HYSTERECTOMY IN YOUNG MALE

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ABSTRACT

Introduction: Disorders of sexual differentiation (DSD) is the term used for neonates without a definite male or female phenotype. Previously, terms like "intersex" and "pseudohermaphroditism" were used for such individuals. However, DSD is the recommended terminology since 2006[1]. According to a study, about 1 in 3000 children is born with ambiguous genitalia[2]. True hermaphrodite or ovo-testicular disorder of sexual differentiation (OVO-DSD) is the rarest variety[2]. The gonads in these individuals are asymmetric with bilateral ovarian and testicular tissue (in varying ratios), which may be separate or combined as ovo-testes. Karyotype varies from 46XX to 46XY to 46 XY/46 XX mosaicism.

Keywords*:* Disorders, ovaries, differentiation, testes, hermaphroditism.

CASE REPORT

A 25-year-old healthy male presented for management of primary infertility. He had been married for 5 years. He was living with his spouse and had never practiced contraception. His wife had been evaluated by gynaecologist and declared normal. He had no significant illness in past. He was a graduate and did his own business, being socially and financially well settled.

Physically he was a young, well built male with well developed secondary sexual characteristics. Systemic examination was normal. However, genital examination revealed empty scrotal sacs bilaterally. An ultra sonography of abdomen, pelvis and scrotum showed absence of testis at all possible sites. MRI pelvis revealed well developed phallus and prostate as well as a well-developed uterus with broad ligaments and two gonads at its distal end which were suspected to be ovo-testes. Serum testosterone level was 1.03 ng/ml. On disclosure of the findings, the patient opted for hysterectomy. Gross examination of the specimen revealed well developed uterus and cervix with unremarkable endometrium and myometrium. Male genital organ including seminal vesicle and unremarkable vas deference were also present. Biopsy of the gonads confirmed them to be ovo-testes with predominance of testicular tissue. Karyotyping was refused by the patient.

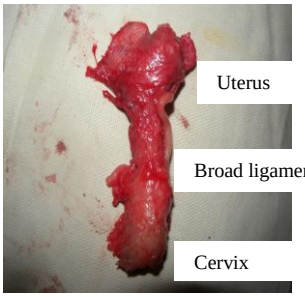

Figure-1: Uterus and Cervix.

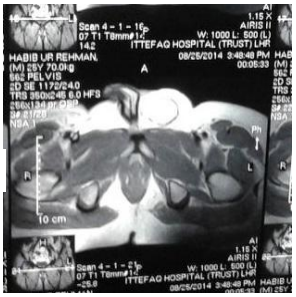

Figure-2: Well developed phallus.

DISCUSSION

Sex differentiation is a complex process involving many genes. Sex of the embryo is determined genetically at fertilization, the gonadal morphological features being acquired at 7-8 week of development[4]. The initial molecular events of sexual development are identical in both male and female embryos. Therefore, abnormalities in sex differentiation can occur which may lead to true hermaphrodites, pseudohermaphrodites and gonadal dysgenesis etc[5].

Microscopic examination of gonads is essential to determine the accurate sex. In true

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Received: 11 May 2015; revised received: 01 Jul 2015; accepted: 13 Aug 2015

hermaphrodites, gonads are almost always asymmetrical and are classified as[6];

- Lateral: Testis and contralateral ovary (30%)
- Bilateral: Testicular and ovarian tissue identified on both sides, usually as ovo-testes (50%)
- Unilateral: Ovo-testis on one side and testis or ovary on other side (20%).

True hermaphrodites may be brought up as male/female depending upon phallus size. Castration is not recommended as it may deprive the individual of beneficial hormonal influences.

A newborn with ambigous genitalia poses a diagnostic challenge and becomes a social emergency regarding gender assigment at the time of birth. A proper diagnosis of DSD, detailed counselling and support of parents, alongwith an optimal and timely medical/surgical managment may help prevent psychosocial issues in such persons and their families.

CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

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