

Persistent Mullerian Duct Syndrome with Transverse Testicular Ectopia

Rare Presentation of Inguinal Hernia

Ashish Gupta,¹ Nilanjan Panda,¹ Makhan Lal Saha,² Shuvro Ganguly,² Samik Kumar Bandyopadhyay,¹ Ruchira Das³

¹ Department of Surgery, R.G KAR Medical College, Kolkata, India

² Department of Surgery, IPGIMER and SSKM, Kolkata, India

³ Department of Surgery, B.S Medical College, Bankura, India

Corresponding Author:

Nilanjan Panda, MS, MRCS Ed, DNB
318 b, CIT Road, Scheme 6M,
Kankurgachi, Kolkata 700054, West
Bengal, India

Tel: +91 974 877 4942
E-mail: drnilanjanpanda2002@
yahoo.co.in

Received April 2011
Accepted July 2011

Keywords: persistent Mullerian duct syndrome, testis, male pseudohermaphroditism

INTRODUCTION

Persistent Mullerian duct syndrome (PMDS) is a rare type of pseudohermaphroditism in genotypically and phenotypically males. They have a uterus, fallopian tubes, and upper part of the vagina; but Mullerian duct fails to regress. In transverse testicular ectopia (TTE), one of the testis moves to the opposite side and both testes pass the same inguinal canal. We report a case of PMDS with TTE presented with a left-sided inguinal hernia and right undescended testis.

CASE REPORT

A 23-year-old man presented with a left-sided reducible inguinal hernia and undescended testis in an empty ill-developed right hemiscrotum. Left scrotum revealed two separate contents, a small 3 × 2 cm oval structure (clinically testis) and another larger structure (clinically the hernia sac with its content) (Figure 1). On taxis, the sac and the left testis both reduced into the abdomen together.

Ultrasonography revealed left-sided normal testis, bulky left epididymis, thickened spermatic cord, and a heterogeneous mass 2.8 × 1.7 × 1.7 mm with minimal vascularity. Partial herniation of the small bowel loops was seen through the internal ring on the left side. Right-sided



Figure 1. Showing normal penis with inguinal hernia on the left side and empty right scrotum.

testis was not seen either in the scrotal sac, inguinal canal, or in any other possible ectopic sites.

In semen analysis, volume was 2 mL and there were no spermatozoa. Serum testosterone level was 862.20 ng/dL (normal range, 280 to 800 ng/dL). Karyotyping showed male XY pattern.

On diagnostic laparoscopy, well-developed uterus, round ligament, fallopian tubes, and the left testis were seen. The left testis was entering the left internal inguinal ring and the right testis migrated to left and was in the left internal inguinal ring. We reduced the hernia and took a biopsy from the right testis to prove TTE (Figure 2). The histopathology revealed seminiferous tubules containing Sertoli cell with no germ cells.

The patient was counseled and re-explored. Total hysterectomy with bilateral salpingectomy followed by bilateral orchidopexy and repair of left-sided hernia were performed (Figure 3). The histopathology confirmed endometrium (proliferative phase), fallopian tube, and vaginal tissue.

DISCUSSION

Nilson first reported PMDS in 1939, with 150 cases reported until now.⁽¹⁾ Mullerian inhibiting substance (MIS), secreted by the Sertoli cells from seven weeks of gestation, causes the regression of the Mullerian duct in the male fetus.⁽²⁾

In PMDS, there is presence of Mullerian derivatives, ie, the

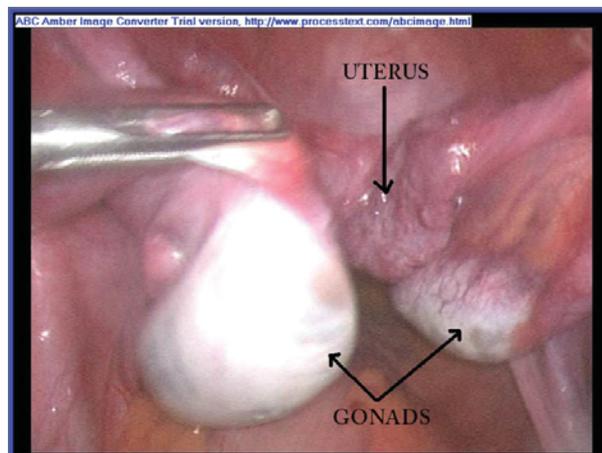


Figure 2. Laparoscopic view showing the right and left gonads with the uterus in between them.

uterus, fallopian tube, and upper part of the vagina in otherwise normally virilized XY male. It results from a failure or mistiming in the synthesis and release, or end organ resistance to MIS.⁽³⁾ The subjects have normal levels of testosterone with normal male secondary sexual characters. The Wolffian duct derivatives and external genitalia develop in a normal male pattern.

More than 100 cases of TTE have been reported;^(4,5) however, the presence of PMDS and TTE together is rare. Persistent Mullerian duct syndrome is mostly found out during surgery for inguinal hernia or cryptorchidism. Transverse testicular ectopia also helps in finding out PMDS, as in our patient.

Persistent Mullerian duct syndrome has two clinical variants. Commoner is unilateral cryptorchidism and contralateral inguinal hernia. The term “hernia uteri inguinalis” is used when the uterus is found in the hernia sac. Sometimes the contralateral testis is found in the sac due to the abnormal mobility of the Mullerian derivatives, known as TTE.⁽⁶⁾ In another variant, bilateral cryptorchidism is seen with the uterus in the pelvis and the testis embedded in the broad ligament.

The undescended testes are at increased risk for malignant transformations into seminoma, embryonal carcinoma, yolk sac tumors, and rarely clear cell adenocarcinoma of the Mullerian duct in PMDS.⁽⁷⁾ Infertility is common with

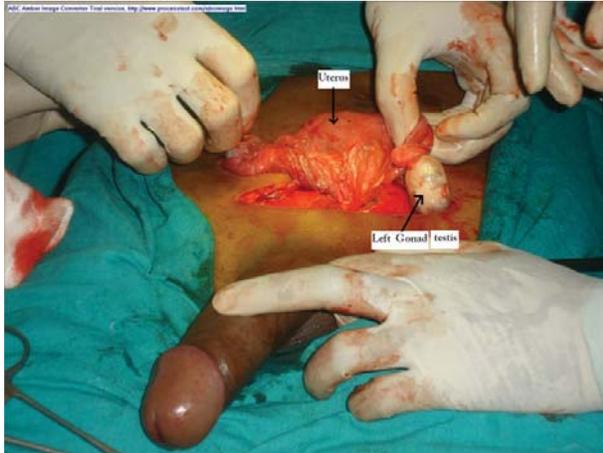


Figure 3. Operative picture showing the uterus with both the testes and normally developed penis.

absence of spermatozoa in semen analysis.

Surgical management is done in stages. Diagnostic laparoscopy confirms anatomy and aids diagnosis with testicular biopsy. Definitive surgery is done in the second stage once biopsy and karyotyping are available; it aims at preserving the testes. Orchidopexy and hysterectomy are done after dissecting the vas deferens from the Mullerian structures. Orchidectomy is indicated if the testis cannot be brought to the scrotum. Orchidectomy should be followed by a lifelong testosterone supplement. Patients with PMDS and TTE are usually infertile. Orchidopexy should be done to prevent malignancy.

CONFLICT OF INTEREST

None declared.

REFERENCES

1. Acikalin M, Pasaoglu O, Tokar B, Ilgici D, Ilhan H. Persistent Mullerian duct syndrome with transverse testicular ectopia: a case report with literature review. *Turk J Med Sci.* 2004;34:333-6.
2. Gujar NN, Choudhari RK, Choudhari GR, et al. Male form of persistent Mullerian duct syndrome type I (hernia uteri inguinalis) presenting as an obstructed inguinal hernia: a case report. *J Med Case Rep.* 2011;5:586.
3. Josso N, Belville C, di Clemente N, Picard JY. AMH and AMH receptor defects in persistent Mullerian duct syndrome. *Hum Reprod Update.* 2005;11:351-6.
4. Mouli K, McCarthy P, Ray P, Ray V, Rosenthal IM. Persistent mullerian duct syndrome in a man with transverse testicular ectopia. *J Urol.* 1988;139:373-5.
5. Fourcroy JL, Belman AB. Transverse testicular ectopia with persistent Mullerian duct. *Urology.* 1982;19:536-8.
6. Karnak I, Tanyel FC, Akcoren Z, Hicsonmez A. Transverse testicular ectopia with persistent mullerian duct syndrome. *J Pediatr Surg.* 1997;32:1362-4.
7. Shinmura Y, Yokoi T, Tsutsui Y. A case of clear cell adenocarcinoma of the mullerian duct in persistent mullerian duct syndrome: the first reported case? *Am J Surg Pathol.* 2002;26:1231-4.