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CYSTOSCOPY AND MUCOSECTOMY: ESSENTIALS IN THE MANAGEMENT OF PERSISTENT MÜLLERIAN DUCT SYNDROME WITH TRANSVERSE TESTICULAR ECTOPIA

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Summary.- **OBJECTIVES:** The concurrence of Persistent Müllerian Duct Syndrome and transverse testicular ectopia is rare. The risk of damage to the vas deferens and the deferential blood supply hinders some surgeons from complete excision of potentially malignant Müllerian duct remnants.

METHODS: We present a unique surgical technique of Persistent Müllerian Duct Syndrome in a patient with right inguinal hernia accompanying transverse testicular ectopia.

RESULTS: During exploration, both testes were detected in the right inguinal canal. When the hernia sac was

opened, a primitive uterus and fallopian tubes without fimbria were identified confirming Persistent Müllerian Duct Syndrome. A 4 Fr catheter was placed into the os of the Müllerian duct remnants via the verumontanum orifice, and then a urethral catheter was placed. The full-thickness excision of proximal Müllerian duct remnants were performed. The distal part of Müllerian duct remnants was layed open and only mucosa was excised for preserving vas deferens. Resection was completed just above its junction with the urethra with the aid of 4 Fr catheter marked at centimeter intervals and the cuff was oversewn. High ligation for right inguinal hernia and bilateral orchidopexy were performed.

CONCLUSIONS: Removal of Müllerian duct remnants is advised in order to reduce the jeopardy of malignancy, urinary tract infections, stones and hematuria. On the other hand, excision down to urethra which can compromise the integrity and vascularity of the vas deferens is difficult, even in experienced surgical hands. Complete excision of these structures by mucosectomy of the distal part of remnant which lay closed to vas deferens is a safe and effective method. Cystoscopy assistance and placement of a catheter into MDRs were essential for the complete excision of this mucosa. To the best of our knowledge, cystoscopy assisted mucosectomy in Persistent Müllerian Duct Syndrome has not been reported previously.

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Resumen.- *OBJETIVOS:* La presencia de síndrome de persistencia de los conductos müllerianos y ectopia testicular transversa es raro. El riesgo de dañar el conducto deferente y la vascularización diversa hace que muchos cirujanos no realicen una extirpación completa de los conductos müllerianos remanentes con riesgo de malignización.

MÉTODOS: Presentamos una técnica quirúrgica única para la resección completa de los conductos müllerianos remanentes en pacientes con hernia inguinal derecha acompañada de ectopia testicular transversa.

RESULTADOS: Durante la exploración física se detectaron ambos testículos en el canal inguinal derecho. Cuando abrimos el saco herniario, se observó un útero primitivo con trompas de falopio sin fimbrias confirmando el síndrome de persistencia de los conductos müllerianos. Se colocó un catéter 4 Fr en la punta del remanente mülleriano a través del orificio del verumontanum y a posteriori se colocó una sonda uretral.

CONCLUSIONES: La extirpación del remanente del conducto mülleriano esta indicada para evitar la malignización, infecciones urinarias, litiasis y hematuria. Por otro lado, la extirpación hasta la uretra puede comprometer la vascularización y integridad del conducto deferente, siendo dificultosa hasta en manos expertas. La extirpación completa de las estructuras con mucosectomía de la parte distal del remanente es segura y eficaz. La ayuda de la cistoscopia y colocación de un catéter en el remanente son muy importantes para la resección completa. Esta es la primera descripción de mucosectomía asistida por cistoscopia en un síndrome de persistencia del remanente mülleriano hasta la fecha.

Palabras clave: *Síndrome persistencia conducto mülleriano. Ectopia testicular transversa. Cistoscopia. Mucosectomía.*

INTRODUCTION

Persistent Müllerian Duct Syndrome (PMDS) is a rare condition of 46XY male disorders of sex development, which occurs as a result of Müllerian inhibiting factor or Müllerian inhibiting factor receptor deficiency. Persistent Müllerian Duct Syndrome is a complex and anatomically variable disorder, which is observed in normally virilized 46XY male patients with the presence of Müllerian derivatives such as fallopian tubes, uterus and upper part of vagina along with normally developed male reproductive organs (1). Transverse testicular ectopia (TTE) is a rare form of testicular ectopia in which both testes are located

in one inguinal side. About 260 cases of TTE have been reported till date (2). The concurrence of PMDS and TTE is extremely rare (2). The risk of damage to the vas deferens hinders some surgeons from complete excision of potentially malignant Müllerian duct remnants (MDRs).

We here present a case with TTE associated with PMDS to emphasise the MDRs excision technique which is essential for preserving vas deferens and the deferential blood supply to the testes.

SURGICAL TECHNIQUE

A 1-year-old boy was admitted with a bulge in his right groin. Physical examination was consistent with right incarcerated inguinal hernia and also bilateral empty hemiscrotum was observed. After the successful manual reduction of hernia, inguinal oedema and erythema in association with discomfort continued to appear. Doppler ultrasonography was performed in order to exclude testicular torsion. It demonstrated that both testes were of a similar size, with normal vascularity and echogenicity in the right inguinal canal. Right inguinal exploration was planned with the prediagnosis of right inguinal hernia accompanying TTE. Written consent form was obtained from parent of the child. During exploration, both testes with their own blood supply, epididymis and spermatic cord were detected in the right inguinal canal. There were microcysts in the polar region of testes. Therefore, bilateral testis biopsy was taken. When the hernia sac was opened, a primitive uterus and fallopian tubes without fimbria were identified confirming PMDS. Cystoscopy was performed in order to evaluate the communication between the MDRs and urethra. A 4 Fr catheter was placed into the os of the MDRs via the verumontanum orifice, and then a urethral catheter was placed. The MDRs were dissected with particular attention to avoid injury to the bladder neck, urethra, rectum, ureters and vas deferens (Figure 1). The distal part of MDRs was layed open and only mucosa was excised for preserving vas deferens. Then, resection was completed just above its junction with the urethra with the aid of 4 Fr catheter marked at centimeter intervals and the cuff was oversewn (Figure 2). High ligation for right inguinal hernia and bilateral orchidopexy were performed. The left testis was placed in the left hemiscrotum through a transseptal approach. No complications occurred in the postoperative period. Histopathological examination confirmed the diagnosis of PMDS (Figure 3). The gonadal biopsies showed normal prepubertal testicular tissues. Karyotype analysis was reported to be 46XY and sex-determining region of Y (SRY)(+).

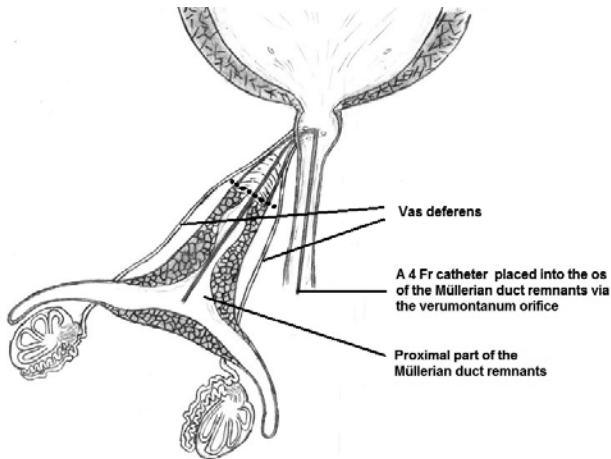


Figure 1. Coronal section of Müllerian duct remnants and right transverse testicular ectopia. Proximal part of the Müllerian duct remnants were dissected and excised.

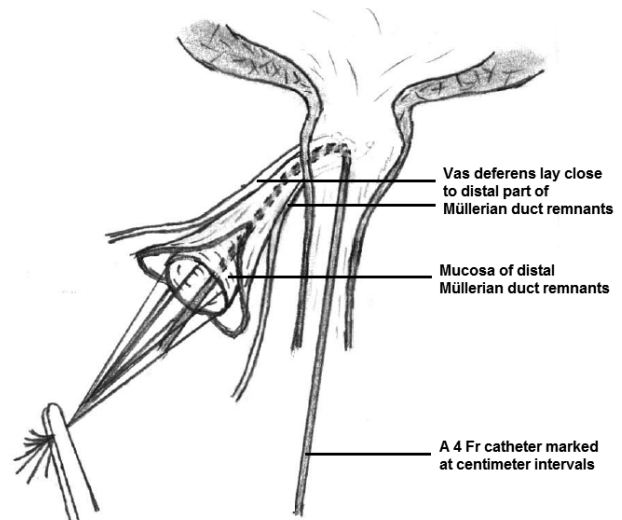


Figure 2. Only mucosectomy was performed in the distal part of Müllerian duct remnants for preserving vas deferens. Resection was completed just above its junction with the urethra with the aid of 4 Fr catheter marked at centimeter intervals.

DISCUSSION

The incidence of PMDS has not been accurately determined. The estimated number of reported cases of PMDS has been approximately 160 till date (1). There are 3 clinical presentations of PMDS:

- A. Unilateral cryptorchidism with contralateral inguinal hernia.
- B. Bilateral cryptorchidism in which testes are localized in the normal position of ovaries.
- C. Both testes are found in one hernia sac (TTE) (1).

The phenotype of our index case was consistent with group C. PMDS associated with TTE is extremely rare and, to our knowledge there are only 59 reported cases in the literature (2).

Management of PMDS with TTE remains controversial. It is often discovered during repair for inguinal hernia or cryptorchidism. Given the fact that most surgeons may not see any patient with the association of PMDS and TTE during their surgical career, it is important to be aware of this rare condition (3). When PMDS is found at surgery for TTE, various surgical techniques can be performed with the aim of prevention infertility and malignancy arising from testes or MDRs. The overall incidence of malignant transformation in these testes is 5-18%, which is similar to the rate of malignancy in abdominal testes in a patient without PMDS. There have been reports of embryonal

carcinoma, seminoma, yolk sac tumor and teratoma in the testes of patients with PMDS (4). Since a full literature review was not performed formerly, some surgeons believed that malignancy was not developed

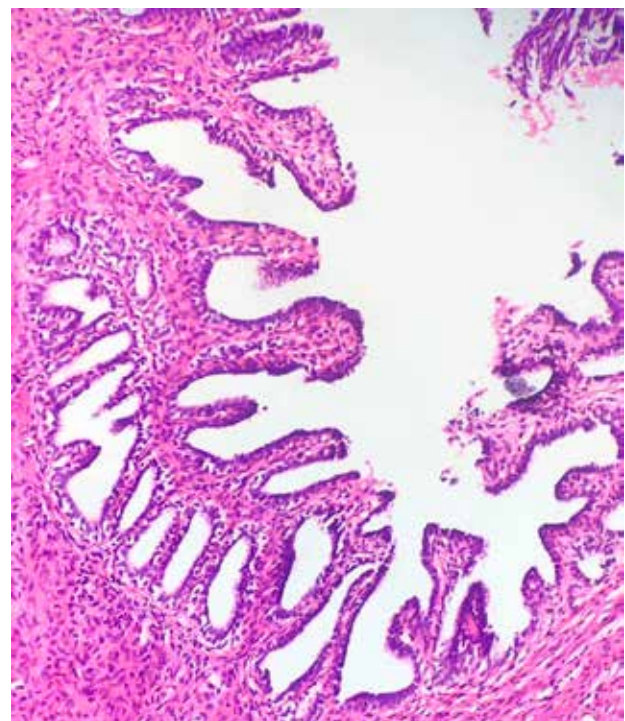


Figure 3. Histopathological appearance of Müllerian duct remnants. Micropapillary wall projections and microtubular spaces which were covered by one layer of tall columnar epithelium is apparent (H&E; x20).

from MDRs. Therefore, they did not recommend the removal of MDRs due to the difficulty of dissection (2,5,6). The vas deferens is in close proximity with the walls of the Müllerian derivatives. It can be incorporated into the wall of the vagina, as well. The surgeon should bear in mind that excision of these derivatives might damage to the testicular blood supply and vas deferens. Splitting of the Müllerian structures in the midline or microvascular autotransplantation are suggested when these derivatives prevented testicular descent (7). However, Farikullah et al performed a thorough search of the literature over the last 40 years and they reported 11 cases of malignancies arising not from the testes but from the retained MDRs, including squamous cell carcinoma, papillary cystadenocarcinoma, clear cell carcinoma, adenocarcinoma, adenosarcoma and clear cell adenocarcinoma. They estimated the malignant transformation of PMDS between 3.1% and 8.4% (8). Furthermore, the MDRs can cause urination discomfort, stones, urinary tract infections or periodic hematuria in the event of estrogen excess (9,10). Although the priority is orchidopexy to preserve fertility and to prevent testicular malignancy in the management of PMDS associated with TTE, the recent report of malignant degeneration of the 11 MDRs led us to remove these remnants in our case (8). We have excised the MDRs, which were located away from the vas deferens. Considering the reported malignancies of MDRs arising from its mucosa, the parts which lay close to the vas deferens were opened and the mucosectomy was performed in our patient. Then, resection was completed just above its junction with the urethra. Cystoscopy assistance and placement of a 4 Fr catheter marked at centimeter intervals were essential for the complete excision of mucosa in our patient. To the best of our knowledge, cystoscopy assisted mucosectomy in PMDS has not been reported previously.

Given the long-term follow-up with magnetic resonance imaging is insistently recommended when MDRs have been preserved, cystoscopy assisted mucosectomy saved our patient from this imaging all through his life. We have been made the follow-up of testes due to the risk of infertility and testicular malignancy by using only physical examination.

Successful laparoscopic removal of MDRs has been reported (8,11). Although laparoscopy provides excellent magnified operative view, we did not prefer laparoscopy for two reasons: First, both testes and MDRs in a hernia sac were detected not intraabdominally, but in the right inguinal canal in our case, Secondly, mucosectomy in the distal part of MDRs may be difficult laparoscopically.

CONCLUSION

Removal of MDRs is advised in order to reduce the jeopardy of malignancy, urinary tract infections, stones and hematuria. On the other hand, excision down to urethra which can compromise the integrity and vascularity of the vas deferens is difficult, even in experienced surgical hands. Complete excision of these structures by mucosectomy of the distal part of remnant which lay closed to vas deferens is a safe and effective method. Cystoscopy assistance and placement of a catheter into MDRs were essential for the complete excision of this mucosa.

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