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Testicular feminization in a male pseudohermaphrodite: A case report

Fadhil Ahmmed Mohialdeen, Fahmi Hussein Kakamad, Mohammed I. M. Gubari

ABSTRACT

Introduction: Hermaphroditism is a condition in which the patient has both male and female sexual characteristics either partially or completely. It is classified into true and pseudohermaphroditism. Patients with male pseudohermaphroditism have male internal genitalia and karyotype (XY). We report a rare case of male pseudohermaphrodite who grew up as a female and insisted to live as female for rest of her life in spite of developing male body build. Case Report: A 26-year-old patient presented with bilateral inguinal swellings since birth. Clinical examination showed bilateral inguinal swellings which were reducible manually with positive cough impulses, hypertrophied clitoris or rudimentary penis, without vaginal orifice. She was grown up as female among her family. Diagnostic workup showed that both hernial sacs contain testicles with the absence of ovaries, fallopian tubes and uterus. Hormonal and enzymatic assay were normal including 5-alpha reductase. During surgical operation; both hernial sacs were repaired, testicles were removed through two inguinal incisions. The histopathological report confirmed mature testes. The postoperative follow up was uneventful. Conclusion: Male pseudohermaphroditism is a rare cause of intersexuality and its etiology could be idiopathic. Patient may insist on being female in spite of development of male body build and presence of male internal genitalia.

Keywords: 5-alpha reductase, Feminization, Intersexuality, Male pseudohermaphroditism

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INTRODUCTION

Hermaphroditism is a condition in which the patient has both male and female sexual characteristics either partially or completely [1, 2]. It is classified into true and pseudohermaphroditism. The former refers to patients who have both unequivocal ovarian tissue and testicular elements regardless of their karyotypes. While pseudohermaphroditism is a condition of developmental sex abnormality in which chromosomal and gonadal gender match, but the external genitalia have characteristics of the opposite gender [1]. Mixed

gonadal dysgenesis (MGD) refers to patients who usually have a differentiated gonad on one side and a streak gonad on the other side [3]. Patients with female pseudohermaphroditism have female internal sexual organ and karyotype (XX) and various degree of external genitalia virilization. External genitalia are musculinized from birth when female fetus is exposed to excess androgenic environment. Congenital adrenal hyperplasia (CAH), mostly 2Group 1-hydroxylase deficiency, is the most common cause [4].

Patients with male pseudohermaphroditism have male internal genitalia and karyotype (XY) and various degree of external genitalia feminization. The most common cause is deficiency of 5cc-reductase-2. The clinical features are related to decreased conversion of testosterone to its active metabolite, dihydrotestosterone. The lack of virilization of external genitalia is so severe in some cases that they are assigned them as female in the newborn period. Various degrees of virilization, however, as the patients grow up, including the development of male habitus, changing of the voice and growth of phallus. The most salient characteristic that accompanies the pubertal changes is a disturbance in the sex related behavior of patients raised as females, as in most cases they convert to a male social gender [5].

We report a rare case of male pseudohermaphrodite who rose as a female and insisted to live as female for rest of her life in spite of developing male body build.

CASE REPORT

A 26-year-old patient presented with bilateral inguinal swellings since birth which were reducible on lying flat. Clinical examination showed bilateral inguinal swellings which were reducible manually with positive cough impulses, hypertrophied clitoris or rudimentary penis, without vaginal orifice (Figure 1) and male like pattern body built and breasts. She was grown up as a female among her family and her friends. She decided to live as a female for the rest of her life regardless of her chromosomal patterns and her body builds. Psychiatric consultation was done, which was going with her wish to become a female. Diagnostic workup showed that both hernial sacs contain testicles with the absence of ovaries, fallopian tubes and uterus. Hormonal and enzymatic assay were normal including 5-alpha reductase, urinary pituitary gonadotrophin, 17 kitosteroid, and estrogenic level. During surgical operation; both hernial sacs were repaired, testicles were removed through two inguinal incisions (Figure 2). The histopathological report confirmed mature testes. The postoperative followup was uneventful and discharged home on the same day's evening and appointment was given to her for future creation of vagina by sigmoid substitution.

DISCUSSION

Pseudohermaphrodites are of two types, either male and female. The determinant factors being the gonads. Accordingly, in male pseudohermaphrodites the gonads are testes, whereas the body build is of female. Conversely, in female pseudohermaphrodites the gonads are ovaries, but male tendencies are seen in the organs of reproduction [6]. In our case, the body build was of female fashion until recently changed to male pattern. Several evidences showed that a person's gender orientation and role produce through the cumulative experiences of long duration of been raised as a female or a male. Regardless of genetics, hormones or gonads, the patient who from early life has been raised as a male or as a female, especially if

Figure 1: Hypertrophied clitoris or rudimentary penis.

Figure 2: Vas Deference.

the external sexual organs have been changed in parallel to this sex, will not ask his own sexuality and behave accordingly [7]. Our patient rose as a female among her family and friends. During diagnosis, her body build and internal sex organ were in favor of been converted to male gender but she refused and psychologist confirmed that her identity suits with female gender. That is why parents and health care professionals should try in early life to decide the sex of the baby. As early as possible, necessary corrective surgeries should be performed. No course of action for changing gender should be undertaken later in life [7].

We did not determine the chromosomal pattern of the patient, partially because this investigation was not present in our locality and partially because it did not change the decision of sex conversion consented by the patient.

It is preferred to determine the gender of an individual according to the anatomical structure of the external sexual organs rather than chromosomal pattern. Trying to create a boy in a person who does not have a well-established male genitalia is not right decision and forces him to a misery [7]. Male patients who have external sexual organs of female appearance invariably convert to female genitalia at puberty that is why operation is not required. It might be indicated to overcome the possible risk of malignancy [8]. Male patient whose external sexual organs at early childhood resemble the male gender may convert to either one at puberty. If it is agreed to raise such a baby as a girl due to the small penis and testicles, operation may be done either to prevent or to treat masculinization. The former course of action often recommended [7].

CONCLUSION

Male pseudohermaphroditism is a rare cause of intersexuality and its etiology could be idiopathic. Patient may insist on being female in spite of development of male body build and presence of male internal genitalia if the patients grow up as a female during childhood. Early intervention and surgical correction is advised.

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Author Contributions

Fadhil Ahmmed Mohialdeen – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Fahmi Hussein Kakamad – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
Mohammed I. M. Gubari – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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