may simulate anorexia (Kornmehl *et al*, 1988). In both these instances, there is a danger of missing the obstruction, with resultant delays in treatment. Thirdly, chronic SMAS may complicate anorexia, in which case either syndrome may be diagnosed but probably not both. The fourth possibility is for chronic SMAS to precipitate anorexia.

The delay in diagnosing anorexia nervosa in this case was partly due to the attitude of the patient, who was reluctant to provide the information required, and that of the parents, who thought it to be irrelevant, and partly because of the severity of the organic condition. Although some residual symptoms of the same nature as the original ones are expected to persist post-operatively, there are two main differences between the pre- and post-operative states: the first is that all patients are expected to regain normal weight (Ylinen *et al*, 1989), and the second that the vomiting mostly stops altogether and the other symptoms are milder and respond to symptomatic treatment. In this patient the vomiting did stop, and in the immediate post-operative period she complained of very little pain or distension. However, she failed to gain any weight, and her gastric complaints increased and failed to respond to symptomatic treatment, which induced the treating physician to seek further psychiatric help. The resistance then exhibited by the patient to such help, her refusal, against all evidence, to admit that she was underweight, and the fact that her menstrual periods had been irregular for much longer than she at first admitted and, at the time of her presentation, were not far from amenorrhoea, all indicate a diagnosis of anorexia nervosa.

There remains the question of which of the two conditions, SMAS or anorexia nervosa, preceded and helped to precipitate the other. This patient's symptoms made their first appearance when she was about 14 years of age, which is indeed a period of rapid growth. She was presumably anatomically predisposed to develop SMAS should any factor intervene to prevent her weight from increasing to match the increase in height, with a consequent failure to build up an adequate retroperitoneal pad of fat. As there seems to be no other apparent reason for it, it may be surmised that it was the onset of anorexia at that stage that provided that factor.

As the number of reports associating SMAS with anorexia increase, it becomes essential to consider both conditions whenever faced with weight loss and vomiting in an adolescent.

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The Association Between Triple X and Psychosis

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Two cases of psychotic illness in association with the karyotype triple X showed specific diagnostic and management problems as well as obstetric complications, EEG abnormalities, and lack of a family history of psychiatric disorder. Routine karyotyping during the investigation of psychosis is becoming relevant to psychiatric practice as research reports increasingly feature genetic and chromosome anomalies in association with schizophrenic psychoses.

British Journal of Psychiatry (1992), **160**, 554–557

The search for congenital and acquired organic abnormalities in the functional psychoses has a long history. Pharmacological agents and cerebral pathology have regularly been reported in association with schizophrenia-like psychoses. Initially, these were seen as rare syndromes, but this did not decrease their interest to those involved in the search for an aetiopathology of schizophrenia. It has now become clear that cerebral abnormality is quite common in schizophrenia, with up to one-third of cases showing enlarged lateral ventricles (Johnstone *et al*, 1976), and minor physical anomalies are also thought to occur more often among schizophrenics (Gualtieri *et al*, 1982).

The association between mental illness and chromosomal or genetic anomalies, both with and without mental handicap, is also increasingly well recognised. This has led to research to locate possible genetic markers: the report of a family with partial trisomy of chromosome 5 and schizophrenia (Bassett *et al*, 1988) led to an international search for a possible locus for schizophrenia on chromosome 5; St Clair *et al* (1990) have reported a family with a balanced translocation on chromosome 11 leading to an increased risk of major mental illness in that family; and Holland & Gosden (1990) have described a family with a balanced 6:11 translocation associated in three generations with psychotic illness.

The association of psychosis and sex-chromosome anomalies has a longer history. Much effort in the 1970s went into linkage investigations of affective psychosis and X-linked colour blindness, and further linkage studies have not revealed an association (Gershon *et al*, 1979). Crow (1990) has postulated that an abnormality of the pseudo-autosomal region of the X and Y chromosomes may lead to the development of psychosis. Crow also feels that the demarcation between schizophrenic and affective inheritance may not be clear cut, and has proposed that the psychoses represent a continuum of variation at a single genetic locus (Crow, 1990). Patients with sex-chromosome anomalies certainly have an increased incidence of psychosis in general, as well as of schizophrenia in particular (Polani, 1977).

We report two patients diagnosed as having schizophrenia and schizoaffective disorder using DSM-III-R criteria (American Psychiatric Association, 1987), who were later found to have the karyotype 47XXX during clinical investigation at the Maudsley and Bethlem hospitals. This illustrates the relevance of routine chromosomal analysis to clinical cases of schizophrenia.

Case reports

Case 1

A is a single 20-year-old woman. She was born by forceps delivery following a long labour, but required no active resuscitation although there had been concerns about foetal distress. Her milestones were normal, except that she did not speak until she was three, and did not use sentences until she was four. She had speech therapy on the advice of a paediatrician, who saw her at three years and told her parents she was brain damaged. A diagnosis of autism was considered but discounted. There was no family history of mental disorder.

She interacted poorly with other children, and had few friends at school. She was in the lowest stream of a comprehensive school, but worked hard and passed seven examinations at the age of 16. Educationally she achieved less than her two sisters and was less sociable. As a teenager her behaviour was like that of a toddler; she began to have screaming rages when she did not get her own way, and was constantly annoying her sisters by demanding attention, pinching, and sometimes hitting until she received it.

At 16 her behaviour became more unusual and socially inappropriate. She complained that when she looked at things their colour changed and she heard voices talking about her and to her; she once appeared naked at a social function, and she kept the family awake at night by banging doors.

At 17 she was admitted to hospital with sleep disturbance, symptoms of bodily change such as complaining that her teeth were rotting, and her throat feeling as if it were shrinking. She was constantly washing her hands and using creams to stop her body 'drying out'. She believed that an evil spirit was trying to kill her and trip her up at night, but it was not felt that this was held with delusional intensity and a diagnosis of a stress disorder was made. On examination she was noted to have self-inflicted scars on her wrists, and her ankles were intermittently swollen. She looked slightly dysmorphic: her eyes were close together, and her pelvic girdle was wide compared with her shoulder girdle. She had a postural scoliosis, with six lumbar vertebrae on X-ray. Electroencephalography (EEG) showed generalised episodes of paroxysmal spike and wave activity. Computerised tomography revealed asymmetrically rounded lateral ventricles. Following a short admission she returned home, where her behaviour remained disturbed and her family were unable to cope with her.

She spent the following two years in hospital, during which time she developed further psychotic features, with thought broadcast, passivity feelings, third-person hallucinations, and loosening of associations; these responded to neuroleptics in high doses, and a possible diagnosis of schizophrenia was considered.

At 18, after two years in hospital, she spent a year living in an adult placement scheme, where she was consistently unable to attend either sheltered employment or day centres on a regular basis. At 19 years she was readmitted to hospital with an incongruous and labile affect, having been found wandering and disorientated. At interview she was mute and withdrawn, offering inappropriate replies to questions when she did reply. Over a few days she revealed that she thought that people could read her mind and that she felt she had to copy other people's gestures when she was spoken to. She reported hearing voices calling her name and telling her she was silly. She behaved childishly, wanting constant supervision in simple tasks.

Physical symptoms, such as incontinence, sensations of ankle swelling, and faintness, had no organic basis. Preoccupations with handwashing were prominent. Intermittent second-person hallucinations remained despite other behavioural symptoms responding to treatment.

A repeat EEG again showed generalised slow activity and spike waves which were felt to be compatible with anoxic brain damage and schizophrenia respectively. She was given a trial of carbamazepine which did not change her mental state or EEG, but she did respond to neuroleptics.

Her mental state fitted the DSM-III-R diagnosis of schizophrenia, and some weeks after this diagnosis was made her chromosomal analysis revealed a karyotype 47XXX. Her parents' karyotypes were normal and there was no other family history of mental illness revealed on interview of family members.

Case 2

B is a divorced woman, who was born four weeks prematurely and whose milestones were normal, except that she did not speak in sentences until five years of age. Throughout childhood she was behaviourally difficult, with temper tantrums and violent outbursts. She was the third child in a family of eight children, three of whom were diagnosed as epileptic, but all attended mainstream schools. She was diagnosed as epileptic at 7, and at 11 had an EEG showing "paroxysmal activity". She then attended a school for children with epilepsy, and from 11 to 14 a school for educationally subnormal (ESN) children. Other siblings, including those with epilepsy, do not have a psychotic illness; this was confirmed by interview with her mother and a sister. From 17 she lived in a hostel for ESN adults and worked for limited periods of a few months as a laundry assistant. She married at 20 and had a child who was taken into care at birth as neither parent was thought able to cope. Her violent behaviour lead to two convictions for actual and grievous bodily harm.

She was first admitted to a psychiatric ward at the age of 20, and then again at 23, since when she has had eight admissions. She has had paranoid delusions, auditory hallucinations, and somatic symptoms prominent in her mental state, which has not responded to neuroleptics on a consistent basis, and has been changeable. Treatment has not always been effective, and her difficult management was thought to be due to a combination of mental handicap and personality disorder.

During recent admissions she has shown thought disorder, mood changes (in a matter of days between mania and depression with nihilistic delusions), agitation, and somatic delusions (electricity passing through her, her body dying, bits of her body missing). Grand mal fits have been thought on occasion to be pseudoseizures, although an EEG at age 38 showed "borderline paroxysmal tendency against a background of relative slowness which may represent medication effects". Her full-scale IQ was assessed at 38 years as being 61, which was the same as when tested in her 20s. Her mental state and subsequently her behaviour have improved with a combination of mood-stabilising medication and neuroleptics.

A diagnosis of a rapidly changing schizoaffective psychosis is thought to be an appropriate diagnosis, although a range of diagnoses have been considered in the past, including personality disorder, schizophrenia, and a psychosis related to epilepsy. However, her epilepsy was generalised rather than being temporal lobe seizures, and fits were never associated with a worsening of her mental state. The characteristics of her mental state, and the pattern of her illness over time, were more in favour of a schizoaffective illness. Chromosomal analysis performed at the age of 43 showed karyotype 47XXX.

Discussion

The association between mental handicap, mental illness, and karyotype 47XXX has been previously described (Kidd *et al*, 1963). Typically, affected XXX women show no gross physical abnormalities and go undetected. The incidence of the karyotype is approximately 1 in 1000 births. IQ can be within the normal range but there is an increased incidence of mental handicap, mosaics being less affected. Even XXX women with a normal IQ underachieve compared with their siblings, and can show deficiencies in certain specific skills such as interpersonal relationships and learning, as do XXY men.

The increased incidence of mental illness in women with triple X was suggested after the discovery that there was an increased prevalence of this chromosomal abnormality in psychiatric in-patients compared with that in the normal population: reported prevalence rates range from 0.28% to 0.38% (Olanders, 1967), compared with rates in the newborn of 0.1%. The prevalence in women with schizophrenia is greater than expected, with rates ranging from 1:277 to 1:105.

Changes on EEG have been noted in both Kleinfelter men and XXX women, but these lack specificity and do not show clinical signs of epilepsy (Kidd *et al*, 1963). EEG abnormalities are also associated with karyotypically normal individuals with schizophrenia.

The case reports illustrate some of the common features seen in XXX such as being slow to speak, poor school performance even with normal IQ (Kidd *et al*, 1963), difficulties in social and interpersonal interactions, abnormal EEGs (not specified as characteristic), and behavioural problems. Commonly, psychotic episodes in XXX, if present, have paranoid and affective features (Kidd *et al*, 1963), and may begin in the early 20s after a background of earlier difficulties. While the association between 47XXX and schizophrenia is recognised, it is unclear whether the mental state has any typical characteristic. Both women had somatic symptoms which were delusional at times, and both improved with treatment, although both were considered management problems, with features in their personal history which contrasted with family background and more typical schizophrenic presentations, but these features are subtle, and seem more obvious *post hoc*. However, in both cases the diagnosis of psychotic illness was made several years after the initial presentation, and the improvement in social behaviour when treated raises the issue of how much of each woman's behaviour was attributable to XXX, to mental handicap, or to psychosis.

It is interesting to note that both had obstetric complications, and before they were found to have a chromosome abnormality this was considered the cause of their early difficulties, and of the later development of psychosis. Eagles *et al* (1990) have discussed the association between specific obstetric complications and schizophrenia, and have pointed out evidence suggesting that abnormality in the foetus may actually lead to obstetric complications. We could find no literature on the relationship between XXX and obstetric complications, but it is tempting to speculate that the obstetric difficulties experienced by our patients were secondary to their trisomy.

The importance of increased awareness of the association of XXX and schizophrenia for aetiological research is undoubted. Whether knowledge of the XXX karyotype affects clinical management is less clear, since such patients respond to neuroleptics and behavioural management like other cases of schizophrenia. It may be that genetic counselling regarding the heritability of 47XXX is the most useful result for the women concerned, although the knowledge of a chromosomal disorder may also help families come to terms with their relative's illness and difficult behaviour. XXX is inherited in a sporadic manner, as it is caused by non-dysfunction of the X chromosome during meiosis and so does not give rise to a family history in most cases.

These two patients had recognisable abnormalities in their mental states, but presented as difficult diagnostic problems. Both have a history of obstetric complications and EEG abnormalities, and no family history of mental disorder. The relationship of these features to each other, to the XXX, and to the development of psychosis, is unclear and represents a challenge to schizophrenia research. The consequences of having a mental illness that was difficult to diagnose has had implications for both women: at various times both had behaviour which was not thought to be due to psychosis and so were not treated, to the detriment of their mental states, which was explained by personality and low IQ. Whether the expression of their psychosis was modified by features of the XXX syndrome is debatable, and in presenting these two cases we are not trying to advance a causal explanation but merely to point out that events leading to psychosis are complex and that chromosomal analysis can be helpful.

Acknowledgements

The authors wish to thank the patients and their families for their time, and Dr P. Nott for referring us the first case.

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