

of γ G, γ A, γ M, and γ D, but the interstitial tissue was positive for γ G.

COMMENT

At first the diagnosis was thought to be gastric carcinoma with clear-cut dermatomyositis, but operative histology and later post-mortem examination revealed undoubted plasmacytoma, with involvement of the bone marrow. This diagnosis had not been suggested by the clinical findings or by the investigations. There had never been a raised E.S.R. or proteinuria, and serum proteins were normal; radiographs of the spine failed to show any deposits.

Extrasosseous lesions in plasma cell myeloma are uncommon (Edwards and Zawadski, 1967) and skin lesions are distinctly rare (Geschickter and Copeland, 1928; Hayes, Bennett, and Heck, 1952). Of primary extramedullary tumours, fewer than 10% are in the gastrointestinal tract (Hampton and Gandy, 1957; Sharma and Shrivastav, 1961); in such cases involvement of the bone marrow is most unusual.

The concentration of L paraprotein in the tumour was low, and the amount in the urine was small for the extent of the tumour. The excess of polyclonal γ G-globulin found in the tumour (though not in the cytoplasm of the contained plasma cells, which had Bence Jones only) suggests an immune reaction. Reaction to an antigen common to the tumour, the skin,

and muscle could be responsible for the clinical picture of dermatomyositis.

It is suggested that when patients with dermatomyositis fail to improve when given large doses of steroids a careful search should be made for an underlying neoplasm, and that this should include studies for M-band and Bence Jones protein.

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XYX Constitution in Prepubertal Child

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A considerable body of information now exists with respect to the physical and psychological concomitants of the XYX chromosome constitution in adult males. We believe this report is the first to be made of the findings in a prepubertal boy of normal intelligence with this karyotype.

CASE REPORT

The patient, a boy aged 8 years 7 months, was 11 days postmature and weighed 7 lb. 6½ oz. (3,365 g.) at birth. He was the firstborn wanted child of a mother aged 23 and father aged 26. Pregnancy, birth, milestones, and early development were unremarkable. He walked unaided and was talking at 18 months, and was clean and dry by two years.

He was referred to one of us (J. C.) when 4½ years old because he was unmanageable at home, destructive, mischievous, and defiant. He would smash his toys, rip the curtains, set fire to the room in his mother's absence, and kick the cat and his 8-month-old brother. He was over-adventurous and without fear; he would climb high ladders in buildings, climb out on to window sills several storeys high, and walk into the sea without regard to depth.

He had sudden periods of overactivity at irregular intervals lasting a few hours to a few days. At those times his face would be flushed and he would pursue his particular activity with grim intent. Between episodes he would play happily and constructively. His mother described two personalities—one considerate and happy, the other disgruntled and unstable.

He started school at 5 years, and his behaviour became intolerable at times. He had a particular interest in sharp-pointed objects, and had been suspended from school several times because of dangerous and aggressive use of sharp instruments. He rammed a screwdriver into a little girl's abdomen. He stole a pair of compasses from his teacher and fired these from an elastic band. He pushed a piece of chestnut piling into another boy's face, narrowly missing his eye. He took one of his father's darts and punctured bicycle

tires with it. School reports indicated periods of stability, co-operation, and politeness.

He began wandering at 2 years. Since the age of 8 years he has become progressively less amenable to normal discipline. He began playing truant and was apprehended by the police on five occasions, having travelled considerable distances by train and ferry, and once was found on the streets at 5 a.m. with his small brother. The juvenile liaison officer of the police felt that his supervision and advice meant little to the boy.

He has always been in excellent physical health. There is no family history of congenital defects, mental subnormality or psychiatric disorders. His brother is normal, and his parents are a concerned, loving, and intelligent couple.

On physical examination he was tall, handsome, and athletically proportioned—height 57 in. (145 cm.), pubic-sole measurement 27½ in. (70 cm.), weight 88 lb. (39.9 kg.). No clinical abnormalities were found. Apart from his size, his appearance was normal.

At 5 years an E.E.G. at the Maudsley Hospital by Dr. D. A. Pond showed more slow activity than usual at this age, and one or two "suspicious spike-like waveforms" were seen. The record was regarded at the most as only non-specifically and mildly abnormal.

His I.Q. was 84 on the Revised Stanford-Binet test at 4½ years. He did better with non-verbal items. At 5 years 2 months tests at the Maudsley on the Revised Stanford-Binet indicated intelligence within the normal range; I.Q. 95.

Cytogenetical Findings.—Cytogenetical investigation was carried out on blood samples of the patient, his parents, and his only brother aged 4 years 9 months. Chromosomal analysis (10 cells) confirmed that the patient had 47 chromosomes, the extra chromosome being a Y. His parents and brother had normal chromosome complements (10 cells were analysed in each case).

DISCUSSION

A history of antisocial behaviour, truancy, and conflict with the police and education authorities from early years has been noted as characteristic in adults with the XYX karyotype (Price and Whatmore, 1967). The patient was examined cytogenetically because of his large size, severe violence, and aggression. His history bears out the view that individuals with this

sex chromosome complement may show aggressive tendencies from a very early age. Price and Whatmore (1967), in examining the criminal records of XYY males, found very little difference between them and XY controls in respect of crime against property. There was, however, a striking difference in the number of convictions for crimes against the person, only four out of nine XYY males having been convicted of this class of crime, compared with 17 out of 18 controls. In the present case the boy's behaviour disorder manifested itself in aggression against both property and persons from an early age.

Abnormalities have been noted in the E.E.G.s of individuals with a variety of sex chromosome abnormalities. Dumermuth (1961) found E.E.G. abnormalities in all but one of 14 boys with chromatin-positive Klinefelter's syndrome, eight of whom were diagnosed prepubertally. Hambert and Son Frey (1964) found E.E.G. abnormalities more often among chromatin-positive males than among controls to a statistically significant degree. Mellbin (1966) found an abnormal E.E.G. in all four sex-chromatin-negative women studied. In the present case borderline abnormalities were found. Such a record is in accordance with the findings in individuals with other abnormal sex chromosome complements, in whose E.E.G. records the abnormalities have tended to be non-specific and borderline. It is difficult to see the causal connexion between anomalies in the sex chromosomes and cerebral dysrhythmia. Hambert (1964) suggested, at least in chromatin-positive men, that sex chromosome abnormality may lead to an abnormality of central nervous system tissue or make it more vulnerable than otherwise. Hambert also suggested that cerebral damage might result from hormonal changes in the presence of sex chromosome anomalies, or that aneuploidy, like other forms of embryopathy, may lead to premature birth, predisposing to brain damage. In the present case, however, the birth was postmature.

Since the first assessment of phenotypical characteristics in a series of individuals with the XYY karyotype (Price, Strong,

Whatmore, and McClement, 1966) it has been noted that these men were abnormally tall. Our patient was above the 97th percentile for height according to the criteria of Tanner and Whitehouse.* His brother, 44 in. (112 cm.) tall at 4 years 9 months, was within the 75th percentile for height. This supports the view that increased height in the XYY syndrome is apparent before puberty.

It is possible that the personality and character disorders not uncommonly associated with sex chromosome anomalies, and the violence and aggression associated with the XYY syndrome in particular, have an organic basis which may be reflected in the E.E.G.

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Acute Renal Failure in Case of Paraquat Poisoning

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Six cases of fatal poisoning with paraquat (dichloride salt of the radical 1,1'-dimethyl-4,4'-dipyridilium), a powerful herbicide, have been reported (Clark, McElligott, and Hurst, 1966; Bullivant, 1966; Almog and Tal, 1967; *Brit. med. J.*, 1967; Campbell, 1968). Respiratory failure was the main cause of death in all of them.

We present another fatal case of paraquat poisoning in which respiratory failure, hypercatabolic renal failure, and gross damage to the kidneys occurred. Death was due to cardiac arrest, probably anoxic.

CASE REPORT

The patient, a healthy farmer aged 44, was admitted to hospital on 31 March 1967. An hour earlier he had taken a mouthful of Gramoxone (paraquat) from an unlabelled bottle thinking it was lemonade, but had spat out most of it. He had no symptoms, but was agitated. There were no oral lesions or abdominal tenderness. The pulse was 88/min. and the blood pressure 190/100 mm. Hg.

After a sodium chloride emetic he vomited blood-stained undigested food. He was given milk with albumin and copious fluids orally. The vomitus was not analysed for paraquat.

Six hours later he was feeling so well that it was difficult to persuade him to remain in hospital. His blood pressure was 150/80, pulse 80, and the chest was clear on clinical examination. Over the next 48 hours his general condition remained good, but his throat gradually became painful and swollen, causing dysphagia.

Three days after the ingestion of paraquat cyanosis was first noted. It deepened over the next 48 hours, becoming increasingly resistant to oxygen administration. Crepitations became audible over both lung bases. A chest x-ray film showed increased vascularity of both lung fields and a little fluid at the left base.

For the first three days his urinary output, though not recorded, was satisfactory. Thereafter it ranged between 600 and 1,600 ml. daily. His blood urea on the fourth day, when first estimated, was 178 mg./100 ml., Hb 14.1 g./100 ml., and W.B.C. 19,400 per cu. mm., with a normal differential count.

He was treated with Crystamycin (benzylpenicillin and streptomycin), cephaloridine, and hydrocortisone 100 mg. daily.

On the sixth day his blood urea had risen to 310 mg./100 ml. and the electrolytes were normal. Because of the rapid increase of the blood urea he was transferred to the renal unit at the Belfast City Hospital. He was fully conscious and co-operative and his only complaint was thirst. Intense cyanosis developed rapidly when the oxygen mask was removed. There were crepitations over both bases. His pulse was 94 and regular, and the heart sounds were normal. The blood pressure was 125/60. No abnormality of the central nervous system was found. His blood urea was 340 mg./100 ml.

Peritoneal dialysis was begun, but he continued to deteriorate and became less co-operative, and diazepam 5 mg. was given but did not control the restlessness. The cyanosis became worse despite oxygen, and cardiac arrest (ventricular asystole) developed