

Gilles de la Tourette's Syndrome

— Report on 24 Korean cases —

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The data on 24 Korean patients with Gilles de la Tourette's syndrome are described. Main symptoms and other clinical data and EEG findings were generally consistent with those on Caucasian and other Asian patients reported by many authors and reviewers. The effectiveness of haloperidol treatment was also good. These data are thought to suggest organic etiology of this syndrome, but emotional components should not be neglected in understanding and helping patients with varied symptoms other than tic movements.

Key Words : Gilles de la Tourette's syndrome, Korean

Gilles de la Tourette's syndrome an unusual condition characterized by recurrent, involuntary repetitive, rapid movements (multiple motor tics) including multiple vocal tics. Coorplalia, an irresistible urge to utter obscenities, may be present in some cases. Since this syndrome was originally described by Gilles de la Tourette in 1885, many cases have been reported from different cultures over the past 100 years. For many years it was considered to be a psychological illness (Ascher, 1948; Dunlap, 1960). Now evidence has accumulated proving that organic and hereditary factors play a predominant role in the etiology of this multiple tic syndrome (Shapiro *et al*, 1973; Lucas *et al*, 1973; Wassman *et al*, 1978; Pollack *et al*, 1977; Klawans *et al*, 1978; Shapiro and Shapiro, 1981). Up to the present, however,

the varied hypotheses on the etiology have not been confirmed.

This syndrome had not been reported outside of Europe and America until the first Chinese case was reported in 1963 (Singer, 1963). The retrieval of bibliographical list, of related articles since 1966 through the Australian Medical Literature Analysis and Retrieval System (MEDLARS), revealed only one report on 5 Chinese with this syndrome (Lieh-mak *et al*, 1979) among hundreds of Caucasian cases throughout the world. The authors of this report attributed the scarcity of reports from Asia to the rarity of the syndrome, the lack of awareness of the condition and the lack of pressure to publish.

In Korea, the first case of Gilles de la Tourette's syndrome was reported in 1968 (Lee and Sul, 1968). Since then 9 cases have

been reported in various Korean medical journals. Those reports focused mainly on descriptive or psychodynamic aspects of the syndrome.

This report will summarize a study of 24 Korean patients with Gilles de la Tourette's syndrome whom the author has seen over the last 6 years.

MATERIAL AND PROCEDURE

Twenty-four patients have been diagnosed as having Gilles de la Tourette's syndrome in the 6 years since 1976 at Yonsei University Medical Center in Seoul. Eight of the 24 cases were referred by neurologists, neurosurgeons, pediatricians and internists.

All 24 patients were evaluated psychiatrically by the author. History of symptomatology, family history and developmental history were also obtained from semi-structured interviews with the patients and their families.

The diagnostic criteria were, 1) onset of symptoms in childhood, 2) presence of recurrent, involuntary repetitive, rapid, purposeless motor movement affecting multiple muscle groups, 3) multiple vocal tic. Coprolalia, although considered pathognomonic, was not considered necessary for a diagnosis of Gilles de la Tourette's syndrome (Shapiro *et al*, 1973).

RESULTS

Demographic Data:

All 24 patients were native to Korea. The incidence of this syndrome can be said to be very rare here in Korea, considering that the total number of out-patients of this medical center is about 3.4 million during the same period of 6 years. Nineteen patients were male

and five were female, or a ratio of 3.8:1. The average age of the patients was 13.0 years (S.D. 6.98) and ranged from 5 to 32 years. One of the 4 adults was married.

Past History:

No significant factor was found in the birth history of any of the patients. However, twelve patients had an abnormal developmental history including hyperactivity and distractability in 7 cases, learning difficulties in 3 cases, enuresis in 2 cases, encopresis in one case, and stuttering in one case. In premorbid personality 13 patients were reported to be impulsive, hot-tempered and competitive. Four patients were obsessive and meticulous and 7 patients were polite and shy. From their medical history, 2 patients had suffered from nephritis, 2 patients from dog bite, 2 patient from pulmonary tuberculosis, one patient from a car accident, and one patient from a febrile convulsion. One patient had had a tonsillectomy.

Family History:

Simple tics were reported to have been present in the siblings of 2 patients. But the presence of any form of psychosis was denied by all the families. Ten patients were the first born, 6 had older and younger siblings and 8 were in the younger group of siblings.

The Age of Onset and Duration of Illness:

The syndrome began before the age of 15 in 23 of the 24 patients. One female patient began at age 16. Most patients (5 patients) began at the age of 6. The mean age of onset was 8.14 years, S.D. 2.81 (range 5-16 years): male, 8.10 years, S.D. 2.23 (range 5-12 years), female, 8.30 years, S.D. 4.68 (range 5-16 years). The mean duration of the illness was 4.68 years, S.D. 6.32 (range 1 month to 22 years).

The Initial Symptoms:

The initial symptom was, in most patients

(9 cases), eye twitching with or without vocal tics (2 cases, grimace (2 cases), arm jerk (one case), head jerk (one case). Five cases began with head jerks, 3 cases with limb jerks, 4 cases with vocal tics such as an expiratory sound, throat clearing or snorting, 2 cases with facial grimaces, and one case with jerking of the trunk.

Present Symptoms:

After the initial symptoms, other parts of the body became involved and new tics developed and replaced or were added to older ones. The details are shown on table 1.

Most patients have shown various facial tics as eye blinking and grimacing. Atypical bark was apparent or reported in 13 patients (54.2%). Other vocal tics such as expiratory sound, sniffing, snorting, throat clearing and other meaningless noises, "uh" or "huh", were apparent or reported in 7 patients (29.1%). Coprolalia was present in 9 patients (37.5%). Echolalia was reported by 3 patients. Other rare behavioral symptoms included spitting in 3 patients, tongue biting in one patient, mannerism in 2 patients, and stuttering in one patient. Compulsive touching, jumping in the air and echopraxia were rarely reported. Symptoms were absent during sleep in all patients.

Only 9 cases showed the classical syndrome with multiple motor tics, barking and coprolalia. The other 5 cases had combined multiple motor tics and barking. Seven cases had multiple motor tics and other vocalizations. However, 3 cases had only severe multiple motor tics. According to criteria of severity by Shapiro and his colleagues (1973), 14 patients (58.3%) were rated as having marked and severe illness, which is defined as progressive and chronic illness without remission, and with serious social, academic and occupational impairment.

Psychopathology:

No patients were evaluated as having schizo-

phrenia or other psychosis. However 4 patients, who were older ones, were diagnosed as having reactive depression. One of them, a 17 year old male, showed serious depression with suicidal ideas and agitation. Three children were considered to have an obsessive-compulsive trait. One adult male had typical obsessive-compulsive disorder. Nine children were reported to have adjustment reaction in school life and peer relationships since the onset of the syndrome. The parents of 5 children reported that the personality of their children had changed to be hyperactive, violent and impulsive since the onset of the syndrome.

Electroencephalography and Computerized Tomography:

Twelve (60%) of 20 patients who took EEG, had abnormal findings: random slow wave in 2 cases, random slow wave and sharp wave mixture in 5 cases, mild focal abnormality (bicentroparietal area) in one case and a burst of high amplitude sharp waves in both hemispheres in 4 cases.

Computerized tomography was done in 5 cases and the findings were normal in all.

Psychodynamic Consideration:

Various precipitating factors seemed to reflect some psychodynamic influences. That is, symptoms developed after minor surgery such as tonsillectomy in 2 cases, and after beating by the father, elder brother or school teacher in 3 cases. Seven cases had a definite psychogenic factor such as emotional frustration, conflicts in child-parent relationship and loss of self-esteem. This precipitated the syndrome or modified its severity. One patient scarcely showed the symptoms at school but frequently did at home. Most patients reported that the symptoms were usually aggravated with anxiety, tension and other emotional stress. Seven patients reported anxiety and emotional uneasiness when they tried to voluntarily control or

inhibit symptoms, especially coprolalia in 4 cases. These findings suggest that psychological factors are closely related with the syndrome, if not an etiological factor.

Treatment:

Among previous treatments were anticonvulsant medication in 5 cases under the diagnosis of myoclonic type epilepsy before referral, herb medicine in 5 cases and acupuncture in 2 cases. Those treatments were eventually proved to be ineffective.

After the diagnosis was made as Gilles de la Tourette's syndrome, all patients were treated with haloperidol. The initial dose was usually 0.5 to 1.0 mg per day and gradually increased by 0.5 or 1.0 mg every day or every other day until an effective dose was determined. The effective dosage was different in each patient and ranged from 1.0 to 12.5 mg, a median of 2.5 to 3.0 mg. The effective dosage did not seem to be positively correlated with the body weight, the severity of the symptoms, or the duration of illness or the age of onset. The period of treatment ranged from 3 weeks to 1.5 year.

With haloperidol treatment, symptoms remitted completely in 18 patients and were alleviated in 4 patients after one month of treatment, showing an average improvement rate of 91.7%. Combined treatment with diazepam, 3-8 mg per day, was helpful in 3 cases in which emotional tension was an obstacle to treatment. The side effects of haloperidol were bothersome to many patients. Drowsiness was most common, and dyskinesia was the major cause of discontinuing haloperidol. As various behavioral problems were not as much improved as the tic movements during the period of drug treatment, other psychiatric aids such as counseling were needed.

Twenty patients discontinued medication against medical advice after 6 to 12 weeks of

treatment. Most of the patients themselves or their parents believed that treatment was enough. But 15 patients visited the hospital again after a mean period of 2.4 months because the symptoms recurred. The common cold seemed to be the precipitating factor in 5 cases. In 4 cases, the dosage needed to be increased to be effective as in previous treatments.

DISCUSSION

These Korean cases of Gilles de la Tourette's syndrome support the contention that this syndrome is very rare but not limited to a single race or ethnic group, and that the clinical manifestations are similar as reported by many authors from different countries (Fernando, 1976; Lieh-mak *et al*, 1979). The sex ratio of male predominance, age of onset, non-significance of birth order, and high incidence of abnormal developmental history are consistent with other reports and reviews (Fernando, 1976; Lucas *et al*, 1967; Marphew and Sin 1967; Shapiro *et al*, 1973; Woodrow, 1974).

However, the finding that tics and other psychiatric problems were rare in the family history of Korean patients, is different from the rather high incidence of psychiatric problems in other studies on Caucasian patients which have provided a hereditary or genetic model for the syndrome (Shapiro *et al*, 1973; Wassman *et al*, 1978; Kidd *et al*, 1980). The low incidence of family cases may be due to the lack of painstaking efforts required to obtain this kind of history.

The initial symptoms, present symptoms, severity of symptoms and the clinical course of a wax and wane pattern were also similar to commonly reported cases (Fernando, 1976; Shapiro *et al*, 1973). But the incidence of coprolalia was a little lower compared with the

incidence of about 60% in Caucasian cases. Spitting in 3 cases can be considered as one of abusive behaviors like coprolalia. Woodrow (1974) had suggested that Gilles de la Tourette's syndrome might be behaviorally related to schizophrenia and biochemically related to amphetamine psychosis. However, in conformity with other studies (Lucas *et al*, 1967; Shapiro and Shapiro, 1981), the findings on psychopathology in this study do not support such a relationship.

Incidence of obsessive-compulsive traits was infrequent as Shapiro *et al* (1973) reported. This is in contrast to commonly reported cases in which such a trait was considered as a major personality characteristic of patients with this syndrome (Ascher, 1948; Dunlap, 1960). However, the finding that a high incidence of emotional trauma and associated aggressive hostility as a precipitating factor seems to be consistent with the classical hypothesis of a psychological etiology, namely, that the patients develop reaction formation and obsessive compulsive defenses and traits against hostility, and finally tics as compromise symptoms (Ascher, 1948; Dunlap, 1960). Corbett *et al* (1969) also reported that the vast majority of patients showed increased anxiety and depressive symptoms. Woodrow (1974) reviewed the literature and concluded that symptoms become worse during affective arousal and improve with relaxation. Fernando (1976) reported several patients who benefited from psychotherapy. But recent studies do not support this psychological hypothesis (Shapiro *et al*, 1973; O'Quinn and Thomson 1980). The author's impression is that psychodynamic components play a significant role in the development of symptoms. Tics and, particularly, coprolalia and spitting, can be considered to indicate a disturbance of normal balance between a need for tension relief by swearing and a capacity to control

such activity (Fernando, 1976).

However, such poor impulse control may be related to organic and/or hereditary factors. Elkins *et al* (1980) reported that obsessive-compulsive behavior was more common in those patients with Gilles de la Tourette's syndrome who had a family history of the syndrome or of tics than it was in non-familial Gilles de la Tourette patients. Consistent with this line of consideration, other evidence such as the high incidence of abnormal EEG findings, abnormal behavioral problems and other neurologic signs and the effectiveness of haloperidol (Lucas *et al*, 1967; Shapiro *et al*, 1973; Bruun *et al*, 1976; Waldo *et al*, 1978) suggest organicity in the pathogenesis of the syndrome. In addition, the findings shown from this study that the common cold with fever is associated with the recurrence of symptoms and that the personality changed to one of being impulsive and violent since the onset of the syndrome, also suggest organicity.

Haloperidol was an effective drug for Korean patients as it was for Caucasians and other Asians (Shapiro *et al*, 1973; Bruun *et al*, 1976; Lieh-Mak *et al*, 1979). However one finding in this study showed that Korean patients tend to need less dosage of haloperidol and less frequent antiparkinsonian medication than Caucasians patients who were reported to need dosages up to 240mg of haloperidol and more frequent prescriptions of antiparkinsonian drugs (Shapiro *et al*, 1973). This reminds the author that Tourette patients can therapeutically respond to remarkably low serum level of haloperidol (Singer *et al*, 1981). However, it is possible that Asians have genetically different constitutional responsiveness to drugs. The finding that the effective dosage was unrelated to age of onset, duration of illness, severity of symptoms and body weight, is compatible with the report of Bruun *et al* (1976). This means that

haloperidol appears to be the most effective medication available but the treatment must be individualized. As O'Quinn and Thomson (1980) stated the less known symptoms such as learning difficulties, speech and language problems and other behavioral problems were not as much improved by haloperidol as the tic movements as also shown by our study.

The effectiveness of haloperidol led some researchers to suggest that Tourette's disorder is mediated by catecholaminergic activities (Pollack *et al*, 1977) or hypersensitivity of the dopamine receptors (Klawans *et al*, 1978). Therefore, any drug with catecholamine inhibiting action such as pimozide or clonidine, can be effective (Ross and Moldofsky 1978; Cohen *et al*, 1980). But this catecholamine hypothesis is not accepted by other investigators (Shapiro *et al*, 1981).

All these findings and suggestions mean that the etiology of the syndrome remains uncertain and it should be understood in terms of emotional, behavioral, developmental and hereditary/organic models, and in the evaluation and treatment a holistic approach is recommended including academic help, counseling and other psychosocial aids.

CONCLUSIONS

The data collected on 24 Korean patients with Gilles de la Tourette's syndrome on sex, age, past history and family history, psychogenic factors, clinical manifestations and course, behavioral problems, EEG findings and effectiveness of haloperidol were generally consistent with those on Caucasian and other Asian patients reported by many authors or reviewers. However, the findings were worthy of notice that, in some patients, personality changes developed after the onset of the syndrome and that physical

conditions such as the common cold were precipitating factors. The effectiveness of haloperidol treatment was uniformly good. However, Korean patients seemed to need less of the drug than Causcassian patients did. This clinical data is thought to suggest that the etiology of the syndrome is an organic impairment in the central nervous system or constitutional make up. But emotional components which were revealed to play a significant role through precipitation and aggravation of symptoms, should not be neglected in understanding the varied symptoms of this syndrome and helping these patients, by treatment should be individualized.

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