Extended Abstract | International Journal of Anesthesiology & Pain Medicine | 2020 | Vol.6 No.3

INTERESTING CASE OF INTERSEX DISORDER

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CASE HISTORY: A 14 year old reared as male presented to the pediatric surgery OPD with complaints of breast tissue enlargement .No history of any drug intake. No history of hypothyroidism, obesity etc

PAST HISTORY - History of bilateral undescended testes at birth. At 4 years underwent Left orchidopexy and Right inguinal exploration.

EXAMINATION:

GENERAL PHYSICAL EXAMINATION: Patient is a young male, moderately built and. nourished , conscious

VITALS – WNL

SYSTEMIC EXAMINATION: CVS / RS- NAD

P/A- Soft, non tender, no palpable mass, bowel sounds heard

BREAST EXAMINATION: Revealed bilateral well formed areola Papilla protruding and forming a secondary mound over the breast

GENITAL EXAMINATION:

- Adequately developed phallus with no signs of hypospadias
- Normal urethral opening and of adequate length
- Testis palpable in the left hemiscrotum .
- Right hemiscrotum is empty
- No other swellings noted over the inguinal region, root of the scrotum or the medial side of the thigh with

INVESTIGATIONS:

ULTRASONOGRAPHY: Normal bladder with infantile prostate noted. Right ovary noted measuring 32*18mm with few follicles seen within Scrotum revealed normal sized left testis 24*15mm with empty right hemiscrotum. INTERVENSION: Gonadal tissue found intra- abdominally with soft follicular ovarian tissue on one pole and firm testicular tissue on the other pole

INTRA-OP: Gonad excised from abdominal cavity contains both ovarian and testicular tissue Ovarian tissue shows corpus albicans, corpus luteum , focal aggregates of hemosiderin laden macrophages. Testicular tissue shows infantile seminiferous tubues containing sertoli cells. Leydig cells seen in interstitium. No granuloma and malignancy. Biopsy of the testicular tissue shows Infantile seminiferous tubules containing sertoli cells. Leydig cells are seen in the interstitium. No definite ovarian tissue identified. No germ cell neoplasia, granuloma and malignanacy

CONCLUSION:

Ovotesticular disorder of sex development is the rarest form among all disorders of sex development and characterized by the simultaneous presence of both ovarian and testicular tissues in the same individual. While it characteristically presents with ambiguous genitalia in neonates or infants, it may also present in adulthood. The management of a patient with ovotesticular DSD must be carried out by a multidisciplinary team.

DISCUSSION: Diagnosed as 46XX Ovotesticular DSD. Disorders of sex development (DSDs) are a group of rare and complex disorders characterized by abnormalities of chromosomal, gonadal, or phenotypic properties that determine sex development. The prevalence of DSD is reported to be 1.8 per 10,000 live births. Ovotesticular DSD rarest with prevalence of 1/20,000, and about 500 affected individuals have been reported to date. Characterized by presence of both ovarian and testicular It has been reported that only about 10% of all patients with ovotesticular DSD have a phenotypically normal or nearly normal male penis and diagnosed later during adolescence. Most commonly detected chromosomal makeup among patients with ovotesticular DSD is a 46, XX karyotype. The present case is an extremely rare one in that the external genitalia is nearly normal, and the patient (and his parents) had no doubt about any sexual abnormality until a progressive breast enlargement occurred,.