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Robot-assisted hysterectomy in a 41-year-old male: A rare case report

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ABSTRACT

Persistent Müllerian Duct Syndrome (PMDS) is regarded as a rare genetic disorder influencing internal sexual male development. PMDS is commonly diagnosed incidentally either during any pelvic surgery or examination of undescended testis. Currently, we have presented a case focusing on the phenotype individual who was presented to our institute with primary infertility as well as bilateral undescended testis, who underwent Robotic-assisted hysterectomy, left orchidectomy, and right orchiopexy.

Introduction

PMDS is a rare genetic male sexual development disorder in which the derivatives of the Müllerian ducts (uterus, fallopian tube, and the upper two-third vaginal area) is persistent in the phenotype normal karyotype male (XY′46).[1]

PMDS takes place due to insensitivity or deficiency of the Müllerian inhibiting factor (MIF) which is excreted from the testis embryonically (Sertoli cells). However, the testosterone secretion from the testis of the Sertoli and Leydig cells is not influenced hence there is no maldevelopment of the Wolffian duct resulting in normal male genitalia appearance.[2] Three anatomical conditions exist for PMDS which are Type 1, Type 2, and Type 3. Type 1 is the commonly occurring which is detected by the testis presence in an anatomic position of ovaries in addition to the intra-abdominal presence of the Müllerian duct derivatives. Type 3 is the condition where one of the testes is present in the scrotum or inguinal canal and is thus regarded as the Hernia Uteri Inguinalis. Lastly, Type 3 is detected by the presence of both of the testes within the hemiscrotum (transverse testicular ectopia).[3]

Identification of PMDS is mostly incidental which takes place during examining the underlying cause of the cryptorchidism or hernia repair surgical process. An. ultrasound, magnetic resonance imaging (MRI) scanning, or computed tomography (CT) where MRI is most effective in the detection of the pelvic structure for confirming the imaging modality.[4]

Case report

A 41 years old adult was referred to our institute with the case of primary infertility for assessment. Patient history indicated that he has been married for 17 years with unremarkable sexual activity. Upon physical assessment, the patient had normal appearing genitalia, secondary sexual features, and male phenotype. However, the scrotum was not completely developed and empty. Laboratory examinations revealed a high FSH and LH range while the semen analysis indicated azoospermia. MRI was performed which confirmed the presence of rudimentary uterus with the cervix and a uterine body. Counselling was offered to the patient regarding surgical implications for the removal of the uterus either through orchiopexy or probable orchiectomy. In line with respecting the patient's wishes disproving to the lifelong hormonal therapy, the patient agreed for orchiopexy. He underwent diagnostic exploration and was found to have bilateral testis with normal vasculature as in normal abdominal testis and an underdeveloped uterus along with bilateral adnexa (Fig. 1). The patient proceeded for

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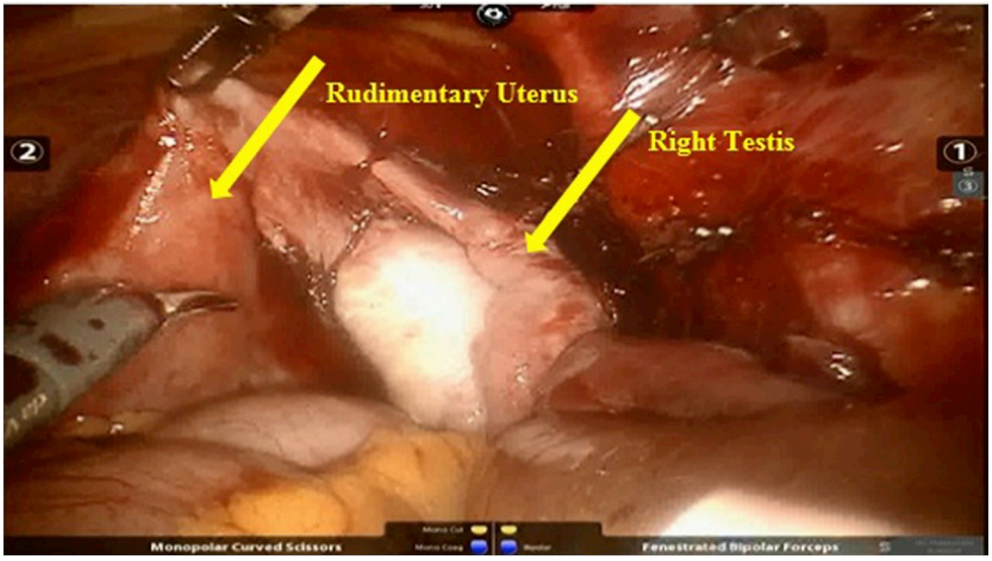

Fig. 1. Intraoperatrive finding showing rudimentary uterus with fallopian tube and right testis.

orchiopexy and robotic hysterectomy (Fig. 2). The patient was placed in a lithotomy- Trendelenburg surgical position; uterus was initially identified followed by cord and gonads. Through ligating round ligaments, mobility of the uterus was ensured. The uterine vessels were ligated for removing the uterus. Vagina was also resected after the identification of the cervix. Bilateral cutting of the fallopian tubes was performed while the gonads were brought down. However, due to shortage of the left spermatic cord as well as the hard left testis on palpitation as opposed to the right testis, it was decided to perform orchiectomy in addition to right orchiopexy. No consideration for staged orchiopexy. The total duration of the operation was 190 minutes and the total loss of blood recorded was less than 100 ml. The patient was discharged on 2nd post-op day. The left testis specimen demonstrated Sertoli cells only with absolutely no evidence of malignancy where the uterus was shown as inactive endometrium (Fig. 3). The patient planned for regular follow up with ultrasound to check the viability of the preserved testis.

Discussion

PMDS is identified as a rare genetic condition leading to primary male infertility with three likely probabilities; external masculine genitalia with a developed internal uterus, external feminine genitalia, or external genitalia of equivocal sexualities. The foremost type is the most commonly occurring which is also present in our case.[5] The diagnosis could be identified during the later stages of adulthood. The contributing factors of this condition include the disease rarity, presentation vagueness, normal lifestyle of the patients, and the disease course. Also, there are no visible characteristic complaints or symptom for the physician to examine and identify. Patients either present with hernia repair or undescended testes. In addition, other complaints usually include vague pain in the pelvic or abdominal region while primary infertility is the cause of identification in our case.[3] Similarly, diagnosis is also incidental and subjected to surgery. Missed post-operative cases of PMDS could be evaluated through imaging and genetic studies e.g. anti-Muellerian hormone type-2 receptor sequencing (AMHR 2). Ultrasound is majorly used during the initial phases for visualizing the uterus

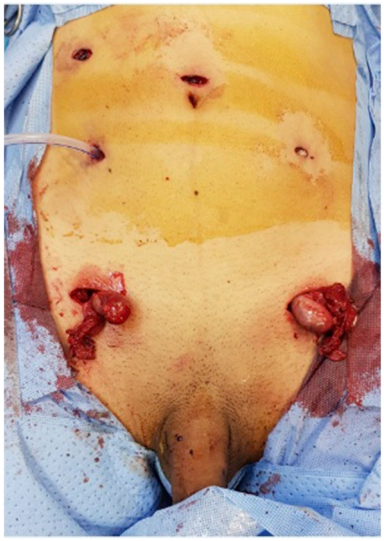

Fig. 2. Intraoperative finding: showing bilateral descending of both testis.

wall and the undescended testes. MRI is confirmatory with the potential to recognize complication PMDS structures. Up to 18% of testicular malignant transformation has been reported.[4] Malignancies usually occur in epithelial and sarcomatous (mesenchymal) transformations in the Müllerian tumor. For the prevention of malignancy, surgical intervention is used. Nevertheless, surgery could injure the vas and hence this process is controversial.[2] Abundantly, surgery is preferred to two prime causes which include prevention of malignancy and fertility preservation. The approach to surgery should be discussed by the surgical team. In the present case, the use of Robot-assisted surgery for PMDS was reported.

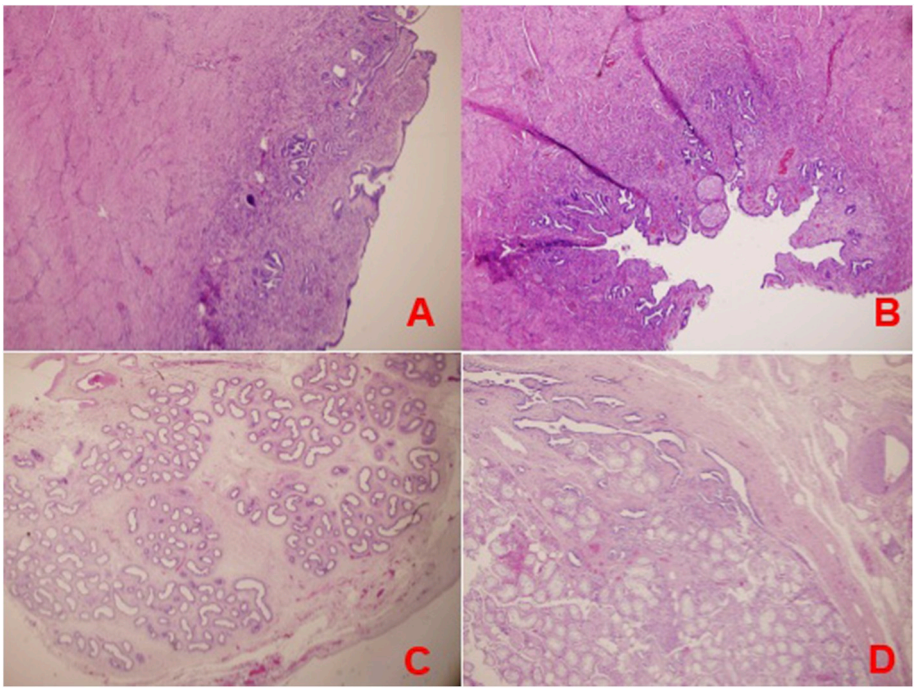

Fig. 3. Histopathology: A. Unremarkable cervix with no well-formed ectocervix. B. Inactive endometrium. C. Unremarkable epididymis. D. All seminiferous tubules show Sertoli cells only, interstitial Leyding cell are present, unremarkable Rete testis.

Conclusion

PMDS is a rare genetic disease whose early identification and treatment are critical for the preservation of fertility and malignancy prevention are some of the cases. Robot-assisted surgery is hence feasible in such case.

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