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PERSISTANT MULLERIAN DUCT SYNDROME

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ABSTRACT

Persistent mullein duct syndrome is a rare type of male pseudo hermaphroditism causes by deficiency in Anti Mullerian hormone (AMH) or defect in its typell receptor. Internal male pseudo hermaphroditism in which Mullerian duct derivatives are seen in a male patient. This syndrome is characterized by the persistence of Mullerian duct derivatives (i.e. uterus, cervix, fallopian tubes and upper two thirds of vagina) in a phenotypically and karyotypically male patient. In this article we present the USG and MRI features of a case of PMDS with bilateral cryptorchidism and left sided inguinal hernia, containing the uterus and fallopian tubes. The optimal surgical management of the testes and mullein duct structures in patients with persistent mullein duct syndrome. Surgical management of persistent mullein duct syndrome and extracted information regarding the etiology, pathogenesis, and treatment of this disorder. In children, doctors typically discover Persistent mullein duct syndrome by chance during surgery correct one or both issues. The diagnosis of the PMDS is made radiologically and that the detection of Mullerian inhibiting factor is mandatory. A diagnosis of PMDs was made on histologically evaluation subsequent to abdominal orchiectomy. The PMDs diagnosed on laparoscopy initially biopsy of these remnants and gonads was done followed by excision of remnants by Laparotomy approach

INTRODUCTION

Persistent mullein duct syndrome (PMDS) was first described by Nilson in 1939. First description of transverse testicular ectopia was made by Lenhossek in 1886. Jordan described syndrome of transverse testicular ectopia with persistent Mullerian ducts. Persistent mullein duct syndrome (PMDS) is a very rare condition with less than 300 cases described in the literature (1,2). This rare entity shows cases adolescent who are phenotypically males with 46 x 4 karyotypes. PMDS Is a rare form of male 46xy disorder of sex development (DSD). Persistent mullein duct syndrome [PMDS] is a condition in which a biological male develops female sex organs specifically a uterus and fallopian tubes in addition to male sex organs. Doctors also sometimes call it "Persistent Oviduct syndrome".

PMDS is a rare form of male pseudo hermaphroditism characterized by the presence of the mullein duct structures in an otherwise phenotypically as well as genotypically normal male. The pathophysiology of PMDS is mostly explained by failure of synthesis or release of IPmullerian inhibiting substance (MIS) by immature sertoli cells, or the failure of end organs to respond to MIS. Clinically, the patient with PMDS present with non- palpable/ undescended testes or an inguinal hernia with a palpable testis with the hernica sac. Although imaging techniques may help to investigate the intersex abnormalities, mostly preoperative diagnosis of PMDS is practically impossible, as the external male genitalia appears to be normal. PMDS is a disorder of sexual development that affects males. Males

with this disorder have normal male reproductive organs through they also have a uterus and fallopian tubes, which are female reproductive organ. The uterus and fallopian tubes are derived from a structure called the Mullerian duct usually breaks down during early development in males, but it is retained in those with PMDS. Affected individuals have the normal chromosomes of a male (46xy) and normal external male gentalia. The first noted signs and symptoms in males with PMDS are usually undescended testes (cryptorchidism) or soft out pouching in the lower abdomen (inguinal hernias). The uterus and fallopian tubes are typically discovered when surgery is performed to treat these conditions. The testes and female reproductive organs can be located in unusual position in Persistent mullein duct syndrome. Occasionally, both testes are undescended (bilateral cryptorchidism) and the uterus is in the pelvis. More often, one testes has descended into the scrotum normally, and one has not sometimes, the descended testes pulls the fallopian tube and uterus into track through which it has descended. This creates a condition called hernia uteri inguinal, a form of inguinal hernia. PMDS is thought to result from the failure of synthesis or release of Mullerian inhibiting factor (MIF), the failure of end-organ to respond to MIF, or a defect in the timing of the release of MIF.

Formation of the urogenital sinus and male external genitalia requires in situ conversion of testosterone into dihydrotestosteron. PMDS patient develop both wolffian and mullein structures due to deficiency of MIF. The gene which is responsible for MIF is localized on the short arm of chromosome 19.

Causes of the persistent mulleinduct:

PMDS are categorized into three types, according to the position of the patients testes and uterus: i) The female type occurs in 60–70% of patients, in which the bilateral testes and epididymis are connected to the fallopian tubes in the abdomen, the bilateral testes are in analogue positions to the ovaries and their inguinal sacs remain empty; ii) the Uteri Inguinal type occurs in 20–30% of patients that exhibit a testis in the hernia sac, or a scrotal testis with a contra lateral testis located in the abdomen; and iii) the transverse testicular ectopia makes up the smallest group as it is only classified in ~10% of patients where the two testes are present in the same hernia sac along with the uterus and uterine tubes .The exact cause of PMDS is not known; however, it is thought to result from the defects of the synthesis or release of MIF or from the MIF receptor defect. The MIF gene has been localized to the short arm of Chromosome 19. MIF, released by the sertolli cells of the fetal testis from seven weeks' gestation onwards, is responsible for the regression of the Müllerian duct in the male fetus. A defect of the MIF gene leads to the persistence of the uterus and the fallopian tube in the male. It is likely that these remnant Müllerian structures produce cryptorchidism by hindering the normal testicular descent mechanism.

Two clinical variants of PMDS are encountered. The more common variant of PMDS is characterized by unilateral cryptorchidism and contralateral inguinal hernia. When in such patients the uterus is present in the hernial sac, the condition is referred to as hernia uteri inguinal. In transverse testicular ectopia, both the testes are located on one inguinal side and the opposite inguinal canal and scrotums are empty. In the rarer variant, patients may present with bilateral cryptorchidism where the uterus is in the pelvis and both the testes are embedded in the broad ligament. In the PMDS-affected individuals, the testis is usually histologically normal, apart from the spectrum of lesions due to longstanding cryptorchidism. Like other undescended testes, the gonads of these patients are at increased risk of malignant transformation. There have been case reports of embryonal carcinoma, seminoma, yolk sac tumor and teratoma in patients of PMDS. The overall incidence of malignant change has been found to be 15%. In our case, neither testicular tissue nor any malignancy was identified in the multiple sections from the mass removed.

Most people with Persistent mullein duct syndrome have mutations in the AMH gene provides instructions for making a protein called "Anti Mullerian hormone"(AMH). The AMHR2 gene provides instructions for making a protein called AMH receptor type2. The AMH protein and AMH receptor type2 protein are involved in male sex differentiation. Al fetus develop the Mullerian

duct, the precursor of female reproductive organs. During development of male fetus, these two proteins work together to induce breakdown (regression) of the Mullerian duct. Mutations in the AMH and AMHR2 genes lead to non-functional proteins that cannot signal for regression of the Mullerian duct. As a result of these mutations, the Mullerian duct persists and goes on to form a uterus and fallopian tubes.

Approximately 45% of cases of PMDS are caused mutations in the AMH gene are called"persistent mullein duct syndrome type 2". In remaining 15% of cases, no mutations in the AMH and AMHR2 genes have been identified, and the genes involved in causing the conditions are unknown. While in the womb, babies (malesor female) grow a mullein duct. Normally, as girls grow in the womb, this duct development into female sex organs. In boys, on the other hand, their genes cause the release of proteins that breakdown this duct so that it doesn't develop further. As a result, the Mullerian duct continues to develop into a uterus and fallopian tubes that serve no useful biological purpose and may cause health problems. AMH: Anti-Müllerian hormone (AMH), also known as Müllerian inhibiting substance, a member of the transforming growth factor-β (TGF-β) superfamily produced by Sertoli cells of the fetal testis from 7 weeks' gestation, is responsible for the regression of Müllerian ducts in male fetuses, the first step of male sex differentiation of the genital tract. AMH signals through receptors located in the mesenchyme of the fetal Müllerian ducts. Testosterone, produced by Leydig cells starting at 9 weeks' gestation, is responsible for the virilization of the male internal and external genital tract, i.e. maintenance and male differentiation of the Wolffian ducts and morphogenesis of a male external genital phenotype. It follows that genetic abnormalities of the AMH or its receptors will interfere only with the process of Müllerian duct regression, while differentiation of male accessory organs and external virilization will proceed normally under the influence of testosterone. Such is the definition of the persistent Müllerian duct syndrome (PMDS), the subject of this review. The other cause of persistence of Müllerian ducts, testicular dysgenesis, usually affects both Sertoli and Leydig cells: persistence of Müllerian derivatives is then associated with external genital ambiguity.

AMHR2: The AMHR2 gene provides instructions for making the anti- mullein hormone (AMH) receptor type2, which is involved in male sex differentiation. The AMH receptor type2 is found on the surface of mulleinduct cells. The Mullerian duct, found in both male and female fetuses, is the precursor to the female reproductive organs. During development of male fetuses, cell of the testes release a protein called the AMH protein. The AMH protein attaches (binds) to the AMH receptor type2, which signals self-destruction (apoptosis) of the Mullerian duct cells. As a result, the Mullerian duct breaks down (regresses) in males. In females, who do not produce the AMH protein during fetal development, the Mullerian duct becomes the uterus and fallopian tubes.

Symptoms: One of the first sign of Persistent mullein duct syndrome in boys (girls don't get it) is that one or both testis don't descend. It's also common to see bulges that poke through the muscles in the groin area (inguinal hernia).

In children, doctors typically discover Persistent mullein duct syndrome by chance during surgery to correct one or both issues. The surgeon notices a uterus and fallopian tubes (and sometime other female structures) in unusual position in the pelvic area. If one testicles have descended, it may pull some of the fallopian tubes and uterus with it.

Other possible symptoms of Persistent mullein duct syndrome include:

- Abnormal growth patterns in internal male genitalia.
- The signs or appearance of external female genitalia in addition to normal male genitalia (malepseudo hermaphroditism)
- Abnormal hormone levels.
- Men with Persistent mullein duct syndrome may also notice blood in the semen. They also have higher risk of testicular cancer.

- More specifically,undescended testes left untreated may Breakdown and developed cancer.

Diagnosis:

The diagnosis of the Persistent mullein duct syndrome is made radiologically and that the detection of Mullerian inhibiting factor is mandatory. The position of the uterus and fallopian tubes different in all patients;

One was in the scrotal sac, another in the abdomen and the third in the left inguinal canal. Between 1993- 2002 we used both diagnostic and operative laparoscopy in the management of 5 cases of PMDS. 2 siblings from two different families accounted for four of the cases. They presented with cryptorchidism and inguninal hernias. The diagnosis was established during diagnostic laparoscopy. The impalpable testes were on the left in three, on the right in one and bilateral in one. A diagnosis of PMDS was made on histological evaluation subsequent to abdominal orchiectomy. Ten patients had a laparoscopic excision of their Müllerian structures while the remaining nine patients had their Müllerian structures left in place. No malignant changes were found in the excised Müllerian tissues. Of the 37 gonadal biopsies taken, 31 (84%) indicated normal testes.

Three adults with PMDS and an associated testicular malignancy were evaluated using physical examination, imaging, measurement of tumor markers, surgical exploration and chromosome analysis. The PMDS was diagnosed on laparoscopy; initially biopsy of these remnants and gonads was done followed by excision of remnants by laparotomy approach. Biopsies taken from gonads in each patient revealed testicular tissue with variable degree of immaturity and dysplasia. The biopsy of mullein remnants did not reveal any malignancy. All patients were genotypically male.

Treatment: The surgical management of Persistent mullein duct syndrome and extracted information regarding the etiology, pathogenesis, and treatmen of this disorder.

We specifically assessed the risk of retained mullein structure versus surgical excision of the infantile uterus and fallopian tubes. Surgically excision of the infantile uterus and fallopian tubes risk damage to Vasa differentia and the deferential blood supply to testis. Surgical excision of Persistent mullein duct structure may result in ischemic and or traumatic damage to the Vas differentia and testes. Optimal surgical management is orchiopexy leaving the uterus and fallopian tubes in situ. Orchiectomy is indicated for testes that cannot be mobilized to apalpable location. Persistent mullein duct syndrome finding either during orchiopexy or during routine inguninal hernia repair in male patients. often the surgeon would face a dilemma about what is to be done with the remnants of mullein duct i.e., fallopian tubes and uterus and proximal vagina. Patients with Persistent mullein duct syndrome, we have removed the mucosa of the retained Mullerian structures, without compromising the integrity and vascularity of the vas deference, thus reducing the chance of malignancy.

Persistent mullein duct syndrome can be managed as single stage procedure however two stage procedure including gonadal biopsies in first stage procedure including gondal biopsies in first stage followed by mullein remnants excision and orchidopexy in the second stage can be opted if there is doubt about gonads and genotype.

History of PMDS:

PMDS: A 24 years' experience the medical record of 27 cases of Persistent mullein duct syndrome (PMDS) operated in three teaching hospitals more than a period of 24 years is retrieved and analysed for demography, clinical presentation, investigations, and treatment.

There were a total of twenty-seven male children with PMDS. The age was ranged between 3 months and 19 years. Ten patients presented with bilateral UDT and unilateral inguinal hernia (4 left and 2 right sided inguinal hernia). And 8 patients presented with right inguninal hernia and left sided

UDT. Eight of twenty-seven patient s showed familial trends i.e. four pairs of brothers had PMDS in our series. In 21 patients, the diagnosis was made incidentally while operating for UDT and inguninal hernia. At operation 5 patients had female type PMDS. In 22 patients had male type PMDS. In 6 patient s (male type), the PMDS was associated with testicular ectopia.

To determine the etiology of this syndrome, we studied the expression of anti- Mullerian hormone (AMH) in six boys, including three brothers, with the persistent mullein duct syndrome. All except one presented with an inguinal her containing the Mullerian derivatives, and in two boys the hernical sac contained the contralateral testies. AMH was normally expressed in the testicular tissue of two patients, as shown by bioassay of anti-Mullerian activity and immunocytochemistry. The testicular tissue of the other patients had no detectable bioactive or immunoreactive AMH, yet they expressed AMH mRNA with a normal transcription initiation site and in the amount expected for their age. These result prove the heterogeneity of the PMDS and suggest that it may sometimes involve peripheral insensitivity to AMH. Approximately 200 cases of Persistent mullein duct syndrome have been reported over the last 50 years. To describe the presentation and management of eight patients with PMDs seen over a 10-years period at our center. 11cases with PMDS and malignancy of the Mullerian remnants were identified.

The hospital records of eight patients with PMDS were retrospectively reviewed between 2001 and 2011.

19 phenotypically male patients (aged 8 months to 27 years) presented with testicular maldescent. All of them had normal male external genitalia. 2 of them had a previous diagnosis of Persistent mullein structures. All patients were karyotyped, and had a hormonal profile, diagnostic laparoscopy, retrograde urethrocystogram, gonadal biopsies, and surgical management according to the findings. Diagnostic laparoscopy showed the presence of Persistent mullein structures in all 19 patients.

From June 2009 to August 2013, 19 phenotypically male patients (mean age 8.2 years, range 8 months to 27 years) presented with testicular maldescent. All of them had normal male external genitalia. Two patients had had a previous right inguinal orchidopexy; one of them was assessed using MRI (to visualize his left impalpable testis), and this detected a uterus. Another patient had surgery for a left inguinal hernia, during which a lump was excised and confirmed to be uterine tissue. All patients were karyotyped and had a hormonal profile (FSH, luteinizing hormone, LH, and testosterone), a diagnostic laparoscopy which confirmed the presence of Müllerian structures, a retrograde urethrocystogram, gonadal biopsies, and surgical management according to the findings. Three adult patients had semen analysed and tumor markers assessed before surgery, i.e., human chorionic gonadotrophin (HCG), α-fetoprotein (AFP), and lactate dehydrogenase (LDH).

All patients had a normal male karyotype (45XY). Ten patients had al laparoscopy excision of their mullein structures while the remaining 9 patients had their mullein structures left in place. No malignant changes were found in the excised Mullerian tissues of the 37gondak biopsies taken, 31 (84%) indicated normal testes. The current study reports the clinical data and results of the genetic analysis of a 17- month old male diagnosed with PMDs. A 67 years old man with clear cell adenocarcinoma of the remnant uterus in PMDs. He had normal penis and scrotum and there was also a vagina and uterus.

Case and reports:

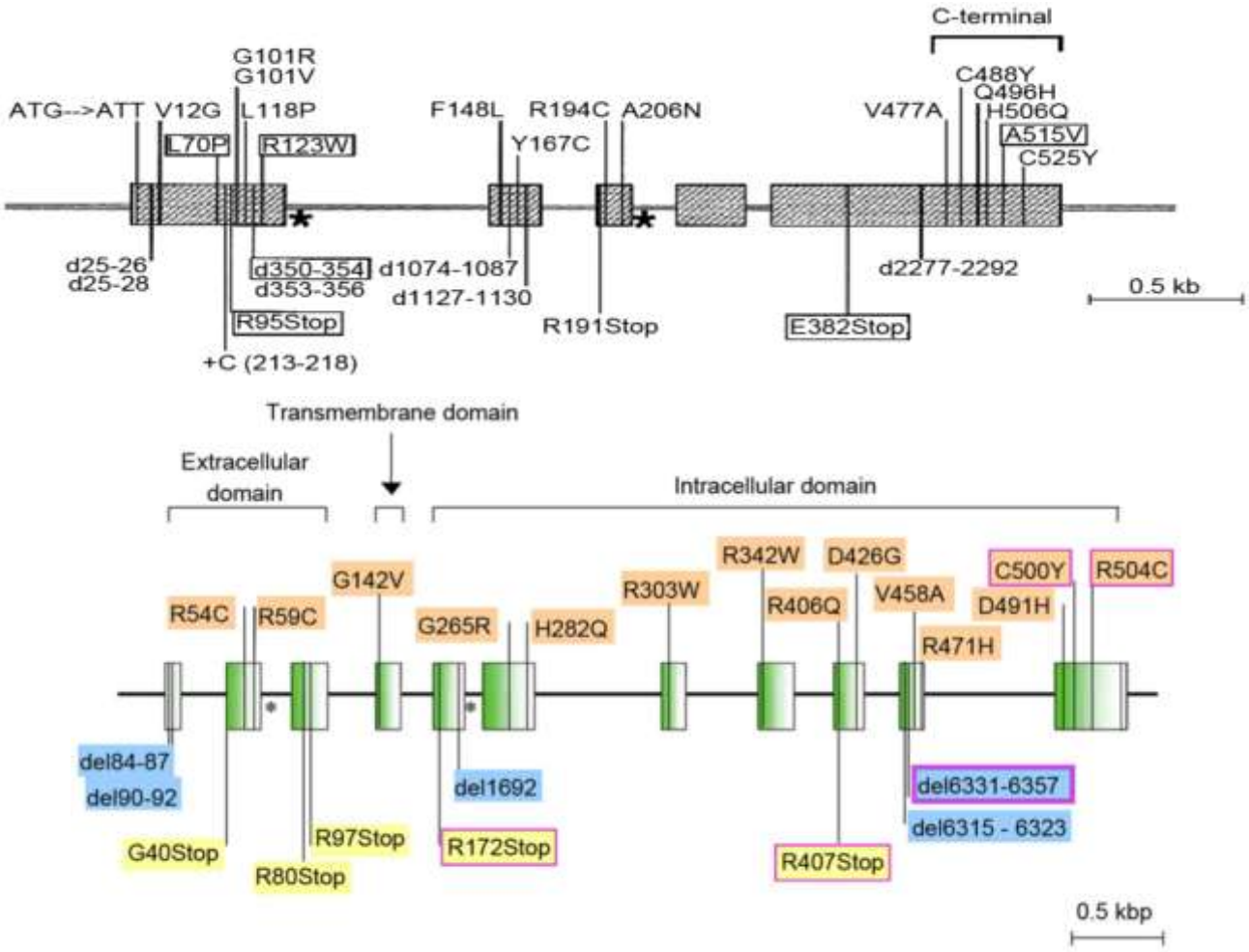
PMDS is a rare condition, with only about 150 cases being described in the literature. In transverse testicular ectopia, the ectopic testis may lie at internal ring, in the scrotum. Since its first description was made, more than 100 cases have been reported. Among all reported cases, the oldest and youngest patients were 77 years and 3 days old, respectively. A majority of cases which have been reported, belongs to pediatric and adolescent age groups. A 38 years old man was found to have a uterus, fallopian tubes, and a gonda in the left hernical sac during herniorrhaphy.

The patient developed a left scortal sac swelling postoperatively. On physical examination, the right scortal sac was empty. The left scortal sac appeared boggy. The patient was phenotypically male, with male pattern of external genitalia and secondary sexual characteristics. The past history included primary infertility.USG showed a well formed uterus situated alongside the bladder and extending into the inguninal canal. The endometrial cavity was distended with a small collection. An oval structure with uniform internal echoes, confirming to a testis, was noted at the level of the left deep inguninal ring. The prostate and seminal vesicles were well visualized and appeared normal. The left scortal sac showed a septate collection, consistent with a hematocele. No testis was identified within the left scortal sac. The right scortal sac was empty. The right testis could not identifying an ectopic location. MRI confirmed the USG findings. It showed a well formed uterus and fallopian tubes alongside the urinary bladder and extending into the left inguinal region. A small collection was noted in the uterine cavity. An oval structure measuring 3.7×1.9cm, with morphology and signal intensity consistent with a testis was detected at level of the deep inguinal ring. A vaginal like structure was seen extending from the uterus towards the left seminal vesicle no structure confirming to the testis was seen in the scortal sac or in any ectopic location. A 73-year-old man presented with complaints of progressive pain in the left lower quadrant associated with constipation. These complaints were reduced by having the patient lie on his right side. There was no history of micturation problems.At physical examination, a palpable mass was detected in the left lower abdomen, together with an ipsilateral reducible inguinal hernia. No testis was found in the right side of the scrotum. Cystoscopy showed no abnormalities. Laboratory studies revealed minimal renal function abnormalities without hematuria and a low level of testosterone (3.1 nmol/L [0.89 ng/mL]). A chromosome analysis revealed a normal male karyotype of 46 XY.

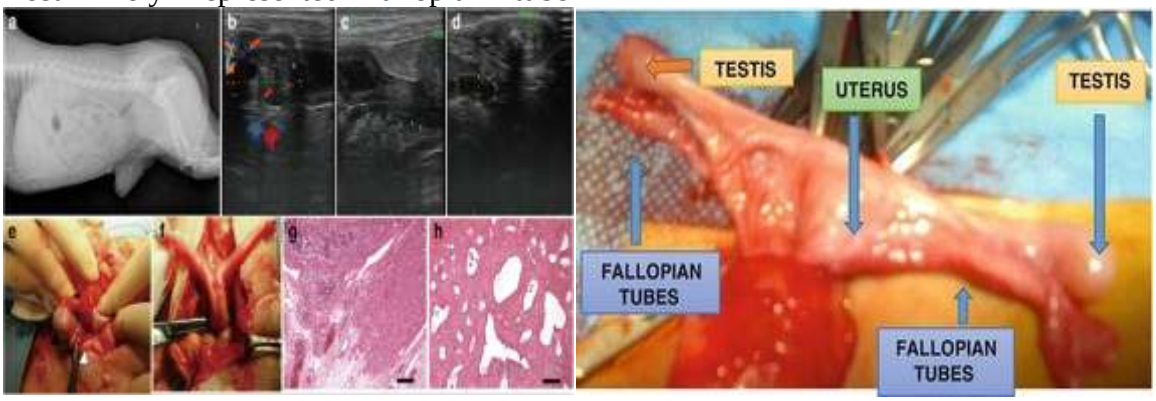
Past medical history included repair of a left inguinal hernia and left orchiopexy (performed in 1945), appendectomy, right nephrectomy for pyonephrosis with nephrolithiasis, and primary infertility. Ultrasonography (US) of the genitalia showed multiple cysts located in the scrotum and extending into the left inguinal canal. A solid mass with a central cystic component was identified in the lower abdomen. Magnetic resonance (MR) imaging of the abdomen showed a lower abdominal mass at the ventral side of the urinary bladder. The wall of the mass was thickened and had low signal intensity at T1-weighted imaging and mixed intermediate and high signal intensity at T2-weighted imaging. Smooth, well-defined lesions with low signal intensity were seen within the wall with both sequences. A portion of the mass was seen to herniate into the left inguinal canal. The mass was filled with fluid that had high signal intensity at both T1- and T2-weighted MR imaging, a finding that was compatible with blood products. A structure with low signal intensity at T1-weighted MR imaging and high signal intensity at T2-weighted imaging was identified on the left side of the scrotum, and a structure with low signal intensity at T1-weighted imaging and mixed intermediate and high signal intensity at T2-weighted imaging was detected in the right side of the abdomen. These two structures represented the testes. Exploratory laparotomy revealed an enlarged uterus. The left side of the uterus was partly situated in the left inguinal canal. The left adnexa were found in the scrotum. At surgery, brown fluid was found in the vagina. The uterus, vagina, and adnexa were removed en bloc.

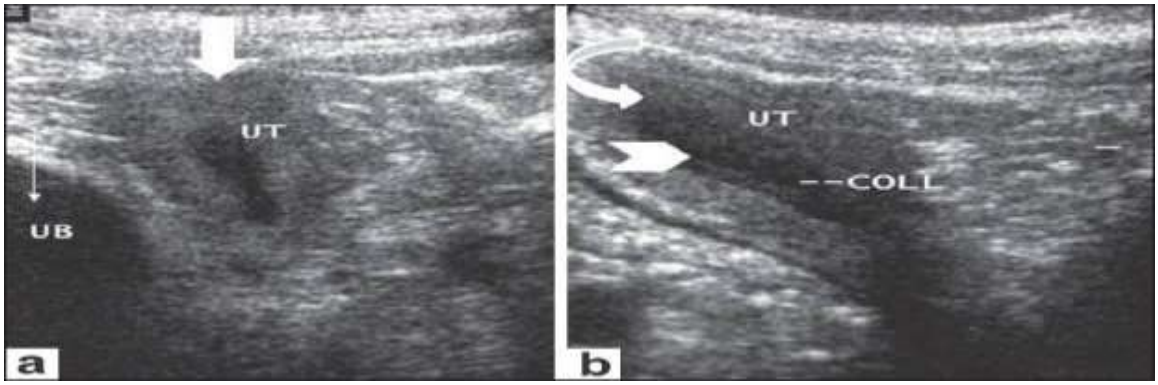
Histologic examination showed the uterus to be composed of a muscle layer lined with a single layer of tubal epithelium without atypia, with foci of underlying endometrial stroma. Multiple leiomyomas without signs of malignancy were observed in the muscle wall. Although the gonad in the right adnexa was thought to be an ovary, at histologic analysis it proved to be a testis with complete atrophy and containing fibrosis and calcifications.

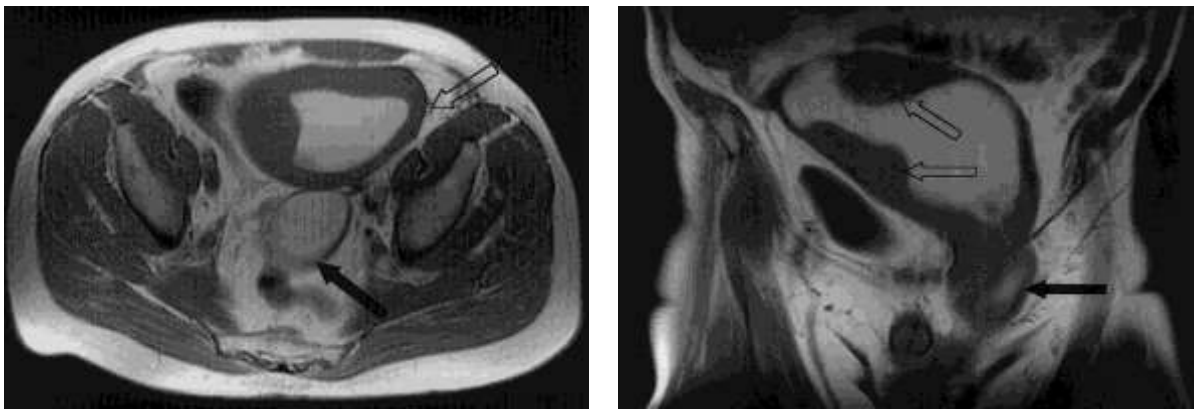
The left testicle demonstrated atrophy and Leydig cell hyperplasia, with no active spermatogenesis. A rete testis with a normal relationship to the testicle was present on both sides. No ovarian tissue was found. Structures lined with tubal epithelium were detected on both sides of the uterus and remnants. Adjacent to the left adnexa, simple cystic structures lined with a single layer of tubal epithelium without atypia were present.

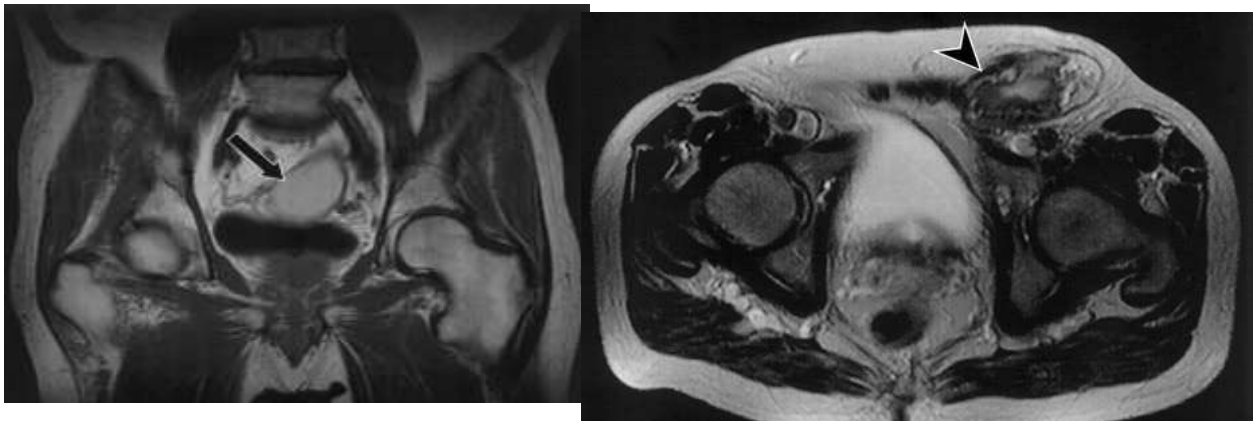


most likely represented fallopian tube









CONCLUSION:

The incidence and prevalence of PMDS are not well estimated. Müllerian structures should be removed whenever possible to avoid the risk of malignant transformation. The early diagnosis of PMDS makes possible the excision of Müllerian structures and a primary orchidopexy.

- The principle aim of orchidopexy with simultaneous laparoscopic removal of the Müllerian structures can be accomplished with minimal surgical trauma and the benefit of no malignancy risk in the future.
- Surgeons should consider excision of the Müllerian remnants where possible.

PMDS can be managed as single stage procedure however two stage procedure including gonadal biopsies in first stage followed by mullein remnants excision and orchidopexy in the second stage can be opted if there is doubt about gonads and

Surgical excision of persistent mullein duct structures may result in ischemic and/or traumatic damage to the vasa differentia and testes. Optimal surgical management is orchiopexy leaving the uterus and fallopian tubes in situ.

Management of this syndrome is difficult because of the limited number of cases. If the

diagnosis can be made before surgery, karyotyping can be useful to decide on orchiopexy or orchioectomy. In suspected cases, laparoscopy and ultrasonographic evaluation of all crytorchidic cases may be helpful for diagnosing this condition before surgery. All patients with this syndrome have a male phenotype; therefore, it is essential to preserve secondary sex characteristic.

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