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LATE PRESENTATION OF A TRUE HERMAPHRODITE

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ABSTRACT One of the rarest variety of disorders of sexual differentiation (DSD) is true hermaphrodite and it represents only 5% of all cases. These cases usually present in early childhood or late adolescence with ambiguous genitalia. We are here reporting a rare case of a true hermaphrodite at 30 years of age with two cellular lines (46XX/46XY), small testis, hypospadias and breast pain. Histology of the gonads revealed right ovotestes.

KEYWORDS : True Hermaphrodite, Ovo-testes, Sexual Differentiation Disorders

INTRODUCTION:

Hermaphroditism is the condition of having both male and female reproductive organs. Conditions that involve discrepancies between external genitalia and internal reproductive organs are described by the term *intersex*. Intersex conditions are sometimes also referred to as disorders of sexual development. In true hermaphroditism, an individual has both ovarian and testicular tissue. The ovarian and testicular tissue may be separate, or the two may be combined in what is called an ovotestis. Affected individuals possesses both the male XY and female XX chromosome pairs. In 46, XX intersex (female pseudo hermaphroditism), individuals have male external genitalia but the chromosomal constitution and reproductive organs of a female. In 46,XY (male pseudo hermaphroditism), individuals have ambiguous or female external genitalia but the chromosomal constitution and reproductive organs of a male.

Intersex surgery was performed to remove the gonads of the opposite sex. The remaining genitalia were then reconstructed to resemble those of the chosen sex. In older individuals the accepted gender may be reinforced by the appropriate surgical procedures and by hormonal therapy.

CASE REPORT:

A 30-year-old male presented to our General Surgery OPD with small penis and hypospadiasis since childhood and enlargement of bilateral breasts for 16 years. Associated with mild pain which was recent in onset for 16 years. He had no history of altered bladder habits, voice change, any comorbidities or any previous surgical history. His secondary sexual features were normal. Upon local examination, he had bilateral gynecomastia, small size penis with single perineal urethral opening on the dorsal surface, bilateral small testis with right side firmer than the left. Routine investigations were under normal limits. USG abdomen showed hypoechoic lesion in left iliac fossa probably left ovary. Scrotal doppler revealed well defined hypo echoic lesions in the right testis with no internal vascularity probably ovary.

Fluorescence in Situ Hybridization revealed presence of 2X chromosomes in all the analyzed cells and he was positive for SRY gene.

Urologist opinion was sought for hypospadiasis correction, which was planned at a later date. Patient was taken up for Bilateral Webster procedure followed by right orchidectomy and laparoscopic left oophorectomy, as dysgenetic testis has high chances of turning malignancy.

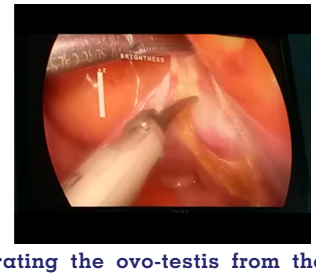


fig 1: separating the ovo-testis from the peritoneum covering

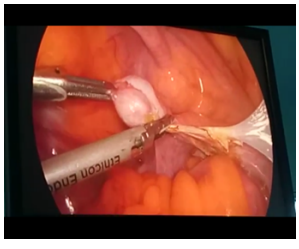


Fig 2: ligating the pedicle fused to the ovo-testis.

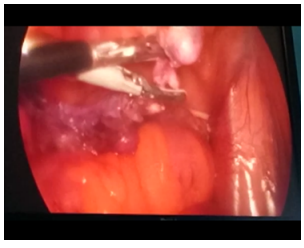
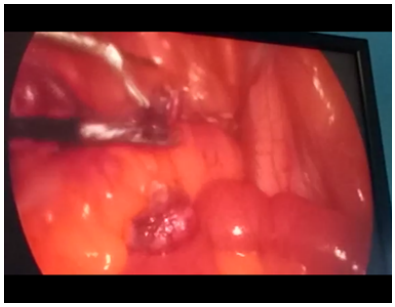


Fig 3 & 4: ovo testis has been cut and removed.



Post-operative period was uneventful.

HPE revealed benign breast tissue and ovo-testicular tissue and benign serous cyst of ovary.

Patient was planned for testosterone replacement after surgery for continuing as male.

DISCUSSION:

The diagnosis of a true hermaphrodite depends on the histologic confirmation of testicular and ovarian tissue in the same individual. 90% of cases present at birth with ambiguous genitalia including micro phallus, hypospadias, urogenital sinus, fusion of penoscrotal labia, or cryptorchidism. Individuals which present with even the slightest of ambiguity in the external genitalia and unilateral or bilateral undescended testicles should be investigated. 70% of cases of true hermaphrodites have 46 XX karyotypes. Only 10% of cases are 46 XY karyotype, while the rest represent complex karyotypes of which the most common is 46 XX / 46 XY. True hermaphrodites can be classified according to the position and the histology of the gonads.

i. Lateral: testis and a contralateral ovary (30% of cases).
ii. Bilateral: both testicular and ovarian tissue, usually represented by an ovotestis that is identified on both sides (50% of cases).
iii. Unilateral: ovotestis on one side and a testis or ovary on the other side (20% of cases).

Imaging modalities that may aid in the diagnosis of true hermaphrodites include ultrasonogram, MRI and genography. Testis and ovaries have textural differences on ultrasound. Management involves medical treatment, correction of the ambiguous genitalia and removal of dysgenic gonads by surgery if patient presents in their childhood or adolescent age.

A soft testicular portion and a firm ovarian portion on palpation should arise a suspicion of ovo testis for diagnosis of a true hermaphroditism

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