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Rare Case of True Hermaphrodite: A Case Report

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Abstract

True hermaphrodite is one of the rarest variety of Disorders of Sexual Differentiation (DSDs) and represents only 5% of cases of all.

A 12 year-old presented with complains of pain and swelling right side lower abdomen with Right sided Undescended testis. On exploration Mullerian structures was present on right side and on left side testis was normal into left hemiscrotum.

Keywords: True hermaphrodite; Persistent mullerian duct syndrome; Disorders of Sexual Differention.

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Introduction

Disorders of Sex Development (DSD) are congenital condition in which the development of chromosomal, gonadal or anatomic sex is atypical.[1] In about 60% of cases, patients have 46XX karyotype.[2] Rarely, 46XY/46XX mosascism may occur.[3] There have been report of 46XX karyotype.[4]

Case Report

A 12 year-old patient presented in Out Patient Department of Jodhpur medical college and hospital Jodhpur. Brought up as a male child with complains of pain and swelling right sided lower abdomen.

On examination right sided undescended testis and bilateral mild gynacomastia was present. Left testis and penis was normal. (Fig 1)

Ultrasonography scan revealed that right testis in right inguinal canal with tortion. Serum FSH, LH and Estradiol levels were normal according to male patient with its age. However serum testosterone level was slightly lower. Other investigations were within limits.

Figure 1

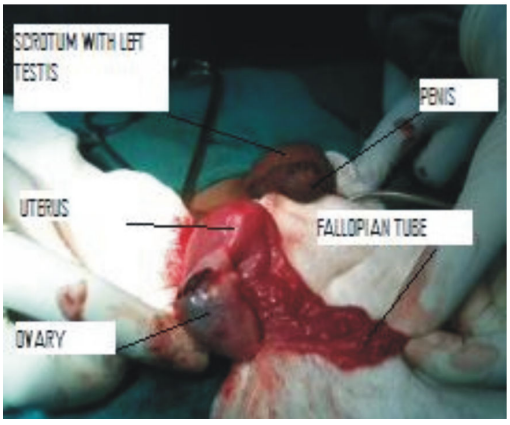


Figure 2

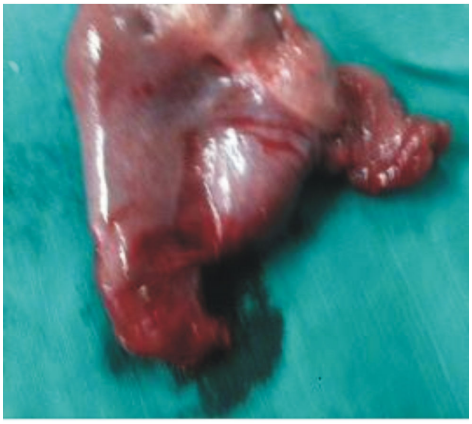
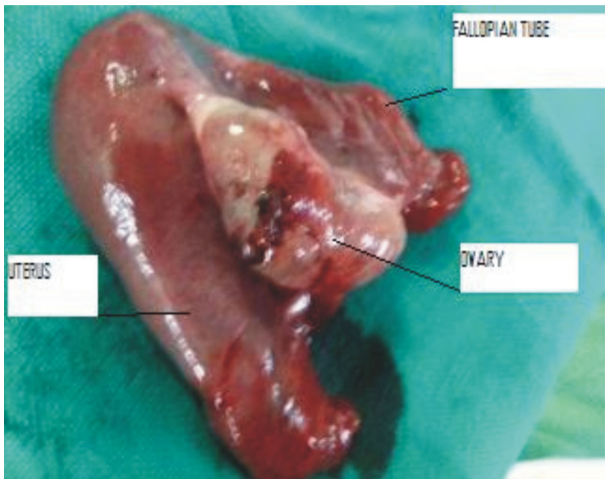


Figure 3



Patient was explored with right inguinal incision and intraoperative finding demonstrated well developed Uterus, Fallopian tube and Ovary instead of right Testis. (Fig 2 - 3) Because patient was by appearance and upbringing a male, these organs were removed. Patient made uneventful recovery.

Biopsy report shows fallopian tube, rudimentary ovary and normal endomyometrium tissue in proliferative phase. No testicular tissue seen in left side.

Karyotypic analysis showed a model number karyotype 46XX with no numerical or structural chromosomal anomalies detected at the banding resolution.

Discussion

Disorder of Sexual Differentiation is the term used for a child born without clear Male or Female phenotype. The term "Hermaphrodite" is derived from Greek mythological God "Hermaphroditos" son of Hermes and Aphrodite, whose body after being merged with nymph Salmakis assumed a more perfect form with both male and female attributes.[5]

Hinman[6] has classified true Hermaphroditis concisely Bilateral: Testis and Ovary (Ovotestis) on each side (50%) Unilateral: Ovotestis on one side and with ovary or testis on the other side (20%) Lateral or alternating: Testis on one side and Ovary on other side (30%). The most common karyotype[7] in true hermaphroditism are 46XX (60%), 46XY (12%) and mosaic (28%) usually 46XX/46XY, 46XY/47XXY or less frequently 45XO/46XY.

The external genitalia of true hermaphrodite are most often ambiguous but can vary from almost normal female to almost male.[8] Internally Mullerian and wolfian derivatives usually coexist.[9] Breast development occur at puberty and virilization may also occur, there may be incomplete development of secondary sex characteristics.[10]

In our case, according to Hinman classification lateral type of true hermaphrodite and karyotype was 46XX. This patient was upbringing as male child so we removed mullerian structured and in follow up period we started testosterone.

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