PARANOID PSYCHOSIS IN A CASE OF KLINEFELTER'S SYNDROME

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SYNOPSIS

A 27-year-old Chinese male with paranoid psychosis and Klinefelter's syndrome is described. The purpose of the article is to draw attention to the psychiatric aspect in this syndrome of which there is little awareness. The cause of his paranoid psychosis is thought to be due to Schizophrenia. Chlorpromazine controlled the disturbed behavior but did not have any effect on the paranoid delusions.

Klinefelter's syndrome is a sex chromosome anomaly occuring in phenotypic males. It occurs with a frequency of 1·7 per thousand or 0·17% among newborn males (Forssman, 1970). First described by Klinefelter *et al* in 1942, it was later shown that the cells in these individuals had one or more chromatin bodies. In 1959, Jacobs and Strong showed that the karyotype was 47XXY following which other karyotypes have been described (e.g. 48XXXY, 49XXXXY, 48XXYY, mosaic 46XY/47XXY and phenotypic males with 46XX).

It is known that the frequency of positive sex chromatin among the mentally retarded or the mentally ill is higher than that for the normal population, being 0·89% and 0·54% respectively (Forssman, 1970). Indeed, studies indicate that males with this syndrome have a higher risk of developing mental illness or sociopathic behavior later on in life (Nielsen, 1969 and Forssman, 1970).

A report of one case of Klinefelter's syndrome associated with a psychiatric disorder does not imply a causal relationship but in the light of studies by other workers, the occurence of mental abnormality might not be entirely fortuitous. Locally, Wong and Chua (1969) reported 10 cases of 47XXY, of which 3 were mentally retarded while none had other psychiatric disease. Kutty *et al* (1968) described a case of Klinefelter's syndrome in a 30-year-old Chinese male who complained of poor memory and impotence but did not suffer from other psychiatric disorder. We report in this paper, a patient with a paranoid psychosis and Klinefelter's syndrome.

CASE REPORT

Our patient, L.T.H. a 27-year-old Chinese male presented with a history of talking to himself and paranoid delusion for one month.

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The patient is the eldest of 3 siblings with a sister aged 23 and a brother aged 22. His birth history and developmental milestones were normal. There was no family history of mental illness. He was a backward student and despite repeated failures was given automatic promotion to Primary 5. He left school at the age of 14 and started work as a road sweeper at 18. He performed his work satisfactorily for the next 9 years.

He is not married but claimed to have normal libido. He did not have any previous sex relations. He has a schizoid personality which accounted for his inability to make friends at school and withdrawal from extracurricular activities. He became more self-conscious and shy from the age of 18 when he grew rapidly in height. In June 1969, he had an operation for a left mixed parotid tumour. Although he was depressed for a short while, follow-up did not reveal significant psychiatric features.

In August 1970, he accused his neighbours of using abusive language at him. He also alleged that they wanted to harm him. He had auditory hallucinations and was admitted to Woodbridge Hospital for observation. He was treated with Diazepam and 3 weeks later he left the hospital being less disturbed but still paranoid. Four months later he was re-admitted with a history of being agitated and was more paranoid for 2 weeks. He had auditory and olfactory hallucinations and complained of a foul odour emanating from his body. He had insomnia and was paranoid at the male nurses whom he accused of putting poison as well as faeces in his food. He also had grandiose delusions of being made a minister by Chairman Mao. However he did not show formal thought disorder or incongruity of affect. He calmed down with Chlorpromazine though this did not affect his paranoid delusion.

Psychologically, his Intelligence Quotient (I.Q.) was 71 (Performance Weschler Adult Intelligence Scale or W.A.I.S.). As the patient is unable to speak English, it was felt that a verbal W.A.I.S. would not

be a reliable assessment. He had poor insight to problem-solving and showed female identification on projective testing (Thematic Apperception Test).

Clinical examination showed that he had the typical features of Klinefelter's syndrome: tall stature (72½ inches), asthenic build, pigmented naevi on the chest, scanty axillary and public hair, relatively normal penis and small testes. His height is above the 97th percentile for his age, sex and ethnic group (Chang *et al*, 1965). There was no gynaecomastia. His blood pressure was 110/80 mm. of Hg. The fundi were normal and the other systems were normal.

Investigations showed that his chromatin sex (buccal smear) was positive. He chromosomal karyotype was 47XXY (Fig. 2). His 24-hour urinary 17-ketosteroids content was 3·6 mg. (normal range 9-24 mg/day). X-rays of the skull and chest were normal.

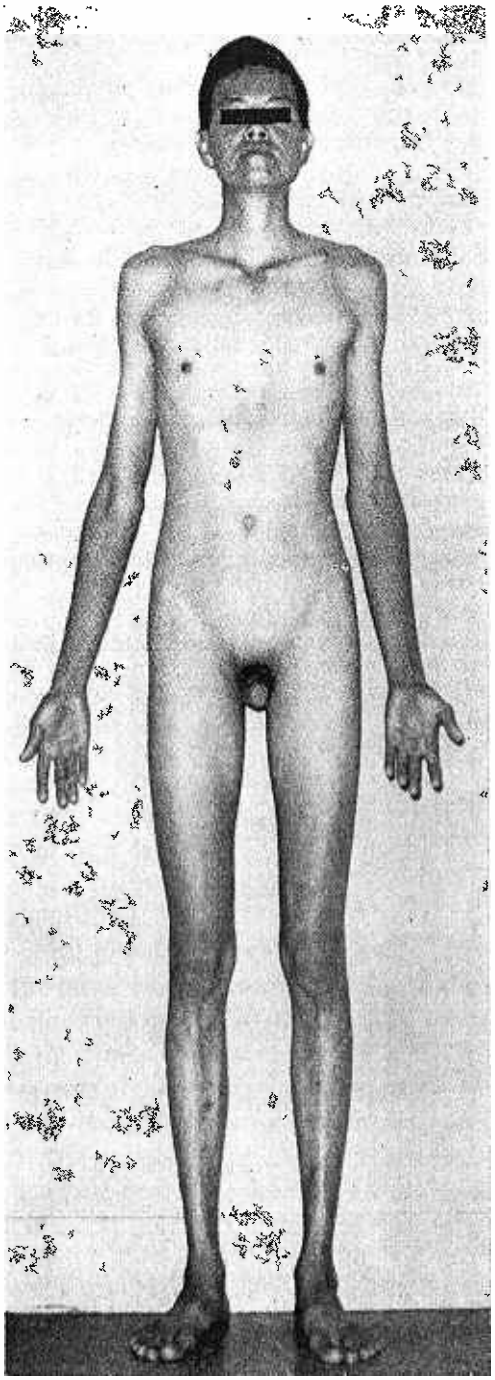

Fig. 1. Physical appearance of the patient.

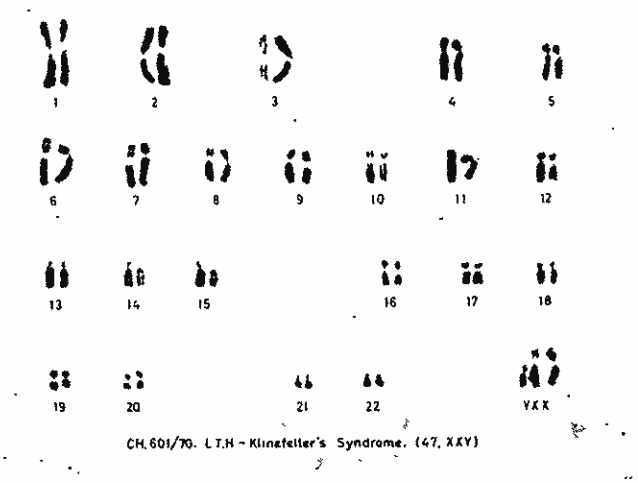

Fig. 2. Chromosomal karyotype (47XXY) of patient.

DISCUSSION

The subject of sex chromosome anomalies and behavior is an exciting one. Court Brown (1962) first drew attention to the association between sex chromosome constitution and delinquency. Fourteen (30%) of his 46 cases of Klinefelter's syndrome had a history of antisocial behavior such as larceny, arson and indecent exposure. His finding was supported by Nielsen (1964b) who found 4 delinquents out of 10 cases of Klinefelter's syndrome. Nielsen (1964a) also found that the prevalence of Klinefelter's syndrome in a mental hospital male population was 5 to 6 times higher than the general population. Becker *et al* (1966) in a study of 50 cases of Klinefelter's syndrome at the Mayo Clinic found about 30% with a significant degree of mental illness and another 30% with a minor psychiatric disorder.

There is a spectrum of psychiatric disorders in Klinefelter's syndrome (Swanson and Stipes, 1969). Some of the abnormalities are personality disorders, neuroses and psychoses which are usually in the form of paranoid states and schizophrenic reactions. In some cases, personality disorders manifest as alcoholism or anti-social behavior. Nielsen (1970) reported a higher frequency of criminality and sexual crimes in this syndrome.

Our patient is mentally retarded with a schizoid personality. There is no history of alcoholism or criminality. It would appear that his psychotic behavior was not due to any specific stress factor. There was no evidence of tension among the family members. It is possible that awareness of his small testes could have affected him adversely with regard to his self-concept but he was ignorant of it before his admission to hospital. It was felt that Schizoph-

renia was a likely cause of his psychosis and this could explain some of the bizarre manifestations and the lack of a precipitating factor. The typical features of schizophrenia such as formal thought disorder and incongruity of affect were however absent.

There are two views on the mental aspects of Klinefelter's syndrome. Kvale and Fishman (1965) emphasize on the disturbances of the body image and self-concept initiated by the physical changes associated with this syndrome during adolescence. Thus it is believed that these individuals do not establish clear masculine identities. It is interesting to note that our patient had a female identification. Forssman (1970) holds the view that there is a cerebral dysfunction caused by the abnormal sex chromosome constitution. This could explain the lower incidence of mental disorders in hypogonadal males due to other causes and also the occurence of similar mental disorders in those with 47XXX or 47XYY karyotypes.

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