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Gilles de la Tourette's Syndrome: Report of Five Cases in the Chinese

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SUMMARY **Five cases of Gilles de la Tourette's syndrome occurring in the Chinese are described. The onset, course, symptomatology and family psychopathology are similar to those reported in Caucasians. On the parameters of APGAR score, psychological testing, EEG and psychiatric examination all had at least one abnormality. The response to haloperidol was uniformly good. Follow-up results showed that all were able to lead a fairly normal life after treatment. Though the cases are few in number, the findings seem to support the hypothesis that the syndrome has an organic substratum subject to psychological influences.**

Introduction

Since Gilles de la Tourette first described the syndrome in 1885 there has been a total of about 500 cases reported in the English literature. Of these 500 only 10 were in Asians. Singer (1963) reported the first case in a Chinese boy. Prabhakaran (1970) reported a case in an Indian boy, and Singer (1970) described two further cases among the Chinese. Teoh (1974) published six cases; one of his patients was Malay and five were Chinese. The reasons for the scarcity of reports from Asia may be not only the rarity of the syndrome but also lack of awareness of the condition and the lesser pressure to publish.

Although the reports gave adequate clinical descriptions, only Prabhakaran (1970) presented a detailed psychological and neurological examination of his patient. As regards follow-up studies, Singer (1970) alone cited one patient whom he saw again after a lapse of nine years from initial treatment. We here present five Chinese patients, diagnosed as having Gilles de la Tourette's syndrome on the following criteria:

(1) childhood onset (below the age of 16);
(2) multiple motor tics;
(3) unprovoked loud utterances progressing to compulsive uttering of obscenities (coprolalia).

All five patients and their families were assessed independently by both doctors. Since all five were born in maternity hospitals, we were able to obtain their birth records. The WISC or WAIS and the Make a Picture Story (MAPS) projective test were administered by a clinical psychologist. The MAPS test was used rather than the Rorschach because the latter has not been properly standardized among the Chinese. The EEG was done by a technician and independently interpreted by two other technicians; they obtained an inter-rater reliability of 0.85. The patients were followed-up for 2–4 years.

Case Reports

Case 1

C.Y.L., a 25-year-old woman, was first admitted to the University Psychiatric Unit in 1975 following a suicidal attempt by overdose when her boy friend refused to marry her because of her tics. Mother's pregnancy, delivery and developmental milestones were normal. Her father was an irresponsible person who left home when she was 2 years old. He started another family in the Philippines, but when domestic pressures became too heavy he returned to Hong Kong 20 years later. Her mother was a fierce and dominating woman who

whenever she could not cope with disciplinary problems would threaten to commit suicide or abandon the patient. There was no family history of tics or mental illness.

At the age of 10, soon after a particularly bitter row during which her mother threatened to jump out of the window, she started to show frequent blinking of the eyes, pouting and grimacing. The tics were aggravated by tension. At 15 her facial tics were accompanied by head-jerking and flinging of the arms. A year later she developed expiratory grunts accompanied by occasional mental swearing. During the next 10 years her symptoms waxed and waned in frequency. Over the years she received various treatment: acupuncture, relaxation exercises, psychotherapy, diazepam, amitriptyline, chlorpromazine and anticonvulsants, but none offered any relief.

In spite of her tics she finished secondary education and was trained as an accountant. Though her tics were a constant source of embarrassment, this did not deter her from holding the same job for three years. Her colleagues found her a rather isolated and eccentric person.

At examination, she showed violent flinging movements of her head, eye blinking, grimacing, jerking of both arms accompanied by occasional explosive utterances which at times would sound like an obscenity. Psychiatric examination showed evidence of reactive depression only.

No abnormality was found on neurological examination. EEG showed an unstable background activity. Plain skull X-ray, brain scan and CSF were normal. On the WAIS she had a verbal score of 110 and a performance score of 103. Results of the MAPS showed narcissistic, immature tendencies with histrionic acting out of conflicts and an inability to form meaningful interpersonal relationships. She was also highly anxious about intellectual achievement.

An initial daily dosage of 3 mg haloperidol was given, later increased to 9 mg daily. Within a week her tics were down to a minimum.

In the two years following treatment she had three relapses, but the tics were not as severe as before. She had in the meantime married, and each of the relapses was precipitated by difficulties with her mother-in-law. At follow-up in July 1977 she was on 1.5 mgm of haloperidol daily; occasional eye blinking and grimacing could still be noted, otherwise she was well and happy.

Case 2

F.S.M., a 15-year-old boy, began to have tics at the age of 6. Just before the onset of tics he had had a febrile episode (temperature of 103°F). After the fever subsided the patient's gait was noticed to be unsteady. A week later he developed wrinkling of his nose and jerky movements of the left arm. A month later, the jerky movements spread to the right arm. A paediatrician diagnosed the patient's condition as Sydenham's chorea, and he was treated with penicillin. He did not improve; instead the jerky movements progressed to his legs and trunk. His parents took him to Canton, where he received acupuncture. For a while his symptoms subsided, only to recur eight months later with greater intensity, accompanied by eye blinking head jerking, sniffing sounds and inarticulate utterances.

His mother's pregnancy was full term; delivery was by forceps because of uterine dystocia. APGAR score was 6 (a physical rating of the newborn's audio-respiratory and muscular condition: maximum healthy score 10, asphyxiated less than 6). Developmental milestones were within normal limits. Family history was negative for tics and mental illness. Being the youngest and the only son, his parents tended to spoil him.

He was first seen at our unit at the age of 11 in 1973. He appeared shy and reticent. Tics included jerking of both arms, wrinkling of the nose, twisting of the head and throat-clearing sounds with occasional loud utterances of the word 'shui' which is the Chinese word for 'no good'.

EEG showed diffused slight abnormality of a non-specific nature. Skull X-ray, brain scan, CSF examination and serum ceruloplasmin were all within normal limits. He obtained a verbal score of 86 and a performance score of 94 with the WISC. On the MAPS he showed evidence of negative self-image, an inability to form close

interpersonal relationships and a strong drive for intellectual achievement. Neurological and psychiatric assessment did not add any positive finding.

He was started on a 6 mg daily dose of haloperidol, and after two days his symptoms subsided. At out-patient evaluation in July 1977 he still manifested occasional eye blinking and arm jerking. At present he is being maintained on haloperidol 1.5 mg twice daily. He has continued to attend school and has managed to be promoted each year.

Case 3

C.K.F., a 15-year-old boy was first seen at our clinic in 1974 at the age of 12. His symptoms started in 1972 at the age of 10, with no apparent precipitating cause. Initially the tics affected his mouth, then spreading to the eyes, neck and shoulders within a period of six months. His tics seemed to diminish during holidays and were exacerbated by school attendance. He was usually at his worst during examinations. After exhausting all the local treatment his parents took him to China, where he was treated with acupuncture and penicillin. His symptoms subsided for two months but recurred soon after he returned to Hong Kong. In the six months before he was referred to us he was noticed to be emitting croaking sounds together with the word 'tiu' (fuck).

He was the second of four children. His father was a supportive and kind person, but mother was negligent, spending most of her time in gambling. This became a constant source of parental quarrelling. He was a full-term breech delivery. APGAR score was 8. Developmental milestones were normal. A maternal uncle suffered from schizophrenia. At the age of 2, soon after being weaned, the patient was placed in a nursery because his mother could not cope with the care of two young children. He was frightened and miserable in the nursery, and so was taken home after a month. For two years after that experience, he remained sickly and clinging.

When seen initially, the patient's tics consisted of: puckering of his lips, chewing movements, shrugging of the shoulders, jerky movements of both arms, croaking sounds punctuated by the word 'tiu'.

No abnormality was found on neurological examination. Psychiatric examination showed evidence of depression. EEG, CSF examination, serum ceruloplasmin, serum copper, liver biopsy and brain scan were all within normal limits. Results on the WISC revealed a verbal score of 86 and a performance score of 104. On the MAPS he showed an inability to handle hostility and aggression. He seemed to perceive his environment as being threatening and fearful, and this prevented him from having any satisfactory interpersonal relationship.

The dosage of haloperidol was gradually increased to 4.5 mg thrice daily, and on discharge 19 days after admission the patient's tics were barely noticeable. When last seen in July 1977, he still had occasional jerking of shoulders, but had not needed medication for six months. He had been referred to a vocational school, where he was reported to be doing well.

Case 4

L.S.C., is a 12-year-old girl and the younger of two children. Her father was a seaman and only stayed at home for one month each year. Even when at home he remained a distant and remote figure. Her mother was a strict and domineering person. Family history is negative for tics and mental illness. She was delivered at term by caesarian section because her mother had a contracted pelvis. Her APGAR score was 5. Neonatal period and developmental milestones were unremarkable. Her mother described her as a rather restless child with poor concentration and short attention span.

At the age of 7 it was noticed that the frequently blinked her eyes, and closed and opened her mouth. Two to three months later she developed coprolalia, which could be controlled by reading aloud. Soon afterwards she began to have jerking of both lower limbs and trunk. She was prescribed various minor and major tranquillizers by several paediatricians, but her symptoms persisted. About a year after onset she began to spit repeatedly and to smear saliva on her face; concomitantly she also developed a tendency to lick everything. During meal times she would smear rice on her face.

When first admitted to our clinic in 1974 she presented with head twisting, protrusion of the tongue, facial grimacing, jerking movements of all four limbs and frequent coprolalia.

She obtained a verbal IQ of 97 and a performance IQ of 117 on the WISC. Projective test showed a socially inhibited girl, who engaged in poor peer and adult interactions. She was also shown to be strongly preoccupied with intellectual pursuits. EEG showed diffused slow activity of a non-specific nature. Brain scan and CSF examinations were within normal limits. Neurological examination did not give any positive findings. On psychiatric examination she was noticed to be manipulative and verbally aggressive.

She was prescribed haloperidol 2 mg daily and when dosage was increased to 9 mg daily there was almost complete remission of her symptoms. She was discharged after 21 days in hospital.

In spite of regular medication she has had three readmissions since discharge; each relapse was precipitated by rows with her mother or teachers. Further increase of haloperidol dosage led to dyskinesia and drowsiness. For this reason mass practice was attempted; to this she responded quite well and was discharged, each time with some residual jerking of the legs and shoulders. At present she is on a total daily dose of 6 mg haloperidol. Though her tics have been reduced to a minimum, her behavioural problems have remained. In spite of joint family therapy and transfer to a special school for emotionally disturbed children, she has persisted as mischievous, aggressive, manipulative and socially isolated.

Case 5

Y.O.T., a 13-year-old boy, started to produce coughing and barking sounds after recovery from an upper respiratory infection at the age of 12. Within one to two months, facial grimacing, flinging of his upper limbs, foot stamping and coprolalia appeared. He responded to chlordiazepoxide, but three months later his symptoms became florid again, this time with echolalia. Both tics and coprolalia were aggravated by school work and examinations.

He was the youngest of three sons. Father was an unassertive man who left all the responsibility of child rearing to mother. Mother was a very perfectionistic, over-protective and religious woman and had symptoms of anxiety neurosis. She was not unduly upset about the patient's tics, but when he started to show coprolalia she suffered a breakdown. There is no known family history of tics or mental illness. The patient's gestation, neonatal period and developmental milestones were normal. He had the cord around his neck at birth and suffered from asphyxia neonatorum. APGAR score was 5.

This boy was seen at our clinic at the age of 11. Investigations, including EEG, lumbar puncture, brain scan, pneumencephalogram, all gave normal results. His verbal IQ was 100 and performance IQ was 110. His performance on the projective test implied conflict between his oral dependency needs and an increasing need to assert himself. He demonstrated the capability of forming meaningful interpersonal relationships and was highly motivated towards intellectual achievement. Neurological examination did not elicit any abnormality. Psychiatric assessment found him to be a timid and inhibited boy with no evidence of any formal psychiatric disorder.

He improved gradually as haloperidol was increased to a daily dose of 42 mg and was discharged after eight weeks with some residual jerking of the right arm. At present he is being maintained on 4.5 mg of haloperidol. Any attempt to reduce the dosage further results in the onset of barking sounds and coprolalia. Coprolalia has also been noticed to be intensified when he is scolded by his mother. At follow-up in July 1977 he was reported to have passed his secondary school entrance examination and had been participating actively in competitive sports.

Discussion

The phenomenological characteristics of all five cases support the contention of Abuzzahab and Anderson (1976) that the syndrome is uniform not only between samples but also across cultures. Although Eastern and Western cultures differ in many facets, the repertory of human behaviour remains remarkably similar; in three of our cases one parent was shown to be

rigid, controlling, punitive and domineering—compare MacDonald (1963) and Lucas *et al* (1967, 1970). Dunlop (1960), Challas *et al* (1957), Moldofsky *et al* (1974) and a host of others have also observed that tics are used as symbolic expressions of hostile impulses directed at authority figures. In line with the findings of Corbett (1971), our series also found a greater incidence of disturbed family dynamics.

Pasamanick and Kawi (1956) found a high incidence of prenatal and perinatal complications in tiqueurs and tentatively postulated 'a continuum of reproductive causality' in the aetiology of tics. Sweet *et al* (1973), however, in 34 cases found only 4 with an abnormal birth history, in line with Corbett (1971). Four of our five patients had an abnormal birth history, but only two were found to have a low APGAR score, one with an abnormal EEG.

Woodrow (1975) cited physical factors like tonsillectomy and chronic sinusitis as precipitants of the illness. Fernando (1976) found tics following stressful events in 4 of his 6 patients. Abuzzahab and Anderson (1976), too, noted a distinct precipitant in 17 out of 50 cases, 8 purely psychological, 7 after a febrile illness and 2 after surgery. Three of our cases had a definite antecedent; one was preceded by her mother's suicidal attempt and two by a febrile illness.

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