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Transverse Testicular Ectopia: Institutional Experience

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Abstract

Transverse testicular ectopia (TTE) or crossed testicular ectopia is an extremely rare anomaly characterized by migration of one testis towards the opposite inguinal canal. Treatment is difficult and includes laparoscopic or surgical exploration.[1] The typical presentation of crossed testicular ectopia is ipsilateral inguinal hernia and contralateral undescended testis.[2] We present our experience of 7 cases of transverse testicular ectopia (TTE).

Keywords: undescended testis; ectopic testis; transverse testicular ectopia; persistent Mullerian duct syndrome; hernia uterine inguinale; laparoscopic management of transverse testicular ectopia.

Introduction

In transverse testicular ectopia (TTE), both testes descend through the same inguinal canal into the same scrotal sac. Cases are usually discovered during clinical examination or surgery for an inguinal hernia or cryptorchidism.[1] Several theories have been put forth to explain the embryogenesis of this anomaly. Persistent Mullerian duct syndrome (PMDS) is a rare syndrome and is a form of male pseudohermaphroditism; the Mullerian duct derivatives (uterus, Fallopian tubes and upper vagina) are present in an otherwise normally differentiated male with a 46, XY karyotype. MIS, is produced by fetal testicular Sertoli's cells, is responsible for the involution of embryonic Mullerian structures in normal males and this hormone also plays a role in decent of the testis which explains crossed ectopia of testis.[3] . Familial association has been found in some cases.[1,2] or by the presence of transverse testicular ectopia (one of the rarest form of testicular ectopia); PMDS associated with TTE is much rarer.[3]

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Materials and Methods

A retrospective study of TTE patients managed in our institution during the period from January 2012 to April 2019 was done in the study. Data regarding the age, side of hernia, persistence of Mullerian structures were noted by clinically or radiologically or during laparoscopic/ open surgery. No separate ethical committee clearance was obtained, but separate consent was obtained from parents of the patients during surgery for publication of their treatment material and photos.

Results

In this study totally seven cases were included, all managed in our institute during the study period. All our patients had contralateral inguinal hernia. Four patients presented with absent testis and clinical examination with ultrasonogram revealed the presence of contralateral hernia (Fig. 1, Fig. 2) and three patients presented with inguinal hernia and routine clinical examination confirmed the presence of non palpable undescended testis on the contralateral side. Left undescended testis with right inguinal hernia was present in 4 of our

patients. Age of presentation was from 8 months to 7 years with a mean age of 2.5 years. Karyotyping was done for all our patients after the initial diagnosis as very high association of persistent Mullerian duct syndrome has ben reported.[1] MRI was done when the ultrasound could not detect the testis and to know the details of the Mullerian remnants.

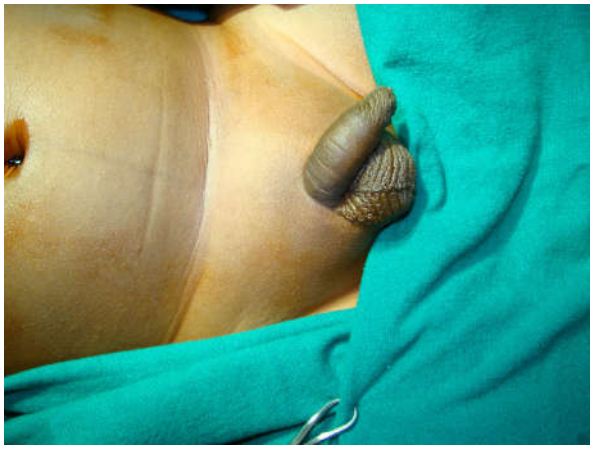

Fig.1: Appearance of genitals, left hemiscrotum hypoplastic.

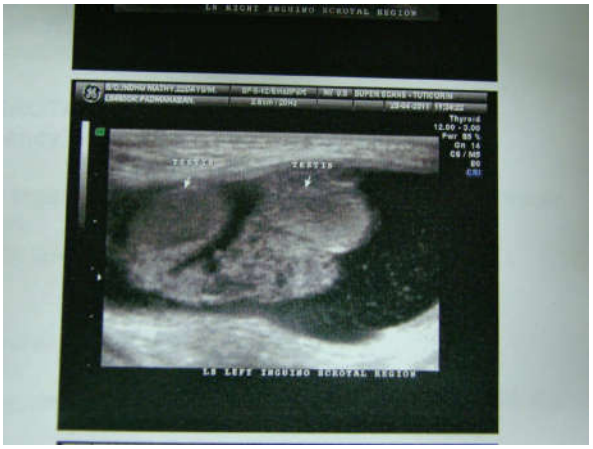

Fig. 2: USG image showing both testes on left side in the hernial sac.

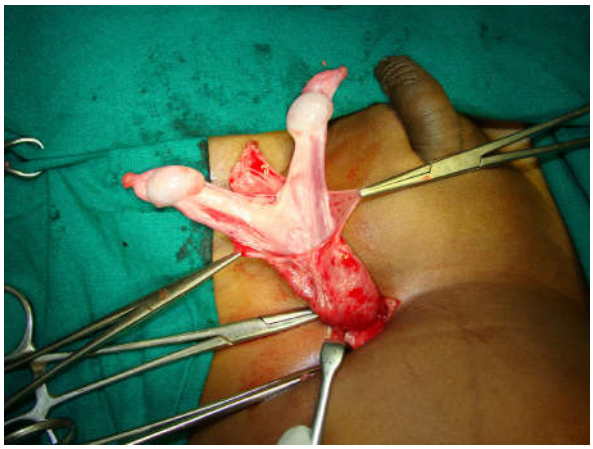

Fig. 3: Both testis in the same hernial sac.

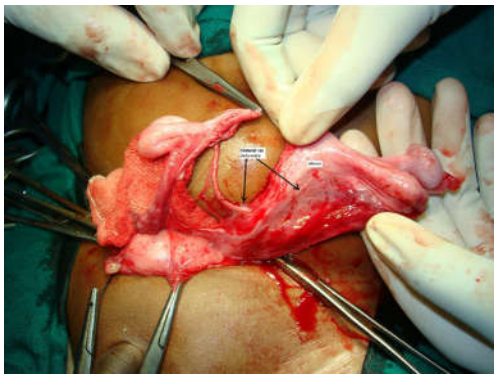

Fig. 4a: Presence of rudimentary uterus and tubes in close association with the vas.

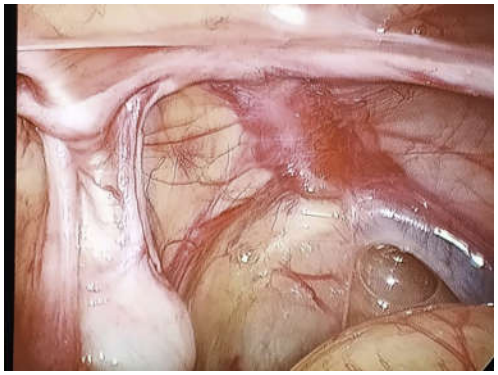

Fig. 4 b: Laparoscopic view of the right testis near the left internal ring, Mullerian structures and hernial sac visualized.

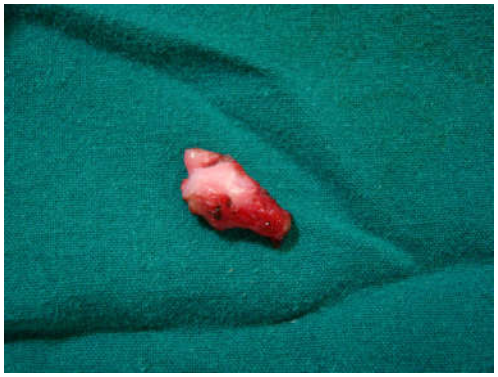

Fig. 5: Excised Mullerian structures.

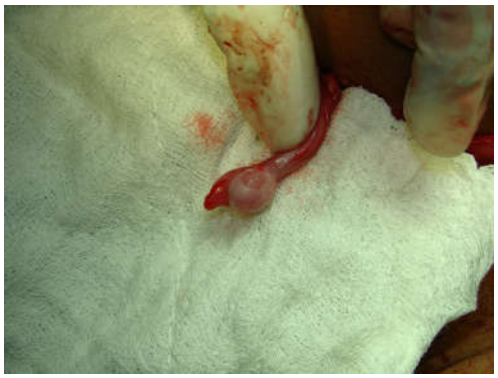

Fig. 6: Surface irregularity of the right testis which was biopsied.

All our patients except the first two in this series underwent a diagnostic laparoscopy to begin with and laparoscopic management of the undescended testis followed by laparoscopic or open repair of the inguinal hernia. All the cases were followed up with ultrasonogram at the end of one year and they were all asymptomatic.

Discussion

Transverse testicular ectopia is an extremely rare but well recognized entity in which both gonads migrate towards the same hemiscrotum.[1] The clinical findings are usually symptomatic inguinal hernia on one side to which the ectopic gonad has migrated and an impalpable testis on the other side. A number of theories have been proposed to explain the etiology of ectopic testis Several theories have been proposed to explain the embryological basis of transverse testicular ectopia. Some of the theories are: both testes arising from same genital ridge, adherence and fusion of the developing Wolffian ducts taking place early and descent of one testis causing the second testis to follow it. Defective implantation of the gubernaculum testis or an obstruction of the inguinal ring preventing testicular descent on the ipsilateral side, defective ipsilateral gubernacular development predisposing to TTE.[4,5,6,7]

Based on presence of associated anomalies three types of TTE are described.[1] Type I: Associated with inguinal hernia (40–50%).[2] Type II: Associated with persistent or rudimentary Mullerian Duct structures (PMDS) (30%).[3] Type III: Anomalies other than PMDS such as hypospadias, pseudohermaphroditism and scrotal anomalies (20%).[1,2]

Presence of both testes in the same hernial sac together with uterus and uterine tubes is a common association and a was found in four (57%)of our patients. Renal anomalies, hypospadias or other system abnormalities were not noticed in our patients.

The treatment of transverse testicular ectopia is focused on the detection of associated congenital abnormalities and placement of ectopic testis into anatomical position. This preserves fertility and allows monitoring for the development of malignancy.

Laparoscopy helps visualizing the anatomy of internal inguinal ring and intraperitoneal space, and bilateral orchipexy in TTE without confusion regarding the laterality of the testes. Especially in case of impalpable testis with disorders of sexual development, laparoscopy is very useful tool for localizing the testis and internal genitalia at one time.[9,10] All our patients underwent diagnostic laparoscopy because in general all our patients with non palpable undescended testis are managed laparoscopically. The first two cases in our series underwent initial inguinal exploration followed by extension of inguinal incision medially to explore the pelvis and complete the excision of Mullerian remnants and orchipexy.

All other patients underwent trans umbilical diagnostic laparoscopy, which allows detection of Mullerian structures and testis..The testis was inside the hernial sac in five of the patients. (Fig. 3) and along the course of the contralateral vas in the perivesical region in two of the patients. The two vas deferences were arranged close to the bladder in most of the cases but they could be separated with some difficulty without affecting their blood supply.

Four out of seven patients had Mullerian remnants (Fig 4a and 4b) in the form of rudimentary uterus. Though it is generally agreed that malignancy arising from the retained Mullerian structures is very rare we did the excision of uterine structure in all cases without damaging the vas deference or the blood vessels.[12,14] The rudimentary uterus is thus biopsied. (Fig. 5). The histopathological examination of the rudimentary uterine and tubal structures showed atrophied structures. The excision and splitting of the residual structure gave some additional length to the cord structures. The blood supply to the ectopically lying testis seemed to be from small branches of the contralateral testicular artery or from the blood vessels of the tunica vaginalis and hernial sac. No attempt was made to dissect and identify the blood supply due to the fear of damaging the blood supply. But in two cases the blood supply was clearly arising from vessels that were crossing from opposite side probably the testicular vessels of the affected side. Testicular epididymal dysjunction was found in all the cases and testis was almost always small as compared to the contra lateral testis. Testis had certain abnormal texture (Fig. 6) and irregularity in two of the cases and biopsy was done in these cases to rule out presence of ovarian tissue. But the histopathology did not show any ovarian tissue but did show hypoplastic seminiferous tubules in the abnormal tissue.

After mobilizing the testis with its vas and vessels along with a band of peritoneal tissue or the Fallopian tube component of the Mullerian tissue

the testis was brought to the contralateral scrotum by direct approach. Though transseptal orchipexy has been described in literature, we found it easier to do the regular orchipexy through direct route which is also well documented in the literature.[13] Trans-septal orchidpexy is recommended when vasa deferentia are fused.[8,11]

Conclusion

Though there have been previous studies of transverse testicular ectopia, this study is one with a significant number of seven patients. In this study we have observed that careful clinical examination and ultrasonogram evaluation can diagnose the location of testis and associated persistent Mullerian duct structures preoperatively. We have done karyotyping for all the patients which helped as to rule out other disorders of sexual differentiation and all of them turned out to be 46XY. Persistent Mullerian duct syndrome was present in 57 percent of our cases, MRI might be useful in some cases where the ultrasound does not show all the details of the anomaly. Laparoscopy is invaluable in all cases of TTE to locate the testis, to visualize and manage the Mullerian remnants, to do the orchipexy and finally repair the hernia. So we found that laparoscopy through umbilical port and two lateral ports (all 5mm) to be very effective in diagnosing and managing these cases. We find it advantageous to excise the uterus and tubes at least in part to facilitate the orchipexy and also to examine the structure histopathologically. The biopsy will be useful from medicolegal point of view. We find orchipexy through direct route to be easier along with midline splitting of residual stump of the uterus after excising the Mullerian structures. Trans septal orchipexy may be useful if the two vases are inseparable due to common blood supply.

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