assumed the proportions of a loud roar. Anyone unprepared for the noise was liable to be startled by it. The roaring bark was occurring every 5 to 10 minutes during the day. Treatment by tracheotomy was suggested to reduce the noise, and even leucotomy was considered. However, no surgical treatment was carried out.

The recent history is that for the past 18 months symptoms have been more severe than ever before. His roaring bark rendered his life at work, and indeed anywhere outside his home, most embarrassing. Due to their violence and uncontrollability, his symptoms forced him to retire from work and he became a recluse, hardly daring to leave his house. The main symptom was a sudden roaring, barking sound, produced by a violent exhalation against a partially-closed glottis. The sound is loud enough to be heard from several hundred yards distance. Sometimes, when commencing to speak, the first word of the sentence would be shouted violently, usually in a distorted form. However, it was only in this circumstance that his vocal tic involved words. Otherwise the exclamation was an unorganized sound, loud, roaring or barking, often emanating from the corner of his mouth, with the head turned to one side. He looked and sounded like a man swearing, though in fact he was not doing so. He felt no compulsion to swear, but the roaring utterance was frequently irresistible. Sometimes he was unable to speak for fear of shouting.

Combined with the loud utterances, but also occurring independently, were violent sudden movements of the body. These were usually symmetrical, involving mainly the shoulders, arms and head. However, anterior abdominal wall muscles, leg, and facial muscles were all occasionally thrown into violent spasm.

Symptoms were always worse when he was under stress, or worried, or emotionally disturbed. Symptoms never occurred during sleep, neither did they occur during general anaesthesia for tooth extraction. Alcohol always improved the condition, and when he had to leave his house a glass of beer made it easier for him to control the vocal tics. Under certain conditions symptoms were always worse, for example at the barber. His dentist protested that extracting a tooth was more unnerving for him than the for patient in view of the loud accompaniment. It also had an unfavourable psychological effect on other patients in the waiting room next door.

He could sometimes, but not always, suppress the symptoms by an effort of will, but he would feel an "inward tremble" until he jerked violently or made his loud sound. He would then feel temporarily relieved for a short time until the next spasm occurred. He was never thrown to the ground by his spasms. Resting quietly in a chair, or relaxing in bed always reduced the spasms. However, they occurred as often when he was alone as when he was in company. The worst time of the day was when he left home for work, and also when undressing at night to retire to bed.

The patient is married and he has two children, one of whom is described as "highly strung". His father was a heavy drinker who died of tuberculosis, and his mother is described as a very anxious, worrying type. Of his two brothers one drinks heavily, and the other died of a stroke at 52. His only sister is well.

The patient's personality is of obsessional type. He is fastidious, accurate, precise, neat and tidy, and a conscientious worker. His present mental state is dominated by a paranoid, suspicious attitude. He suspects that he is disbelieved when he tells his story. He frequently emphasizes that he has "proof", for example, that he was in the Army, or attended the National Hospital. He will not go outdoors if he can avoid it because he feels everyone looks at him and talks about his peculiar shouting. This, of course, is probably true. But it has resulted in a hostile, aggressive and defensive attitude towards society.

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The patient was treated, empirically, with chlorpromazine. Commencing on 25 mg. t.d.s., the dose was increased from 50 to 75 mg. t.d.s. There was a rapid and remarkable response to this drug. The motor spasms became much less frequent and the vocal tics almost completely disappeared. He found the courage to leave his house. He visited his doctor's surgery without giving his usual warning barks, usually audible before he reached the house. He queued for a bus in silence. At the time of reporting this case the patient has been on the drug for six weeks and he is almost symptom free. For 18 months prior to the exhibition of chlorpromazine his symptoms had been worse than ever before. Only occasionally does he shout or have motor tics now. A day or more may pass without a single spasm, whereas previously vocal and motor spasms were occurring every five minutes or so.

Case History: 2

This patient is a man of 67 who complains of violent, spasmodic movements of the head and limbs, with grunting, barking and coughing exclamations. His symptoms commenced at the age of six with head shaking, followed by spasmodic jerking of the limbs. Later he developed involuntary exclamations. He was diagnosed as suffering from chorea at Great Ormond Street. There was a slow increase in his symptoms up to the age of twenty, after which he improved slightly. But from thirty onwards his symptoms have gradually increased in intensity. His present condition is that he has frequent, violent, sudden jerking movements of head, face, arms and legs. At the same time he makes an exclamation—usually a loud "Oh!" or "Ah!" Sometimes the words "No! no! no!" are shouted. As in the first case, words are only exclaimed at the beginning of a sentence, when they are usually produced in a distorted form. Frequently he disguises the exclamation behind a cough. His symptoms are always worse when he is excited, and they disappear during sleep. He is a rent collector and he rides his bicycle regularly without difficulty. His symptoms have never stopped him from working. Though he can voluntarily suppress his symptoms for a short period, he says that they "Come out in a rush of jerks and noise" eventually.

The patient is a very conscientious, hard-working man who has held one job for 31 years. His personality is of obsessional type, and he has some features of an obsessional neurosis, including folie de doute with frequent checking and rechecking his actions. His father had a tic consisting of blinking, and he stammered. The patient is right-handed.

In 1952 he was treated with electro-convulsive therapy at a General Hospital. This had no effect on his symptoms. He then had narco-analysis and psychotherapy, again without effect. Recently the patient was seen at the National Hospital where a diagnosis of Convulsive Tic was made.

As in the first case, the patient was treated with chlorpromazine, but unfortunately after a few weeks he developed chlorpromazine jaundice, and the drug had to be discontinued.

Discussion

The remarkable similarity in the histories of these two patients is notable. By a coincidence both patients were seen within the space of a few weeks in the same general practice. Possibly the condition is not so rare as has been thought in the past. The excellent response to chlorpromazine in Case 1, and the early signs of improvement in Case 2 suggest that this drug should be the first line of treatment.

Both patients have many features in common with Michael's case (1957). In all three cases the onset was in childhood with multiple motor tics diagnosed as chorea. Vocal tics developed later. Then there was a latent period with abatement of symptoms, and a later recurrence of the condition in a more serious form. The natural history of the disease is marked by exacerbations and remissions, and a psychotic termination is a recognized outcome, as was mentioned earlier. In this connection, the paranoid features present in Case 1 are significant.

In considering the aetiology of the condition it is noteworthy that the majority of cases described in the literature have had features of an obsessive-compulsive neurosis. In view of this one must consider whether the vocal exclamations are compulsive components of such a neurosis. Alternatively, they may be tics. In a compulsive neurosis there is a conscious compelling impulse to perform a certain action. A motor tic (or habit spasm), on the other hand, is a much less organized action, presumably involving lower motor centres. It is frequently involuntary, but may be controlled with an effort.

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Purves-Stewart and Worster-Drought (1952) define a tic as a co-ordinated purposive act, perpetuated as a recurrent involuntary movement, which occurs automatically, without conscious attention being directed to it. An obsessional compulsion, on the other hand, absorbs the patient's attention during its performance, and is accompanied by a feeling of irresistible obligation to perform it.

However, the difference between a tic and a compulsive action is not so obvious when it involves speech. The main differences here are:

1. Voluntary inhibition of a compulsion produces anxiety; inhibition of a tic does not produce anxiety.
2. It is frequently difficult and sometimes impossible to inhibit a tic, whereas a compulsion can nearly always be inhibited. In other words a tic may become involuntary, whereas a compulsion is always voluntary.
3. There is always some psychological symbolism underlying a compulsive action, whereas a tic is a more primitive less organized reaction with little or no symbolism.

The relationship of Tourette's disease to chorea is interesting and important in that it suggests that there may be an organic basis to the condition. Several of the cases described in the literature have been diagnosed as suffering from chorea in childhood. It will be noted that this diagnosis was made at the age of 6 at Great Ormond Street in the cases here described. Tics are well known to occur after chorea, and Lewis (1956) points out that Tourette's disease also may follow chorea. Turner and Critchley (1925) described the respiratory tics that may occur during or following encephalitis lethargica. The tics include yawning, sniffing, coughing and spitting. In addition, simple motor tics occur. The vocal tic of Tourette's disease involves the respiratory musculature in a very similar manner to these post-encephalitic tics.

Summary

Gilles de la Tourette's disease is described, and a further two cases of this rare condition are presented. Treatment with chlorpromazine, not previously described, was remarkably effective.

The prognosis is usually poor, the disease often terminating in psychosis.

The aetiology is reviewed, and the differences between tics and compulsive neurosis are discussed.

The occurrence of tics after organic brain disease suggests that there may be an organic basis to this disease.

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