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

PRAKASH KUMAR J
Department of General Surgery, GOVERNMENT MOHAN KUMARAMANGALAM MEDICAL COLLEGE

Abstract : Transverse Testicular Ectopia (TTE) is an extremely rare congenital anomaly where in both the testes lie in the same scrotum. It is mostly associated with inguinal hernia. The current literature reports around 100 cases of TTE out of which 20-49 had remnants of the Mullerian duct. Careful management of such a case is required to safeguard the fertility of patients. Hence, I describe a 2 year old male child who was admitted for inguinal hernia and presence of both the testes on the same side. On exploration, found to have mullerian duct remnants too which is a very rare entity.

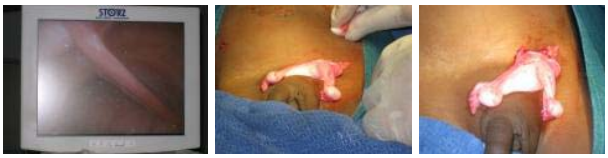
Keyword : Transverse Testicular Ectopia, Persistent Mullerian Duct Syndrome, Herniotomy, Orchidopexy.

INTRODUCTION:

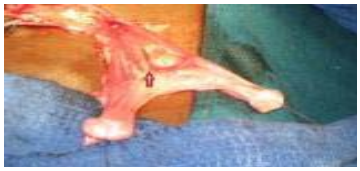
Transverse Tesicular Ectopia (TTE) is a condition wherein both the testes descend through the same inguinal canal and remain in the same scrotum and the other scrotum remains empty. Persistent Mullerian Duct Syndrome (PMDS), in spite of being an intersex condition, the affected persons are genetically males, who can retain their fertility. Only about 100 cases of Persistent Mullerian Duct has been reported so far in conjunction with TTE . This was first described in Autopsy specimen by Lenhossek in 1886.1 TTE are classified into 3 different types according to the association of various anomalies:2 Observed only with Inguinal Hernia (40-50%) Observed with PMDS (30%) Observed with additional anomalies like hypospadias, pseudohermaphroditism, scrotal anomalies (20%)

CASE REPORT:

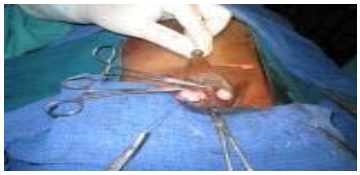
A 2 year old male child, oldest of the two children born to non consanguineously married healthy parents was brought with complaints of left sided inguinal swelling and absent testis in the right hemiscrotum which they had noticed in the recent past. The parents gave no other complaints. The child had attained normal developmental milestones. On examination, the child was alert. No obvious congenital abnormalities on inspection. General condition was stable. Systemic examinations were unremarkable. Per abdomen was soft, no obvious mass palpable. Left hernial orifice was full. External genital examination showed maldeveloped hemiscrotum on the right side with absent testis on the right hemiscrotum. Two firm swellings were palpable one in the left hemiscrotum and the other in the left inguinal region. A swelling was palpable on the left inguinal region that was spontaneously reducible and cough impulse was positive. Penis appeared normal. Child was evaluated routinely for surgery. Basic blood investigations were within normal limits. An Ultrasound of the Abdomen and Scrotum were done that showed left indirect inguinal hernia, left testis within the scrotum and the right testis within the left inguinal canal with rudimentary structures ? Rudimentary uterus. Child was taken up for surgery and a diagnostic laparoscopy was done to visualise both the testes separately.



Vas evident on either sides



Pointer showing mullerian duct remnants



Bilateral orchidopexy

Immediate post op picture

We found that the left testis was within the left scrotum and the right testis along with the vessels and vas had crossed over to occupy the left inguinal canal and had dragged the peritoneum along with it producing a left inguinal hernia. The ports were closed and a left inguinal skin crease incision was made and left herniotomy was done followed by bilateral orchidopexy. Orchidopexy of the right testis was done by pulling the testis and its vasculature and vas deferens towards the opposite median raphae and to our surprise we found that a rudimentary structure was closely applied to the region where both the vas deferens separated. A careful biopsy was taken from the structure, and orchidopexy was performed. The histopathological examination reported that the biopsied structure was a Persistent Mullerian Ductal Structure and that there was no evidence of malignancy in it. Post procedure child was comfortable and wound was healthy and hence discharged.

DISCUSSION:

PMDS in spite of being a intersex condition, the affected persons are genetically males who can retain their fertility. This condition is an autosomally recessive condition. This condition is also described as Hernia Uteri Inguinalis as it is always associated with inguinal hernia.3

ETIOLOGY:

1. Lack of MIS.
2. Insensitivity to MIS receptor.
3. Defective AMH gene.

PATHOLOGY:

Anti Mullerian Hormone (AMH) is secreted by the primitive Sertoli cells as one of the earliest sertoli cell product and causes regression of the mullerian ducts. Mullerian ducts are only sensitive to AMH action around the eighth week of amenorrhoea and Mullerian duct regression is complete by the end of ninth week. The AMH induced regression of the Mullerian duct occurs in cranio caudal direction via apoptosis. The AMH receptors are located on the Mullerian duct mesenchyme and transfer the apoptotic signal to the mullerian epithelial cell presumably via paracrine mediators. The Wolffian duct differentiates into epididymis, Vas deferens, and seminal vesicles under the influence of testosterone, produced by the fetal Leydig cells. Another important hormone that causes mullerian duct regression is Mullerian Inhibiting Substance (MIS) that is either absent or insentivity to its receptor leads to persistence of mullerian duct remnants.

MANAGEMENT:

A child with TTE and PMDS is a genetic male and retains his fertility and hence removal of the duct structures is not necessary. Since this condition is almost always associated with inguinal hernia, herniotomy and fixation of the bilateral testes to the scrotum, to provide accessibility to examine the testis suffices. But, if the testis could not be mobilised further, an orchidectomy may be done as the testis remaining intra abdominally has high chance of being malignant4. Mullerian duct structures need not be removed as there will be no malignant degeneration of those structures.1 These children are said to be fertile and hence any heroic attempt to remove the mullerian duct structures should be avoided as this act may inadvertently injure the vas deferens or testis and render the child infertile. The parents are counselled and reassured about this condition and a semen analysis at a later date to prove the child's fertility may be done for the psychological acceptance of the parents.

CONCLUSION:

This entity should be borne in mind when a child presents with inguinal hernia on one side and undescended testis on the opposite side. PMDS can present in adult males too. These individuals can lead a healthy sexual life provided Orchidopexy is done.

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