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Undescended Testes: The Hidden Normal or Abnormal Anatomy and The Value of Laparoscopy

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1. Abstract

1.1. Background

Persistent Mullerian duct syndrome (PMDS) is characterized by the presence of a uterus, fallopian tubes and upper vagina in otherwise a phenotypically and genotypically normal male. Not uncommonly, it is seen in association with transverse testicular ectopia (TTE).

1.2. Methods

Three male children (14 months, 15 months and 3 weeks old) were referred because of bilateral undescended testes. Clinically, they were found to have bilateral impalpable testes and sub-coronal hypospadias in two of them. MRI of the lower abdomen and pelvis was done for one and showed the right testis in the pelvis but the left one could not be visualized. Abdominal and pelvic ultrasound of another revealed a uterus like structure seen posterior to the urinary bladder measuring grossly about 2.4 x 1.4 x 1 cm and bilateral undescended testes. All had a normal male karyotype (46XY). Two of them underwent laparoscopy and in the third, the diagnosis was made during a right inguinal herniotomy.

1.3. Results

In one, laparoscopy revealed no left testis but there was a uterus-like structure, 3-cm long on the right side with tubes on both sides. The right gonad was seen on the right side. Exploration was done through the right inguinal region incision. There was a uterus-like structure with two fallopian tubes and two testes on the same side. Bilateral gonad biopsies were done which revealed normal testicular tissue with fibrosis. The diagnosis of PMDS with transverse testicular ectopia (TTE) was made. Ten weeks later, he underwent bilateral orchidopexy and excision of the uterus, fallopian tubes and remnant of the vagina while preserving both vases and vessels which were running adherent to the uterus. Laparoscopic exploration of the second patient showed the presence of a normal looking uterus with fallopian tubes and bilateral undescended testes that were not fully developed. The diagnosis in the third patient was made at the time of right inguinal herniotomy.

1.4. Conclusions

PMDS is rare anomaly, seen in an otherwise normal males with normal external genitalia. Most cases are diagnosed as a surprise at the time of surgery for an inguinal hernia or undescended testes. Physicians caring for these patients should be aware of this and surgeons should be familiar with the different surgical options. PMDS should be considered in all cases of bilateral impalpable undescended testes.

1.5. Introduction

Undescended testes are one of the common congenital anomalies in the pediatric age group and although the majorities are palpable and amenable for surgical correction, there is about 20% that are not palpable [1]. The recent advances in minimal invasive surgery have simplified the management of those with impalpable testes [2, 3]. Occasionally however the procedure is not without surprises. This report describes three cases of PMDS associated with bilateral impalpable undescended testes and TTE in one of them. Aspects of diagnosis and management are also discussed.

2. Case Reports

2.1. Case 1

A 14-month-old male infant was referred to our hospital because of bilateral undescended testes. Clinically, he was found

to have bilateral undescended testes and subcoronal hypospadias. Both testes were not palpable. His penile length was normal. MRI of the lower abdomen and pelvis showed the right testis in the pelvis but the left one could not be visualized and no other abnormalities could be detected. The child was planned for laparoscopic assisted left orchidopexy and right orchidopexy. Laparoscopy revealed a closed left internal inguinal ring. No left testis was seen. There was however a uterus-like structure, 3-cm long on the right side with tubes extending on both sides. The left testis, vas and vessels were not visualized. The right gonad was seen coming in and out of the right internal ring. Exploration was done through the right inguinal region. There was a uterus-like structure with two fallopian tubes and two gonads on each side (Figure 1). It was decided to do bilateral gonad biopsies and delay the definitive procedure till after further evaluation. Post-operatively, his testosterone level was 0.32 nmole/L (normal 0.00-1.0 nmol/L). His chromosomal analysis showed a 46 XY pattern. The pathological examination of both gonad biopsies revealed normal testicular tissue with fibrosis. The diagnosis of PMDS with TTE was made. Ten weeks later, he underwent bilateral orchidopexy and excision of the uterus, fallopian tubes and remnant of the vagina while preserving both vases and vessels which were running adherent to the uterus. The pathological examination of the resected specimen was that of a rudimentary uterus and fallopian tubes. Post operatively, he did well and on follow-up 2 years later, he was found to have a right inguinal hernia which was repaired laparoscopically and both testes were palpable in the scrotum. They were of normal size but the right one was slightly smaller than the left one.

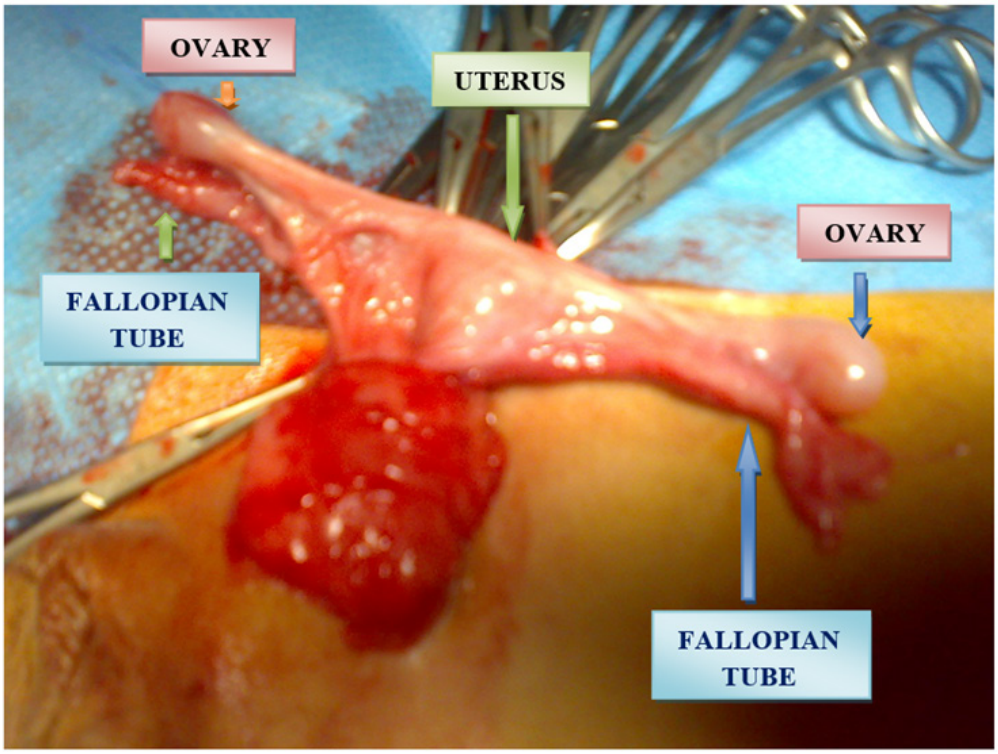


Figure 1: Intraoperative photograph showing all the features of persistent mullerian duct syndrome.

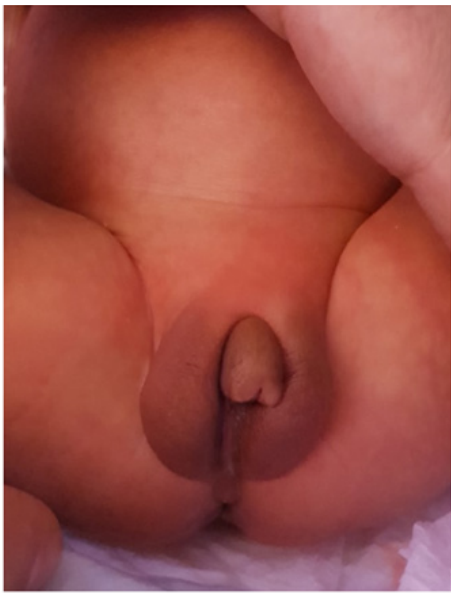


Figure 2: A clinical photograph showing a child with hypospadias and bifid scrotum.

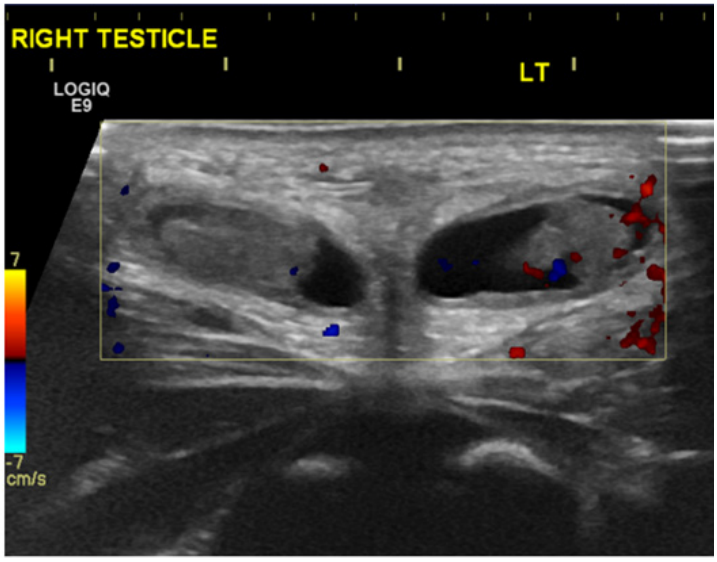


Figure 3: A pelvic ultrasound showing bilateral undescended testes.

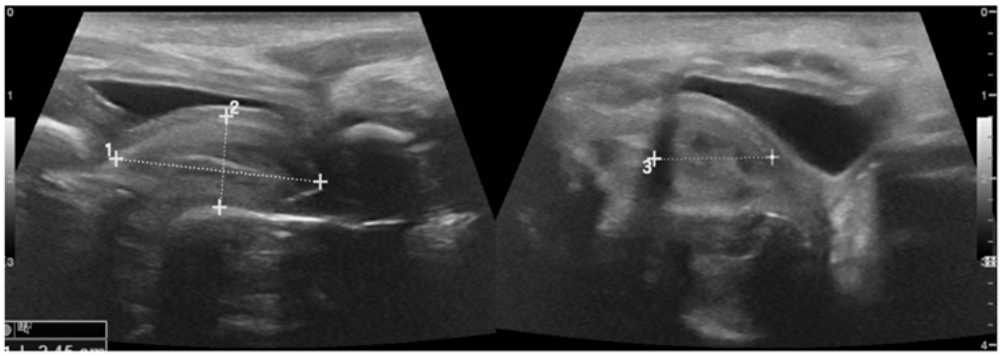


Figure 4: A pelvic ultrasound showing a uterus.

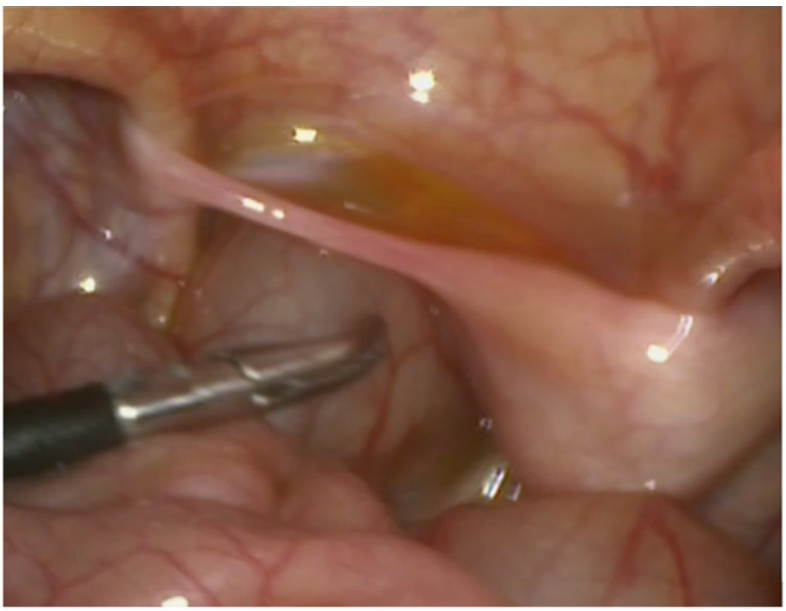
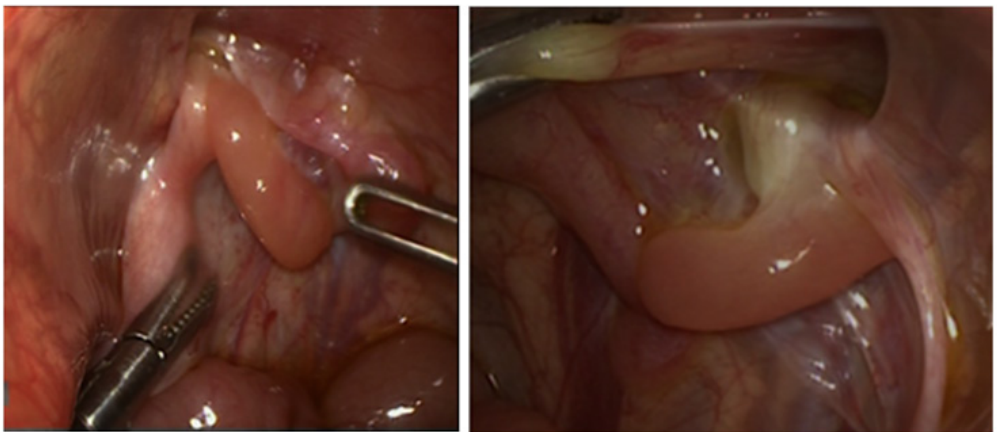


Figure 5: A clinical intraoperative picture during laparoscopy showing a uterus and a fallopian tube.



Figures 6A and 6B: Clinical intraoperative pictures during laparoscopy showing a rigt and left undescended testis.

2.2. Case 2

A 15-month-old male presented to another hospital with right inguinal hernia. He was also found to have right multicyclic kidney. He had an isotope scan which showed a normal left kidney and a non- functioning right kidney. He underwent right inguinal herniotomy and during surgery a uterus and a fallopian tube were found in the hernia sac. The contents were reduced and right inguinal herniotomy was done. Postoperatively, he did well and was referred to our hospital for further evaluation. His chromosomal analysis showed a normal male karyotype (46XY). He was found to have a normal penis with bilateral impalpable undescended testes. He was operated on and through a transverse lower abdominal incision, the peritoneal cavity was opened. He was found to have bilateral undescended testes which looked normal. There was also a uterus, two fallopian tubes and a small vagina. He was diagnosed as persistent mullerian duct syndrome. The uterus was divided in the midline to protect the close by vas and bilateral orchidopexy were done. Postoperatively he did well and on follow up, he was found to have a well-developed scrotum with palpable testes in the scrotum. Six months later, he underwent right nephrectomy for a large multicyclic dysplastic kidney and postoperatively, he did well.

2.3. Case 3

A 3-week-old male, a product of full term by caesarian section was noted to have ambiguous genitalia at birth. There was a strong family history of ambiguous genitalia. Two older siblings were positive for MAP3K1 gene mutation and presented with hypospadias. Their hypospadias was surgically corrected and they are developing well with no abnormalities. Clinically, he was noted to have, a developed normal penis with hypospadias and bilateral inguinal hernias. His karyotype showed 46XY and al hormonal studies (FSH, LH, Estradiol, Testosterone, DHEA, 17 OHP and ACTH) were normal. Abdominal and pelvic ultrasound revealed a uterus/cervix like structure seen posterior to the urinary bladder measuring grossly about 2.4 x 1.4 x 1 cm.

Laparoscopic exploration showed the presence of a normal looking uterus with fallopian tubes and bilateral testes that were not fully developed. The parents wanted to delay the definitive surgery.

3. Discussion

The first laparoscopic diagnosis of impalpable undescended testes was made by Cortesi, et al in 1976 [2]. Since then, several series were published describing the various laparoscopic techniques for both the diagnosis and treatment of impalpable testes [3]. Besides its advantages as a minimal invasive approach and its better cosmetic results, laparoscopy proved to be valuable in diagnosing other unsuspected abnormalities including the diagnosis and treatment of intersex disorders [4, 5]. This was the case in two of our patients who had bilateral impalpable undescended testes and because of laparoscopy they were discovered to have PMDS associated with TTE in one of them. The diagnosis in the third patient was made at the time of right inguinal herniotomy. All three patients had bilateral impalpable undescended testes.

PMDS which is also called hernia uteri inguinal is a rare congenital abnormality which results from a mutation in the gene encoding anti-Mullerian hormone or by a mutation in the anti-Mullerian hormone receptors [6]. Embryologically, the fatal testes secret two hormones. The Ledge cells secrets testosterone which is necessary for the development of the Wolffian ducts into the epididymis, vas deferens and seminal vesicles. The Sertoli cells on the other hand secrete the Mullerian inhibiting hormone which causes regression of the Mullerian ducts that usually develop into the uterus, fallopian tubes and upper third of the vagina [7]. PMDS can be caused by deficiency or failure in the production of the Mullerian inhibiting hormone or abnormality in its receptors. As a result of this, the Mullerian ducts fail to regress and develop into a uterus, fallopian tubes and upper vagina in otherwise a normal male with testicular gonads and 46XY chromosomes [6, 8]. The presence of consanguinity in some of the reported cases as well as its occurrence in several pairs of brothers supports an autosomal male-restricted mode of inheritance [8]. Others suggested an x-linked mode of inheritance [9].

Classically, PMDS is seen in an otherwise normal male with normal external genitalia who presents with unilateral or more commonly bilateral undescended testes and or inguinal hernia. It is also called hernia uteri inguinal because at the time of hernia repair, a uterus and fallopian tubes may be found in the hernial sac as seen in two of our patients. An association between PMDS

and hypospadias was reported before [10, 11]. This was the case in two of our patients who also had subcoronal hypospadias. The reason for this association is not known. Most cases of PMDS are diagnosed as a surprise at the time of surgery for an inguinal hernia or undescended testes. Rarely, the diagnosis is suspected preoperatively during evaluation of undescended testes as was seen in one of our patients. There is however a strong association between PMDS and TTE. PMDS is present in 30%-50% of all cases of transverse testicular ectopia and in these cases cross-orchidopexy becomes a necessity [12, 13]. One of our patients had PMDS associated with TTE. The surgical management of PMDS is still controversial. Since these cases are discovered incidentally, a staged procedure is the most commonly accepted option. During the initial surgery, bilateral testicular biopsies are done followed by replacement of the uterus, fallopian tubes and testes into the pelvis and inguinal herniotomy. Once the diagnosis is confirmed, definitive surgery is planned. The confirmation of the diagnosis includes chromosomal analysis, hormonal assay including HCG stimulation test and the result of testicular biopsies. There is however still controversy whether to remove the Mullerian remnants or not. There are those who advocate leaving the Mullerian remnants to avoid injury to the vas deferens and testicular vessels at the time of their resection [11]. On the other hand, there are those who strongly recommend their removal [6, 14]. Although very rare, there were two reports of clear cell adenocarcinoma of the remnant uterus in PMDS [15, 16]. Add to this the fact that the remnant uterus can hypertrophy causing pain and discomfort and removal of the uterus facilitates orchidopexy. The most commonly performed procedure is bilateral proximal salpingectomy leaving the fimbriae with the epididymis, hysterectomy and bilateral orchidopexy [6]. It is important to avoid injury to the vas and vessels at the time of hysterectomy. One way to achieve this is to leave a pedicle of the myometrium and the fimbriae attached to the epididymis. Others advocated splitting the uterus in the midline bringing the testis with the vas deferens and attached uterine tissue into the scrotum [17]. In one of our patients, it was possible to separate the vas deferens from the rudimentary uterus and vagina. In another patient, the uterus was split in the midline and the testis with the vas deferens and attached uterine segment were mobilized into the scrotum. We like others advocate removing the Mullerian duct remnants unless this is going to result in injury of the closely related vas deferens where we advocate splitting the uterus in the midline to increase the length and facilitate orchidopexy. With the recent advances in minimal invasive surgery, laparoscopy is being increasingly used both for the diagnosis and management of PMDS including testicular biopsy, orchidopexy and herniotomy [10, 12, 18]. Follow-up of these patients is also important. There is a 5%-15% risk of testicular malignancy in these patients which is not different from that in patients with undescended testes [19, 20]. Most of these patients however are infertile because of azoospermia, low motility index or ductal obstruction [21]. It is also important to check the result of testicular biopsy. These usually show testicular tissue with variable degree of fibrosis which may necessitate testosterone replacement at the time of puberty in those with hypoplastic or fibrotic testes.

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