PERSISTANT MULLERIAN DUCT SYNDROME ASSOCIATED WITH TRANSVERSE TESTICULAR ECTOPIA

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ABSTRACT:

Persistent mullerian duct syndrome with transverse testicular ectopia is a rare syndrome. We report a persistent mullerian duct syndrome (PMDS) associated **with** transverse testicular ectopia (TTE) in a 10-month-old male discovered during surgery for inguinal hernia and undescended testis.

Index words: Mullerian duct syndrome, transverse testicular ectopia, cryptorchidism

INTRODUCTION:

Persistent mullerian duct syndrome (PMDS) is a rare form of male pseudo-hermaphroditism, characterized by the presence of a "uterus and fallopian tubes, due to failure of mullerian duct regression in **genotypical** normal males[1,2]. More than 150 cases have been recorded, most of them in adults. The association between the persistent mullerian duct and transverse testicular ectopia is even more uncommon. In 1886 Von Cenhossek described the first case of transverse testicular ectopia and in 1895 Jordan described the syndrome of transverse testicular ectopia with persistant mullerian ducts[3].

CASE REPORT:

A 10-month-old- boy presented with left inguinal hernia and absence of right testis since birth. Examination of the groins showed normal phallus, left inguinal hernia, empty right hemiscrotum, impalpable right testis and normal left testis.

Urinalysis, complete blood count and serum, electrolytes were normal. **At exploration through left inguinal incision two spermatic cords were found separated by rudimentary uterus, thick walled vagina and ligamentus structure corresponding to the broad ligament. The two vasa deferentia were adherent to the thickened vagina. The left testis was lying in the left hemiscrotum but pulled up into the operative field during dissection . The right testis ends at left mid inguinal canal (Fig. 1). The two testes were biopsied, the two cord structures were carefully separated from the persisting mullerian structures and limited resection of rudimentary uterus and vagina was done. The associated inguinal hernia was repaired and bilateral orchidopexy with transposition of the right testis to the right hemiscrotum through suprapubic subcutaneous tunnel was performed.**

Post operative course was uncomplicated. The karyotype **confirmed** a male gender 46xy. Renal ultrasound was normal.

DISCUSSION:

Normally the testis is located in the scrotum at birth. Ectopic testes have been reported at different sites including superficial inguinal pouch, suprapubic, femoral, perineal region and at the base of the penis[4].

Migration of the testis to the opposite side where both testes **pass** through the same inguinal canal is known as transverse testicular ectopia (TTE). Over 100 cases of TTE has been reported in the literature[5,6].

Persistant mullerian duct syndrome is rare, characterized by the presence of well developed or rudimentary uterus, cervix, vagina and fallopian tubes in normal 46xy male.

The presence of PMDS with TTE is even more uncommon. In most cases the PMDS is discovered during surgery for inguinal hernia or cryptorchidism or by the presence of transverse testicular ectopia . Our patient was found to have PMDS with TTE during surgery for herniotomy in a child with contralateral undescended testis. PMDS represents only a small number of male pseudo-hermaphroditism. The external genitalia in these patients is normal in shape and size with a central **glanular** urethral meatus.

The preoperative diagnosis of PMDS and TTE is practically impossible. Adamsbanm et al[7] recommended routine pelvic and inguinal area ultrasonography in bilaterally cryptorchidism patients and in patients with inguinal hernia of unusually hard consistency. Recently, with experience of laparoscopic surgery for impalpable testis, the diagnosis of transverse testicular ectopia was possible. Fairfix et al[8] reported a 14-month-old child with transverse testicular ectopia diagnosed laparoscopically.

As PMDS and TTE is usually discovered incidentally during surgery for undescended testis or inguinal hernia, the optimal surgical approach should include testicular biopsies, herniotomy, orchidopexy, and excision of mullerian duct remnants. The **vasa** deferentia are usually adherent to the lateral walls of the **vagina** and for this reason maximum care should be taken to dissect the vas deferens away from the mullerian duct structures. Some authors believe that excision of mullerian duct structures and scrotal orchidopexy are not possible without sacrificing the **vasa** deferentia[2]. In our patient it was possible to dissect the **vasa** deferentia from the rudimentary uterus and vagina, perform scrotal orchidopexy and excision of mullerian duct structures without risking the **vasa** deferens. We believe that every effort should be made to preserve vas deferentia and testes for possible future fertility. This approach is reported by many authors[1,3,7].

Fertility has been reported rarely in **a** few cases[2,9], Martin et al[3] had reported a 32-year-old man with transverse testicular ectopia and persistant mullerian duct who had normal sperm count but the motility index was zero implying an intrinsic defect in spermatogenesis. The other possible cause of low sperm counts and poor motility in most of these patients is the partial duct obstruction.

There have been reports of embryonal cell carcinoma, seminoma, choriocarcinoma and teratoma in patients with cryptorchidism and persistant mullerian duct. However, the malignancy incidence appears to be similar to that of normal **cryptorchid** child[2,10,11].

In conclusion, conservative surgical approach for this rare syndrome in the form of orchidopexy and partial excision of mullerian duct remnants without risking vas deferens is recommended, long follow up will be needed for assessment of the fertility in these patients.

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LEGENDS:

Figure 1: Intraoperative view showing both testes, fallopian tubes and **rudimentary** uterus.

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