

Senile onset Tourette's syndrome

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INTRODUCTION

Gilles de la Tourette's syndrome (named after a French neurologist) is a relatively rare neuropsychiatric disorder of childhood that presents with fluctuating vocal and motor tics.

Tics are sudden, brief, intermittent, involuntary or semi-involuntary movements (motor tics) or sounds (phonic or vocal tics). Many patients remit in adolescence, but in a minority the syndrome is a lifelong disorder. It is rarely encountered for the first time in the elderly and so far only two cases have been published (Sutula and Hobbs, 1983; Filman et al, 1991). Interestingly, the first well-described patient reported by Itard in 1825 was of an 85-year-old man (Brunn, 1988).

Tourette's syndrome can be primary (idiopathic, inherited) or secondary (as a result of head injury, stroke, infections such as encephalitis, carbon monoxide poisoning or drug-induced (stimulants, levodopa, anticonvulsant and neuroleptic), Chouinard and Ford, 2000). Both genetic and neurobiological links have been found between autistic spectrum disorders and Tourette's syndrome (Rapin, 2001).

Vascular dementia is the second most common form of dementia after Alzheimer's disease. It can be caused by a single strategic infarct, multiple large infarcts, lacunae or white matter lesions.

This article describes an unusual case of vascular dementia presenting with Tourette's syndrome.

DISCUSSION

Although transient tics are a common occurrence in childhood, the prevalence of Tourette's syndrome is 0.7% (Comings et al, 1990). However, first onset of Tourette's syndrome in old age is extremely rare. Tics are the hallmark of Tourette's syndrome.

Motor tics could be simple (i.e. blinking, twitching, grimacing, jerking) or complex (head shaking, trunk bending, touching, making lewd or obscene gestures (copropraxia) or echopraxia).

Simple phonic tics typically consist of snuffling, throat clearing or grunting while complex phonic tics include shouting of obscenities, profanities or otherwise socially inappropriate words (coprolalia). Coprolalia is the most recognizable and certainly the most distressing symptom of Tourette's syndrome. A significant proportion of patients with Tourette's syndrome suffers from distressing co-morbid psychiatric disorders. The most common of these are attention deficit hyperactivity disorder, obsessive-compulsive disorder and depression (Jankovic, 2001).

Neurobiological studies in Tourette's syndrome have implicated the basal ganglia and the related cortical and thalamic structures, especially the cortico-striato-thalamo-cortical circuits that concurrently subserve a wide variety of sensorimotor, oculomotor, cognitive and limbic processes (Chappell et al, 1990; Peterson, 2001). Lesions in this region also explain the co-morbid psychiatric disorders like attention deficit hyperactivity disorder and obsessive-compulsive disorder.

Although white matter lesions commonly occur in old age, periventricular

CASE REPORT

A 78-year-old ex-fireman who had a history of recurrent depressive disorder since 1995 was admitted to the psychiatric ward in December 2000 with a relapse of depression and obsessional thoughts of harming his wife. Over the 18 months before this admission, there was a suspicion that he might be suffering from cognitive decline and there were episodes of grossly disinhibited behaviour completely out of keeping with his usual personality.

He was living at home with his wife and had two grown-up children who lived away. His medical history indicated that he had hypertension, arthritis and prostatism. There was no family or childhood history of tics.

During his stay on the ward it was noticed that he would repeat words under his breath. There were spontaneous outbursts of shouting and swearing (he would keep saying the word 'shit'). He was aware of these outbursts and was very apologetic but was unable to control them. The outbursts would disappear in his sleep or while he was concentrating on pleasurable activities (for example watching cricket on the television) but would increase under periods of stress. He also started developing various motor tics such as blinking, nose twitching, spitting and grimacing. He had recurrent intrusions of the word 'evil' in his mind. He also had thoughts about hurting or stabbing his wife, which were obsessional in nature (he realized that the thoughts were irrational and tried to control them but failed to do so).

Neurological examination revealed mild dysarthria, soft pyramidal signs on the left side and frontal release signs. Neuropsychological examination revealed mild dementia (mini mental state examination score= 21), deterioration of his performance on the Weschler Adult Intelligence Scale (IQ score fell from 91 to 56 in 2 years) with significant impairment of frontal lobe functions (as tested on Behavioural Assessment of Dysexecutive Syndrome). A computed tomography scan of head revealed mild generalized atrophy but with marked periventricular ischaemia. Single photon emission computed tomography scan showed bilateral reduction in perfusion to anterior cerebral hemisphere.

He was treated with paroxetine (20 mg) and a small dose of haloperidol (0.5 mg twice daily). His shouting and swearing gradually settled, his mood lifted and he was discharged to an elderly mental illness residential home where he is currently doing well.

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white matter lesions were reported to occur in vascular dementia 11.6 times more frequently than in Alzheimer's disease and 3.5 times greater than in healthy people (Boone et al, 1992). Patients with periventricular white matter lesions commonly present with a particular cognitive profile that includes a deficit in frontal or subcortical function. DeCarli et al (1994) point out that this pattern is the result of lesions in the same cortico-striato-thalamo-cortical circuit as is hypothesized to be involved in Tourette's syndrome.

The previous two reported cases of senile onset tics did not have any plausible aetiological factors. To the best of the authors' knowledge, this is the first reported case of senile onset Tourette's syndrome with a definitive aetiology.

CONCLUSIONS

This case demonstrates all the typical features of Tourette's syndrome including co-morbid psychiatric disturbances. It is probably the first documented case of vascular dementia presenting with an unrelated neuropsychiatric disorder. This case also demonstrates that careful observation led to early identification and treatment of a rare but distressing syndrome in an elderly patient. **HM**

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IMAGES IN MEDICINE

The third dimension: it's behind you!

E Britton, IS Shergill, DL Stoker

A 55-year-old man underwent an oesophagectomy through a thoraco-abdominal approach. A postoperative chest X-ray raised alarms that a swab had been retained (see arrow, *Figure 1*). However, the linearity of the findings led to closer external examination of the patient. The epidural site on the patient's back had been held with a surgical swab (radiologically visible) rather than a blue swab (radiologically invisible) as is customary. X-rays serve to provide a two-dimensional view of three-dimensional patients. This third dimension should always be carefully considered when interpreting X-rays. **HM**

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Figure 1. A postoperative chest X-ray showing a linear radio-opacity (arrow).

