CASE REPORT

PERSISTENT MULLERIAN DUCT SYNDROME WITH TRANSVERSE TESTICULAR ECTOPIA: A RARE CASE REPORT

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ABSTRACT: We report a case of Persistent Mullerian Duct Syndrome (PMDS) with Transverse Testicular Ectopia (TTE) in a 27 yrs old male discovered during surgery for right irreducible inguinal hernia. Tension free hernioplasty and excision of pmlds was done. The specimen sent for histopathologic examination (hpe). Hpe revealed pmlds.

KEYWORDS: Irreducible hernia-two cord like structure separated by uterus like structure with both cornua leading to gonads-rudimentary uterus.

CASE REPORT: A 27 yrs. old unmarried male came with right groin swelling for 6 months which was reducible earlier, turned irreducible for past 8 hours. Patient had pain in right lower abdomen and vomiting. Vitals found to be normal. On examination tenderness present over right groin swelling, absent cough impulse, swelling irreducible, absent testis on left side, testis was palpable on right side. External genitalia and secondary sexual characters seem to be normal. Patient was not aware of the absence of the left testis. Patient was diagnosed as a right irreducible inguinal hernia with left undescended testis. All basic investigations taken and was found to be normal. Planned for emergency inguinal exploration and hernia repair.

INTRAOPERATIVE: Under spinal anaesthesia, rt parainguinal incision made, inguinal canal opened and following findings noted:

1. Indirect sac.
2. Sac contents- two cord like structure separated by uterus like structure with both cornua leading to gonads;
 - The uterus like structure was removed and both cord like structure along with gonads preserved and sent for hpe.
 - Hernioplasty done for right irreducible inguinal hernia.
 - Two cord like structure along with gonads placed in right hemiscrotum.

HPE REPORT: Macroscopic Appearance: Section shows slit like endometrial cavity measuring 5.5*4.2cms.

Microscopic Appearance: Section shows bundles of smooth muscle fibres enclosing endometrial glands with stroma.

No evidence of any gonads in section studied hpe report: confirmative of rudimentary uterus.

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DISCUSSION: Male pseudohermaphroditism (pmds) characterized by presence of uterus, cervix, fallopian tubes in normal 46xy male.^{1,2}

Tte- migration of testis to opposite side where both testis pass through same inguinal canal. Initially both male and female embryos have wolffian ducts and mullerian ducts.

Around 6-9 weeks of gestation, mullerian duct regresses due to mullerian inhibiting substance produced by sertoli cells in male.^{3,4}

Failure of regression leads to pmds.¹

Pmds-autosomal recessive disorder with uterus, cervix and fallopian tubes in 46xy male.²

Cryptorchidism-testicular descent affected due to entanglement, testis may be present in ectopic sites.

Very rarely associate with tte.^{5,6}

PMDS Presentations:⁶

1. Discovered during hernia repair.
2. Abdominal surgery.
3. Incidental findings in usg /mri.
4. Infertility.

Anatomical Variants:⁷

1. One testis in scrotum, uterus with derivatives in inguinal canal or brought made into it by gentle traction in presenting testis (most common type).
2. Contralateral testis (tte) in sac.
3. B/l cryptorchidism with testis embedded in broad ligaments in ovary position in respect to uterus.

Etiopathogenesis, differential diagnosis, operative strategy in pmds –not defined completely.

Figure 1

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Figure 2

Figure 3

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