

GILLES DE LA TOURETTE'S SYNDROME

Pages with reference to book, From 248 To 249

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INTRODUCTION

Gilles de la Tourette's syndrome is a rare condition and very few cases have been published from Pakistan¹, India^{2,3} and Sri Lanka⁴. The scarcity of report from Asia maybe not only the rarity of the syndrome but also lack of awareness of the condition and the lesser pressure to publish⁵. Three cases of Gilles de la Tourette's syndrome seen at the Department of Neuropsychiatry, Jinnah Postgraduate Medical Centre, Karachi are being reported here.

CASE 1

F.O., 12 years, female, student of class VII was brought by her parents from Naushera with 8 years history of episodic hyperactivity, facial tics, jerky movements of different parts of the body and peculiar voices in an otherwise normal child with good sleep and appetite. Her early developments were within normal limits. She was the eldest of three sisters staying in a nuclear family set up. There was nothing significant in the past history except occasional episodes of high grade fever. No history of epilepsy or joint pain could be elicited. Nine years back she was diagnosed and treated as chorea. As she did not respond, the family resorted to various treatments including spiritual means. Her IQ was 85 on 'progressive matrices'. EEG could not be done.

CASE 2

S.L., 13 years, male, student of 7th class was brought from Paposh Nagar, Karachi with three years history of generalised jerky movements, unprovoked vocal utterances, increased thirst with a tendency to take ice cool drinks and shakes. During this period he had been irregular at school. Parents' earliest impression was that of imitations of action seen on a T.V. serials. He was the only son, third in order of birth with three sisters living in a nuclear family system. His early developments were within normal limits. His father a radio cum T.V. mechanic had been busy with his shop from early morning till late evening since long: Neurologically nothing abnormal; high voltage delta activity on EEG was the only finding. Psychological assessment on 'Bender Gestalt' and 'Progressive Matrices' showed no sign of mental subnormality or brain damage.

CASE 3

A.A., 12 years, male from Chakiwara was referred by a general practitioner with 3 years history of abnormal jerky movements, excessive desire for very cold water to relieve thirst, abnormal eruption of teeth and unprovoked vocal utterances. Initially the parents related his problem to imitation of some T.V. acting to attract attention. Manual efforts to control them failed badly. Before visiting us, he had received sodium valproate beside anxiolytics. He was the youngest of four sibs including two who were born dead after full term pregnancies. Two brothers aged 19 and 16 supported the family as unskilled labour because their father had been incapacitated due to mal-united fracture of right femur bone and pulmonary tuberculosis. Polydactyly was seen in mother and the maternal grandmother, two of the deceased sibs and the eldest brother. His early developments were within normal limits without schooling because of financial limitation. Psychological assessment on 'Progressive Matrices', 'Bender Gestalt' and 'Draw A Man' tests excluded any evidence of subnormality of intelligence or brain damage. There was no neurological deficit. Repeat EEG revealed no abnormality.

DISCUSSION

This peculiar syndrome was described by Itrad in 1825 and later established by George Gilles delaTourette when he was working with Charcot⁶. The incidence of reported cases in childrens' psychiatric clinics varies from one case in 1,000 patients to one in 12,500 patients. The tics are coordinated (less than one minute in duration), rhythmical, purposeless, rapid and may vary from one to thousand tics per hour. The tics are usually multiple involving more than one muscle group. Dystonic components are occasional concomitants of this syndrome⁷. Tics cease with sleep, decrease with attention span, sexual activity and voluntary control of the urge to stop them⁸. They are typically worse during times of stress⁹. A simple single facial tic usually occurs at first; but other parts of the body or vacalization may be involved⁷. This severe disorder of childhood onset found in all social classes is characterized by multiform motor, phonic, behavioural and psychological symptoms-mostly phobias and obsessive compulsive symptoms^{8,10}. Imitation of movements observed or viewed on television has also been reported by Friel¹¹. Yamane et al has endorsed Sanders¹² observation of imitation as conscious mimicry thereby attributing imitation as a symptom of the disease. Infact substantially higher frequencies of obsessive compulsive symptoms may be found in Tourette's patients than in general population¹³. In two of our three cases there is a history of increased thirst and a tendency to take ice cool drinks. This has not been reported in any of the reports available to us in the literature. Does this syndrome have any link with the hypothalamus? Yamane et al reported a case accompanied with alcoholism. Their patient described alcohol somewhat effective on his tics and so he drank more. The syndrome shares features with other psychiatric disorders including the attention deficit disorders, learning disabilities, obsessive compulsive disorder. Clonidine has been described as effective in the amelioration of compulsive symptoms, aggression and behaviour difficulties¹⁰. Other treatments that have been tried with little success are physostigmine¹⁴ and self administered aversion therapy with naloxone⁹. Discovery of haloperidol in 1960's generated new investigatory and clinical interest in the disorder and focussed research on the role of dopaminergic mechanism. Unfortunately blood level of haloperidol is unrelated to therapeutic response. However the minimum effective blood level in one study was found to be about 2.0mg/ml. Other dopamine agonists such as pimozide and penfluridol are alternative drugs that should be tried in patients with inadequate response to haloperidol or who develops adverse side effects¹⁵. Shapiro et al¹⁵ note that occasionally symptoms exacerbate either spontaneously or in reaction to stress. This effect may indicate not only tolerance to haloperidol but the spontaneous increase in symptoms characteristic of the disease. The medication dosage may have to be increased.

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