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Ovotestes Without A Uterus

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Case Report

A two-and-a-half-month-old boy presented to the Emergency Department with fever, periorbital edema, hypoalbuminemia, decreased oral intake for a day, two episodes of non-bloody yellowish vomit, decreased urine output of two wet diapers (normal of six per day), irritability, abdominal distention, and a swollen right scrotal sac. The patient was later found to have Streptococcus pneumonia bacteremia and was subsequently treated with ceftriaxone for ten days along with vaccination upon discharge.

His past medical history is significant for congenital nephrotic syndrome, Ebstein's anomaly, ambiguous genitalia (46XX/46XY = true hermaphrodite), hypothyroidism, and anemia of chronic disease. Genetic testing revealed a chimera 46, XY/46, XX/46 karyotype or true hermaphrodite (chimerism = two genotypes from different zygotes). A previous aCGH test also showed duplication of chr 2p22.1 which proved to be a familial polymorphism that is maternally inherited.

Obstetrical history is significant for premature delivery at 36 weeks, born to a 27-year-old G2P2002 woman from a normal spontaneous vaginal delivery. No teratogenic exposures during prenatal period were reported. He has two elder siblings who are healthy with no complications.

During his admission, the child's physical examination was significant for a reducible umbilical hernia, ambiguous external genitalia, an empty left scrotal sac, a 2cm phallus, and a very firm, solid, and enlarged right hemiscrotum. His laboratory findings were significant for an alpha fetoprotein over 1000 ng/ml along with marked leukocytosis of 22.500 cells/cc. No uterus or ovaries were identified on pelvic ultrasound. Scrotal ultrasound showed a complex mass adjacent to his right testicle demonstrating heterogenous echotexture. A radical

orchiectomy procedure was subsequently performed. The right testis, portion of right spermatic cord, and right epididymis were removed. Gross pathologic examination of the mass demonstrated suppurative and necrotizing inflammation with chronic inflammation and fibrosis. Ovotestis within the right testis was also noted. There was no evidence of neoplasm.

Discussion

Ovotestes falls under the classification of intersex disorder. True hermaphroditism occurs when both ovarian and testicular tissue are found within the gonads. It is very rare with only about 500 cases identified worldwide [1]. In 2006, a task force of 50 specialists sponsored by the European Society for Pediatric Endocrinology (ESPE) and the Lawson Wilkins Pediatric Endocrine Society (LWPES) proposed "Ovotesticular Disorder of Sex Development (DSD)" as the new nomenclature [2]. The new nomenclature as well as early gender assignment are important in normal psychological development.

The clinical features of true hermaphroditism include all have uterus, most have fallopian tubes, some have vas deferens [3]. In our presented case, ambiguous genitalia on physical exam and 46XX/46XY with chromosomal testing were consistent with true hermaphroditism. However, no uterus was identified on pelvic ultrasound.

The characteristic imaging feature of true hermaphroditism is the presence of an ovotestis or one testis and one ovary in the same patient [4]. A structure containing both testicular echotexture and follicles is the classical appearance of ovotestis on ultrasound. The ovotestis in our case did not have the classical appearance. Right scrotal ultrasound of our patient demonstrated a testicle with loss of its normal homogenous echotexture, otherwise nonspecific (Figure 1,2,3). French and Rodriguez suggested improved sensitivity of ultrasound detection of ovotestes with use of FSH injections to induce ovarian follicular development [5].

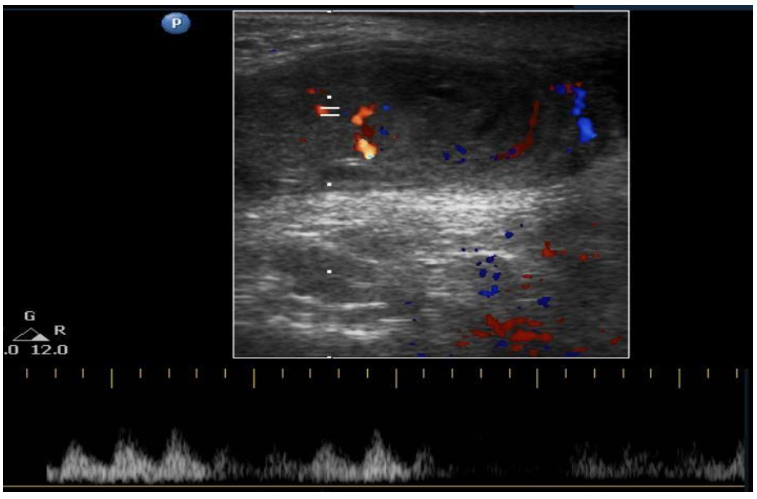

Figure 1: Scrotal ultrasound - Right testis longitudinal view. Right testicle is in the scrotum and measures 1.7 x 0.9 x 0.9 cm with normal vascular flow (green arrow). Thickened, echogenic, hypo vascular right epididymis (yellow arrow).

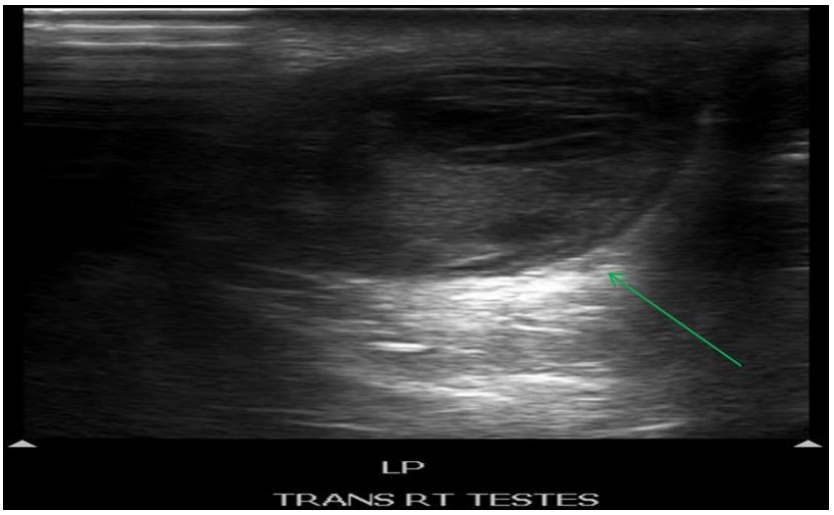

Figure 2: Scrotal ultrasound - Right testis transverse view. Transverse view of right testicle lower pole in the scrotum (green arrow).

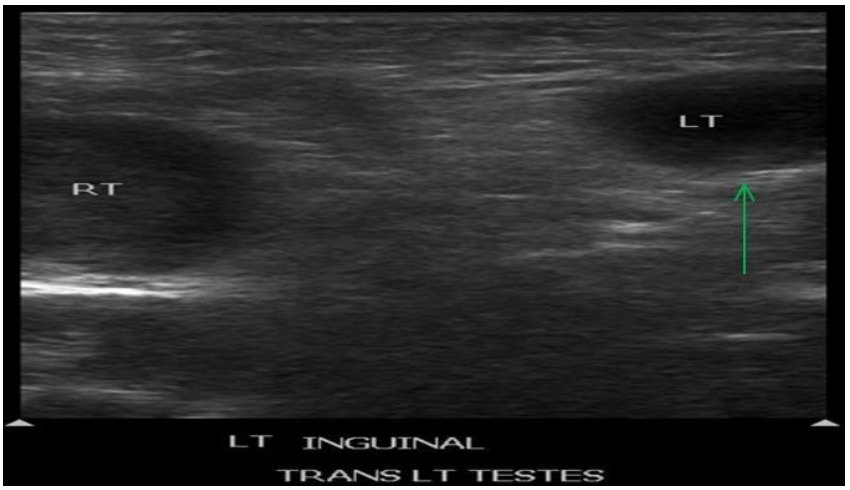

Figure 3: Scrotal ultrasound-transverse view of bilateral testes. There is no left testicle in the scrotum (green arrow). The undescended left testicle was identified in the inguinal canal, measuring 1.7 x 0.9 x 1.0 cm with normal vascular flow. Left epididymis is unremarkable (not included in this image).

It is difficult to diagnose ovotestes via imaging. Generally, ovotestes is first suspected on clinical exam, then confirmed with chromosomal testing and histopathology (Figure 4,5,6). The Ovarian portion of the ovotestis is frequently normal, whereas the testicular portion is typically dysgenetic. The risk of gonadal malignancy is approximately 10% in 46, XY true hermaphroditism and 4% in 46, XX true hermaphroditism. The most important aspect of management in true hermaphroditism is gender assignment, and may include gonadectomy followed by reconstructive surgery, and some patients may also need hormone replacement therapy during puberty [6].

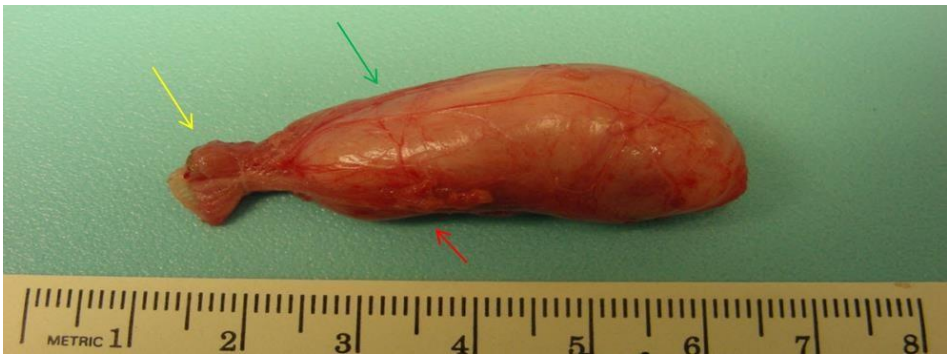

Figure 4: Gross - Right testicle. The specimen consists of right testicle covered by tunica vaginalis (green arrow), complete with a fragment of the spermatic cord (yellow arrow), epididymis (red arrow).

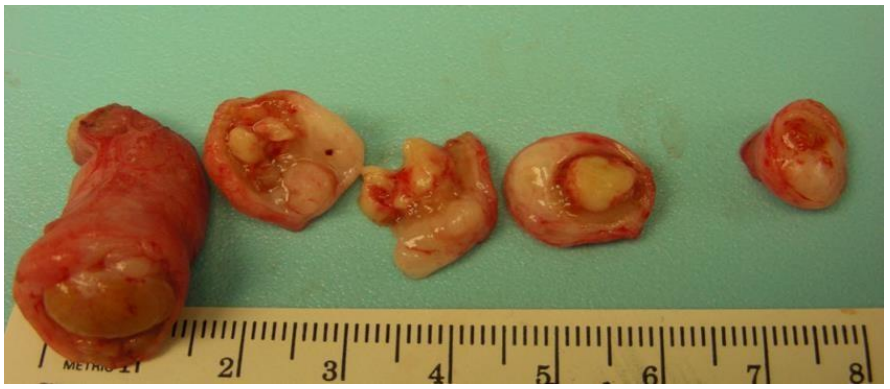

Figure 5: Gross - Right testicle. Upon cross-sectioning there is a pale-yellow soft mass measuring 0.7 cm in diameter at the lower pole of the testicle (yellow arrow). There is also normal testicular tissue present on sectioning located above and adjacent to the mass.

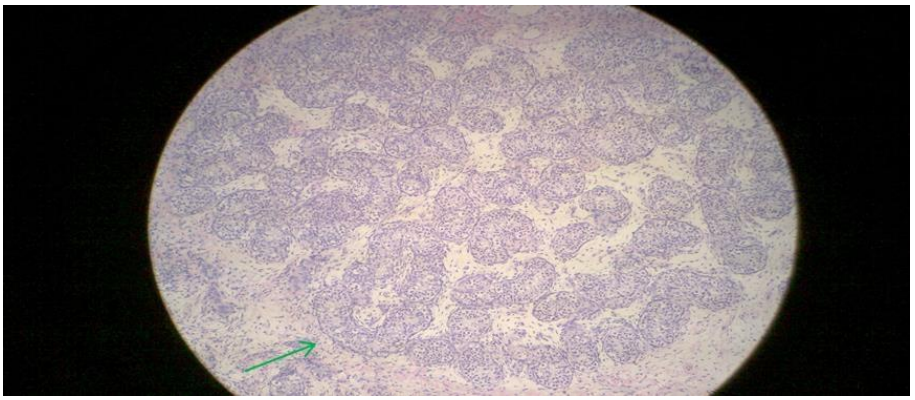

Figure 6: Histology - Right testicle. Ovotesticular Disorder of Sex Development (DSD) is characterized by the presence of both testicular and ovarian tissue. In this example, seminiferous tubules are seen (green arrow).

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