

Case Report

True Hermaphrodite: a case report

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Abstract: True hermaphrodite have gonads of both sex in the same individual. They contain ovary and testis with ambiguity of genital organs. We report a case of true hermaphrodite in a male patient of 26 years having absent of beard and moustaches, medium pitch voice, partial fusion of labia majora and testis, enlarged clitoris with non penile urethra, no vaginal and opening having uterus.

Keywords: hermaphrodite, gonads, testis, uterus

INTRODUCTION

It is the presence of gonads of both sexes in same individual. Hermaphrodite = Hermaphrodites, son of Hermes & Aphrodite, united with nymph of Fountain of Salamis = one person with characteristics of both sexes.

Nomenclature of hermaphrodite: Female pseudo hermaphrodite (Female intersex)--Association of female gonads with male external genitalia. Male pseudo hermaphrodite (Male intersex)--Association of male gonads with female external genitalia. True hermaphrodite-- In true hermaphrodite gonads of both sex are present in the same individual, ovary and testis with ambiguity of genital organs [1].

CASE SUMMARY

A married, apparently male patient, aged 26 years, was seen on 26-6-2010 with complaints of cyclical pain in lower abdomen from one year. There was h/o bilateral mastectomy done 6 year ago. On examination his skin was smooth, beard and moustaches were absent. Voice was medium pitched. Scar of bilateral mastectomy seen. There was partial fusion of labia majora and testis was palpable in right side of labia majora. Clitoris was enlarged (1.5") with non-penile urethra. No vaginal opening. P/R examination –uterus of normal size was palpable. USG –Revealed normal size uterus and left ovary. Neutrophil showed Barr bodies. Biopsy of testis showed spermatogonia.

Laparotomy was done on 29-6-2010---Body of uterus, left ovary and left tube were normal in size. Round ligament was present on both sides. There was no right tube and ovary. Cervix & vagina were absent.

Hysterectomy with removal of left tube & left ovary done. Cut section of uterus showed no uterine cavity. Postoperative period was un-eventful. Patient was discharged on 8th postoperative day with advice for regular follow up.

Histopathology---confirmed uterine, tubal and ovarian structures with maturing follicle.



Fig-1

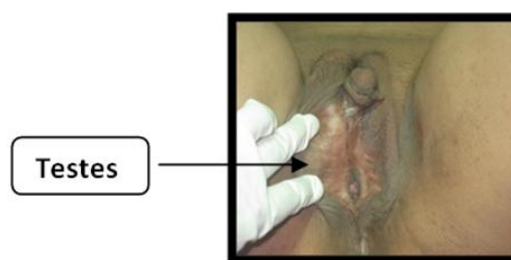


Fig-2

Barr body in
the
neutrophil

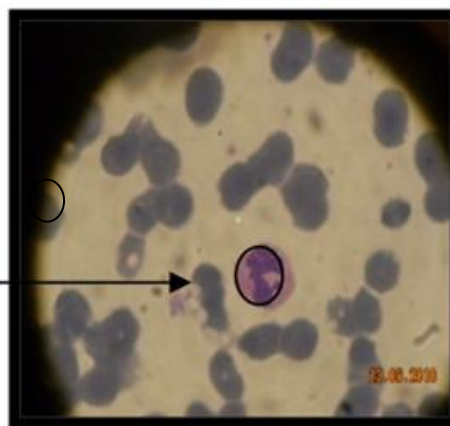


Fig-3

Normal sized uterus

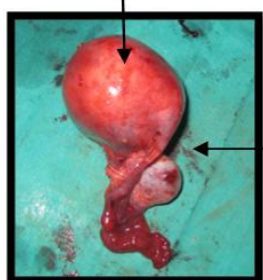


Fig-4

Cut section of uterus



Fig-5

Left tube
and ovary

spermatogonia

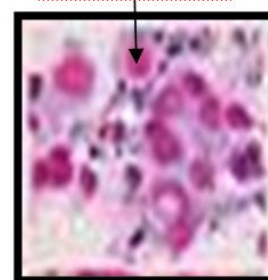


Fig-6

DISCUSSION

In true hermaphrodite gonads of both sexes are present in the same individual - ovary and testis [2]. True hermaphrodite is rare in Europe and common in South Africa among Bantu tribes. Both chromosomal defect and genetic cause are held responsible [3]. Both testicular and ovarian elements are present in different combinations. 1)-Bilateral—ovotestis present on both sides (most common). (2)-Lateral—Testis on one side and ovary on other side. (3)-Unilateral—Testis or ovary on one side and ovotestis on other side [4]. In pseudo-hermaphrodite, gonads are of one sex while external genital organs are of opposite sex. True hermaphrodite is Very rare – only 750 cases are reported in the literature since 1899 [5]. It is probable that fertilization by sperm carrying one X chromosome which contains some male determining material from Y chromosome gives rise to this condition [6]. Common presentation is ambiguity of external genitalia. Internal structures depend on the degree of differentiation of the associated gonads on that side [7].

Investigations

Sex chromatin is usually positive. Karyotype usually 46 XX, 46 XY, rarely XXXY. Confirmation done by gonadal biopsy [8]. Management in this is the change of sex, depends on sex of rearing, psychologic & anatomic sex. Genitalia inconsistent with the sex assignment should be surgically removed or modified [9]. In general, it is possible to change external genitalia from male to female but not from female to male.

Gonads are to be removed and followed by substitution therapy as there is a high chance of malignancy if left behind.

CONCLUSION

- Extremely rare anomaly, most uncommon variant of intersexuality in humans with gonads of both sexes.
- Diagnosis on the basis of history, clinical examination, karyotyping & histopathology.

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