CASE REPORT

Gilles De La Tourette's Syndrome

by

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Abstract.

A case of Gilles de la Tourette's Syndrome is presented. This is the second reported case in Indonesia.
The diagnosis was based on four essential features : childhood onset, multiple motor tics, vocal tics and changing pattern of the symptoms.
The exact etiology of this syndrome is still unknown. It is proposed that patients with this illness could have hyperactivity of the dopaminergic systems in their basal ganglia.

Symptomatic improvement has been achieved with haloperidol. The response to this drug could not be evaluated in this patient because of refusal to continue the treatment.

Introduction

This rare disease was first reported by Itard in 1825 and later described as a syndrome by Gilles de la Tourette in 1885.
Although most case reports are from the United States and Europe, the condition is found in all parts of the world (Shapiro et al., 1972). Lieh–Mak et al (1979), reported five cases in China. A case diagnosis as Gilles de la Tourette's Syndrome were reported by Loebis et al. (1981) from Medan.

The exact aetiology of this syndrome is still unknown. The illness is characterized by multiple motor tics, vocal utterances including inarticulate noises and obscene words (coprolalia).

This paper presents the second case of Gilles de la Tourette's Syndrome in Indonesia in an attempt to raise the level of awareness of this condition among pediatricians.

Case.

A 10–year–old Indonesian girl visited the Pediatric OPD Dr. Pirngadi General Hospital in Medan on May 16th, 1983 with complaints of shoulder shrugs, jerking, jumping movements, vocal tics and eye blinking. The complaints of shoulder shrug, jerking and vocal tics was of about one and half months duration. The shoulder shrug and jerking preceded the vocal tics. "Ah" sounds was snapped repetitively as a component of unprovoked vocal tics. These symptoms appeared every five to ten minutes and disappeared during sleep.

Bilateral eye blinking was noticed one year earlier and replaced by shoulder shrug and jumping movement after one month. She was free of symptoms in the 10 months there after.
She never has had head trauma and there was no history of tics in her families.

She was spoiled by her parents and her siblings and was slight hyperactive compared to the other siblings. She was moderately intelligent at school and except for teasing by her classmates because of these symptoms, she never showed any difficulties in social adjustment with her peers.

On physical examination, except for these multiple tics there were no other abnormalities. Neurologic examination was negative.
She was slightly tense and anxious during examination.
Treatment was started with haloperidol 0,5 mg. thrice daily. On follow up one week later, the vocal tics had disappeared and the other tics had decreased. The drug has been stopped by the father because of excessive sedation and refused to continue the treatment.

Discussion.

This patient was diagnosed as having the syndrome described by Gilles de la Tourette based on four essential features : (1) childhood onset (under 16 years of age), (2) multiple motor tics, (3) unprovoked vocal utterances and (4) changing pattern of symptoms.

Shapiro et al (1976) suggested the following criterion required for the diagnosis of this syndrome : (1) Age of onset between 2 and 15 years, (2) Multiple involuntary muscular and verbal tics, (3) Symptoms wax and wane, (4) Slow change in symptoms, usually over a 3 months period, in which old

symptoms may disappear and new symptoms may replace or be added to pre existing symptoms and (5) Chronic, lifelong illness. Coprolalia, copropraxia, echolalia, echopraxia and palilalia are confirmatory symptoms but not essential for the diagnosis. History of hyperactivity in childhood, abnormal EEG, soft neurologic abnormalities and subtle signs of organic dysfunction on psychological testing are frequently concomittants of the disorder but not essential for the diagnosis.

The diagnosis of Tourette's syndrome at the present time is a clinical one. The minimal diagnostic criteria, especially in the pediatric age group seem to be muscular and vocal tics. Motor tics are usually the first to appear and often begin as simple tics involving the face or head. Complex or stereotyped movements may also occur and include jumping, hopping, squatting, smelling or touching especially the genitalia. Vocal tics may consist of sounds or words. Sounds include coughing, hissing, barking, and throat clearing. Stammering, stuttering, or echolalia may occur, or there may be repetitive utterings of partial or whole words. The most dramatic vocal tics is termed coprolalia, the involuntary use of obscene language.

Many aetiological theories have been proposed for Gilles de la Tourette's syndrome. Psychological theories have been proposed previously based on evidence of psychologic factors in patients with this abnormalities. Recent work favoured an organic aetiology in this disorder.
Dopamine is the predominant neurotransmitter in the corpus striatum and putamen. It has been proposed that patients with Gilles de la Tourette's syndrome could have hyperactivity of the dopaminergic system in the corpora striata.

Haloperidol, a butyrophenone with potent neuroleptic activity, especially on the basal ganglia and their connections can alleviate the synptoms of this syndrome.
Haloperidol is a potent dopaminergic blocking agent. This drug has been helpful in most cases. Symptomatic improvement has been achieved with this drug.
It is interesting that haloperidol controls but does not reverse the basic pathology. Cessation of the drug is being followed by relapse. At higher dosages, extrapyramidal symptoms develop. The major side effect at lower dosages is sedation, and this is usually the factor which limits the dose.
The dosage varies between 1 – 180 mg per oral daily. Most children improved with dosages less than 4 mg/day. Dosages larger than 2 mg/day require concurrent administration of an antiparkinsonian agent.

The response to haloperidol cannot be evaluated in our case because the father refused to continue the treatment. Loebis et al. (1981), reported their case being treated successfully with haloperidol in a dose of 3 mg daily. All symptoms were alleviated in two weeks.

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