SHORT REPORT

Late onset startle induced tics

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Abstract

Three cases of late onset Gilles de la Tourette's syndrome are presented. The motor tics were mainly induced by an unexpected startling stimulus, but the startle reflex was not exaggerated. The tics developed after physical trauma or a period of undue emotional stress. Reflex tics may occur in Gilles de la Tourette's syndrome, but have not been described in late onset Tourette's syndrome. Such tics must be distinguished from psychogenic myoclonus and the culture bound startle syndromes.

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The normal startle response consists of a brief flexion response, most marked in the upper half of the body, elicited by an unexpected auditory, and sometimes somaesthetic, visual, or vestibular stimulus.[1] An exaggerated motor startle reflex is one of the main features of hereditary hyperekplexia,[2 3] but it has also been described secondary to other neurological disorders, such as cerebral palsy,[4] postanoxic encephalopathy,[2] and brainstem abnormalities.[5–9] Compared with the normal startle response these startle reflexes are greatly exaggerated in amplitude and more extensive in distribution. It has been suggested that the startle reflex may be exaggerated in Gilles de la Tourette's syndrome,[10] although this has recently been contested.[11] The issue is compounded by reflex tics, which may particularly occur in response to a startling stimulus.[12–14] Reflex tics may occur in the setting of idiopathic Gilles de la Tourette's syndrome, but have not, to our knowledge, been described in the late onset disease. Here we present three cases of late onset Tourette's syndrome, where startle induced tics dominated. The tics developed after physical trauma or a period of undue emotional stress. Such tics must be distinguished from psychogenic myoclonus and the culture bound startle syndromes.

Case reports

PATIENT 1

This African woman had a road traffic accident in 1988 at the age of 33. She was not rendered unconscious by the impact but noted pain in the neck and right sided weakness the next day. Over the next few days she developed sudden jerks affecting the head, neck, and left arm, with involuntary vocalisations (screams, yelps, and grunts). These were subsequently noted to be startle sensitive as well as spontaneous. They could be triggered by unexpected bright lights or taps. The involuntary movements were more likely to occur when she was angry or stressed. In 1991 coprolalia and kissing tics started. Typically these would come in flurries lasting several minutes and consisted of coprolalia with high pitched yelping noises, facial tics including grimacing and pouting, and jerking movements of the left shoulder and left arm. No obvious urge or build up of tension preceded the spontaneous movements. The suppressablity of the jerks remained unclear. There was no family history of tics or obsessive-compulsive behaviour. Tetrabenazine, but not sulpiride, led to considerable improvement. On neurological examination she had multiple motor tics of the face, jerks of the left arm, and startle induced loud high pitched screams. Neuropsychological studies, EEG, brain MRI, and screening blood tests were normal.

Neurophysiological testing of the auditory startle response (stimulus 90 dB tone) produced an isolated response in the orbicularis oculi muscle (latency 48.6 ms) followed later by complex and variable patterns of muscle activation with mean latencies ranging from 113 ms (mentalis) to 250 ms (forearm muscles). Percussion of the left side of the body produced further complex patterns of muscle activation associated with vocalisation. The latency to onset of the muscle activity after each stimulus was variable and ranged from 80 to 500 ms in different muscles of the body.

PATIENT 2

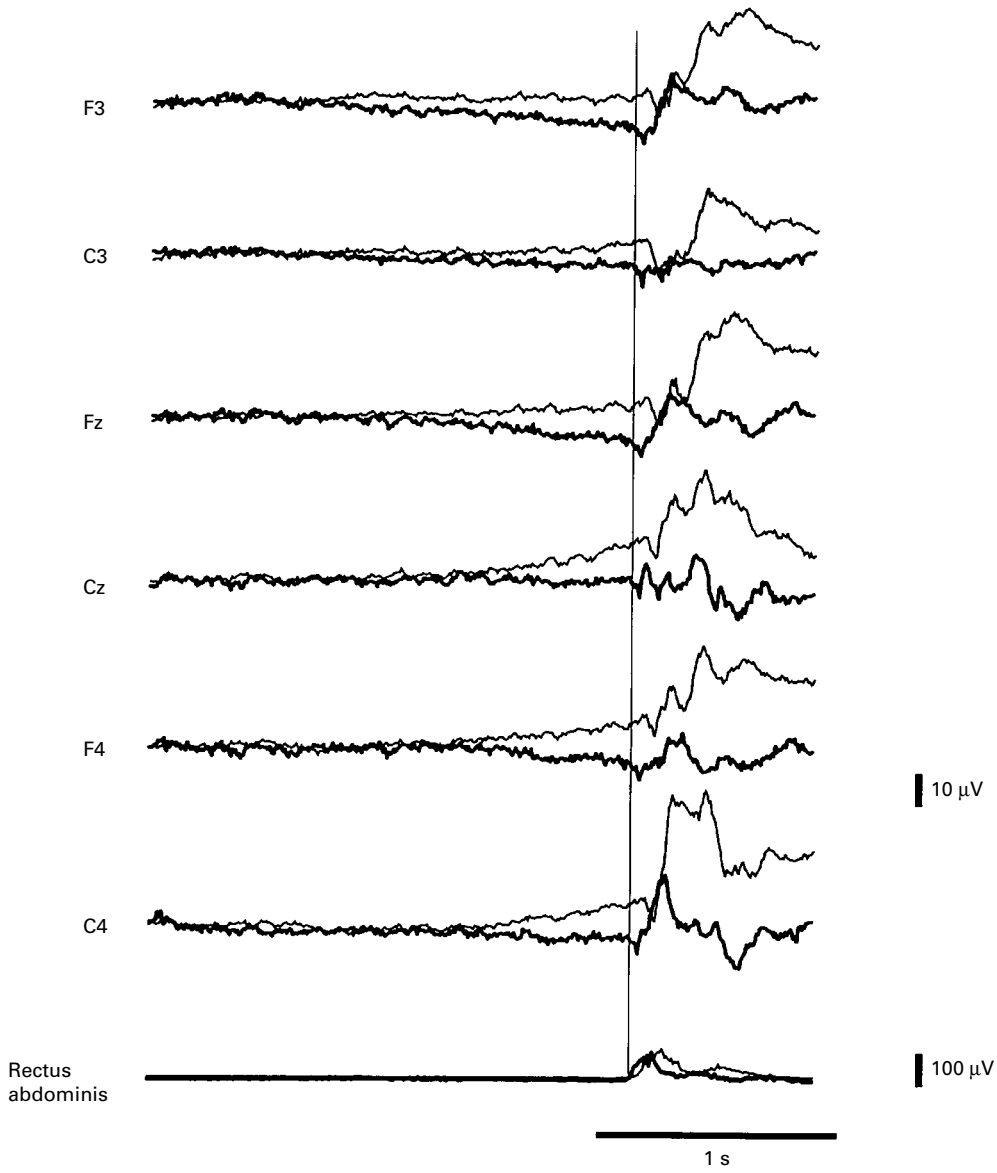
This 52 year old man developed excessive anxiety and panic attacks with hyperventilation during a stressful period. The panic attacks were accompanied by jumps. The anxiety disorder resolved but the jumps persisted. The jumps consisted of flexion of the trunk, hips, and knees followed by twitching of the hands and feet. The jumps occurred spontaneously or were triggered by unexpected touches and tap. They tended to occur in flurries of up to 10 jumps and were occasionally accompanied by grunting. The movements were preceded by a poorly defined subjective feeling and were suppressible. The attacks were virtually absent

when distracted. As a child he had a minor and suppressible repetitive involuntary movement of one leg and an involuntary dancing movement of the fingers when under stress in middle age. Family history was negative for tics or obsessive-compulsive behavior.

On examination, a tap to the jaw, biceps, or quadriceps tendon would set off a flurry of jerks lasting up to 2 minutes. These jerks involved various parts of the body including the face, hands, feet and abdomen and did not follow a stereotyped pattern. The duration of the EMG activity during the jerks varied between 75 ms and 1 s. The earliest jerk was recorded 120–160 ms after a tap to the patella or biceps tendon. Variable jerks would also occur spontaneously when completely relaxed. These included eye blinks, brief flexion movements of the trunk, and hand and feet movements. Suppression of the jerks was followed by an even more marked flurry of jerks. The EEG activity preceding the spontaneous truncal flexion jerks was back averaged. A premovement potential was absent before these jerks, but was present when the patient voluntarily mimicked the same movement (figure).

PATIENT 3

This 32 year old African woman was knocked down by a car. She was unconscious for 3 days. Thereafter she had anterograde amnesia for seconds and post-traumatic amnesia for several months. She had anxiety, panic attacks, and forgetfulness, and complained of right sided weakness and sensory disturbance. A year after the head injury she developed high pitched inspiratory screams associated with facial grimacing and jerky movements of the arms. The screams and movements would occur spontaneously or follow an unexpected and



Premovement potentials in patient 2 preceding spontaneous axial tics (thick trace, average of 42 trials) and voluntary axial flexion jerks mimicking the spontaneous tics (thin trace, average 100 trials). A thin vertical line marks the onset of movement. There is no pre-movement potential preceding the tics. The scalp electrodes were referenced to linked ears.

startling stimulus, such as the slamming of a door. Vocalisations could be temporarily suppressed. There was no definite urge or build up of tension preceding spontaneous jerks and vocalisations.

On examination she had a slightly slurred speech and mild right sided weakness and sensory loss. Tone and tendon reflexes were normal and plantar responses flexor. She had spontaneous vocalisations with inspiratory wheezing sounds associated with facial grimacing and eye closure. Similar vocalisations could be precipitated by an unexpected stimulus such as a tap to the face or loud noise. These had a latency of about 500 ms and were repeated every few seconds. The spontaneous vocalisations were often accompanied by variable head movements or raising of the left arm and shoulder. She also had spontaneous upwards deviation of the eyes. Clonazepam was ineffective. Brain MRI showed bilateral frontal high signal change consistent with her previous head injury. There was no premovement potential before spontaneous vocalisations.

Discussion

We present three cases of late onset Tourette's syndrome, in whom startle induced tics dominated. The tics developed after physical trauma in two patients and a period of undue emotional stress in the third. In patient 2 a pre-existing Tourette's syndrome was possibly unmasked by stress as he had a motor tic and obsessive traits in childhood.This supports the idea of a genetic basis or predisposition to Tourette's syndrome.[15] Post-traumatic tic disorders have previously been seen, but did not include startle induced phenomena.[15–17] There was no family history of tics or obsessive compulsive disorders. Since the onset, there has been waxing and waning of the tics in patient 1. In two patients the tics were suppressible and in one patient tics were preceded by a perceived urge. The tics tended to be complex and even gave the appearance of being distractible at times; yet they were not preceded by a premovement potential in two cases—evidence against a psychogenic movement disorder. The absence of a premovement potential before spontaneous simple tics has been previously reported.[18]

The startle evoked responses in our patients were of very long latency and active muscles had a variable order of recruitment. They were therefore not exaggerated startle reflexes,[1 3] but startle induced involuntary movements. Usually electrophysiological examination is necessary to discriminate between these two types of response. Historically, Gilles de la Tourette[19] (1884) linked Tourette's syndrome to the jumping Frenchman of Maine, latah of Malaysia, and miryachit of Siberia.[20] These culture bound startle syndromes consist of an excessive startle response followed by echolalia, echopraxia, and forced obedience. Whether the startle response in these syndromes is a true exaggerated startle reflex has never been studied. The current opinion on these phenomena is that they are culture specific psychological rather then neurological disorders.[21 22] The occurrence of multifocal tics distinguishes the present cases from these culture bound syndromes.

In summary, reflex tics may develop de novo in middle age, in the setting of physical or emotional trauma. It is important to distinguish them from psychogenic reflex jerks, which they may mimic.

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