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GENDER IDENTITY AND ROLE IN A PEDIGREE OF ARABS WITH INTERSEX DUE TO 5 ALPHA REDUCTASE-2 DEFICIENCY

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SUMMARY

Between 1986 and 1995, a pedigree of six Arabs with male pseudohermaphroditism due to 5 alpha reductase-2 deficiency have been identified. All, were raised as girls since birth. At the time of diagnosis, three were post-pubertal, one pubertal and two pre-pubertal. The external genitalia of 'pseudo-vaginal perineoscrotal hypospadias' was identical in these subjects. Although these individuals were a homogeneous group in terms of their sex of upbringing, phenotypic appearance, endocrinological profile and socio-cultural background, the development of the gender identity and role was not uniform in these six cases. Their psycho-sexual make-up was closely related to the transaction of their life experiences. These cases provide further insight into the interaction between various factors involved in the development of gender identity and role in male pseudohermaphrodites in an Eastern culture. © 1997 Elsevier Science Ltd.

Keywords—Intersex; Gender identity; Arabs; 5 Alpha reductase-2 deficiency.

INTRODUCTION

Male pseudo-hermaphrodites with 5 alpha reductase-2 deficiency suffer from impaired metabolism of testosterone to dihydrotestosterone, which results in severe ambiguity of the external genitalia. Those who are thought to be girls at birth and raised accordingly, exhibit various degrees of virilization at puberty and beyond (Peterson et al., 1977). In this study, the author reports the experience of this condition in a pedigree of Arabs, focusing on the development of gender identity and role, and on what appeared to be the determining factors in the development of their gender status.

PATIENTS AND METHODS

Six Omani Arabs with intersex are the subjects of this study. All, belong to interrelated families of low socio-cultural background. These families are geographically distributed in various parts of Oman and the UAE. All subjects were born to consanguineous parents. The interrelated marriage amongst cousins is commonly practised in these families. Patients I and II were delivered in a hospital in Saudi Arabia and have been living in Abu Dhabi

(UAE) since their childhood. Patients III, IV and V were delivered in a peripheral hospital in an area of Oman adjacent to the city of Al-Ain in Abu Dhabi Emirate and were still living there. Patient VI was delivered at home in a rural area of Inner Oman and still lives there to date. Like most Eastern communities, the Arabs favour patriarchal control and males have social, cultural and financial advantages over the females. Moreover, the demarcation between unmarried males and females is observed more by UAE nationals and Omanis compared to other Arabs in UAE. Patients with intersex in the Arab community are called 'Khunthi'. These six subjects were unequivocally raised as females. The ambiguity of their genitalia was identical and had the appearance of 'pseudo-vaginal perineoscrotal hypospadias'. The testicles were at different inguino-scrotal levels. Facial, chest and abdominal hair was scanty and gynaecomastia was absent in adults. Patient III had few acne spots. Patients I, II and VI had rectal examination and cystoscopy, which showed the presence of rudimentary prostate. The karyotype studies and most of the appropriate endocrinological assays were carried out locally. Simultaneous assays for serum testosterone and dihydrotestosterone were initially performed (in Metpath and in JSPJ Laboratories, UK), and recently in the laboratory of Professor J. Imperato-McGinley, USA. Because of financial constraint, the test of urinary chromatography for steroid metabolites was performed only for patients I and IV (in Northwick Park Hospital, and in JSPJ Laboratories, UK). All patients underwent pelvic ultrasonography. Only patients I and II agreed to be interviewed by a psychiatrist. The relevant data on pre- and post-pubertal sexual perception and the analytic conclusion of each case, were based on direct interviews of the patients and parents.

LABORATORY RESULTS

The karyotype of all patients was 46-XY, with absent Barr bodies. The Y chromosome appeared short in cases III, IV and V. This, did not produce additional dysmorphic features in these patients. Mullerian derivatives were absent in the group. Serum androstenedione, 17 alpha hydroxy-progesterone and DHEA sulphate were of normal values. All, except the child (patient V), had normal values of serum testosterone, and low dihydrotestosterone, forming high T/DHT ratios. Urinary 5 beta/5 alpha steroid C19 and C21 metabolites were markedly elevated in cases I and IV. The above findings, along with the positive family history, identicalness of features and absence of pre-testosterone biosynthetic enzyme deficiency were consistent with the diagnosis of male pseudohermaphroditism due to 5 alpha reductase-2 deficiency (Imperato-McGinley et al., 1979; Peterson et al., 1977; Thigpen et al., 1992).

CASE DESCRIPTIONS

Patient I; Present Age 27

This subject was a 17-year-old schoolgirl in 1986 when the diagnosis was made. Although primary amenorrhoea was the main concern of the parents, the subject was seeking an operation to change sex after becoming increasingly doubtful over the nature of self-gender identity. The self awareness, sexual arousal and response were of male nature since the age of 13, and the subject had been experiencing morning phallic erection and nocturnal emission, but had not been engaged in any sexual activity. The phallus was small, testicles were scrotal, and the muscular habitus was well developed. Despite the full awareness that

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