

PERSISTENT MÜLLERIAN DUCT SYNDROME ASSOCIATED WITH TRANSVERSE TESTICULAR ECTOPIA

Abdulrahman A. Al-Bassam, FRCS(Ed)

Persistent müllerian duct syndrome (PMDS) is a rare form of male pseudohermaphroditism, characterized by the presence of a uterus and fallopian tubes due to failure of müllerian duct regression in genotypically normal males.^{1,2} More than 150 cases have been recorded, most of them in adults. The association between the persistent müllerian duct and transverse testicular ectopia is even more uncommon. In 1886, von Cenhossek described the first case of transverse testicular ectopia and in 1895, Jordan described the syndrome of transverse testicular ectopia with persistent müllerian ducts.³

Case Report

A 10-month-old boy presented with a left inguinal hernia and absence of the right testis since birth. Examination of the groins showed a normal phallus, left inguinal hernia, empty right hemiscrotum, impalpable right testis and normal left testis.

Urinalysis, complete blood count and serum electrolytes were normal. At exploration through the left inguinal incision, two spermatic cords were found separated by a rudimentary uterus, thick-walled vagina and ligamentous structure corresponding to the broad ligament. The two vasa deferentia were adherent to the thickened vagina. The left testis was lying in the left hemiscrotum, but were pulled up into the operative field during dissection. The right testis ended at the left mid-inguinal canal (Figure 1). The two testes were biopsied, the two cord structures were carefully separated from the persisting müllerian structures and limited resection of the rudimentary uterus and vagina was done. The associated inguinal hernia was repaired and bilateral orchidopexy with transposition of the right testis to the right hemiscrotum through a suprapubic subcutaneous tunnel was performed.

The postoperative course was uncomplicated. The

karyotype confirmed a male gender 46XY. Renal ultrasound was normal.

Discussion

Normally, the testis is located in the scrotum at birth. Ectopic testes have been reported at different sites, including the superficial inguinal pouch, suprapubic, femoral and perineal areas, and at the base of the penis.⁴

Migration of the testis to the opposite side, where both testes pass through the same inguinal canal, is known as transverse testicular ectopia (TTE). Over 100 cases of TTE have been reported in the literature.^{5,6}

Persistent müllerian duct syndrome is rare, characterized by the presence of well-developed or rudimentary uterus, cervix, vagina and fallopian tubes in a normal 46XY male.

The presence of PMDS with TTE is even more uncommon. In most cases, the PMDS is discovered during surgery for the inguinal hernia or cryptorchidism or by the presence of transverse testicular ectopia. Our patient was found to have PMDS with TTE during surgery for herniotomy in a child with contralateral undescended testis. PMDS represents only a small number of male pseudohermaphroditism. The external genitalia in these patients are normal in shape and size with a central glandular urethral meatus.

The preoperative diagnosis of PMDS and TTE is practically impossible. Adamsbaum et al.⁷ recommended routine pelvic and inguinal area ultrasonography in bilateral cryptorchidism patients and in patients with inguinal hernia of unusually hard consistency. Recently, with experience of laparoscopic surgery for an impalpable testis, the diagnosis of transverse testicular ectopia was possible. Fairfax and Skoog⁸ reported a 14-month-old child with transverse testicular ectopia diagnosed laparoscopically.

As PMDS and TTE are usually discovered incidentally during surgery for undescended testis or inguinal hernia, the optimal surgical approach should include testicular biopsies, herniotomy, orchidopexy and excision of müllerian duct remnants. The vasa deferentia are usually

From the Department of Surgery, King Khalid University Hospital, Riyadh.
Address reprint requests and correspondence to Dr. Al-Bassam: P.O. Box 86572, Riyadh 11632.

Accepted for publication 7 September 1996. Received 7 February 1996.



FIGURE 1. Intraoperative view showing both testes, fallopian tube and rudimentary uterus.

adherent to the lateral walls of the vagina and for this reason, maximum care should be taken to dissect the vas deferens away from the müllerian duct structures. Some authors believe that excision of müllerian duct structures and scrotal orchidopexy are not possible without sacrificing the vasa deferentia.² In our patient, it was possible to dissect the vasa deferentia from the rudimentary uterus and vagina, perform scrotal orchidopexy and excision of müllerian duct structures without risking the vas deferens. We believe that every effort should be made to preserve the vas deferens and testes for possible future fertility. This approach is reported by many authors.^{1,3,7}

Fertility has been reported rarely in a few cases.^{2,9} Martin et al.³ had reported a 32-year-old man with transverse testicular ectopia and a persistent müllerian duct who had a normal sperm count, but the motility index was zero, implying an intrinsic defect in spermatogenesis. The other possible cause of low sperm counts and poor motility in most of these patients is the partial duct obstruction.

There have been reports of embryonal cell carcinoma, seminoma, choriocarcinoma and teratoma in patients with cryptorchidism and persistent müllerian duct. However,

the malignancy incidence appears to be similar to that of a normal cryptorchid child.^{2,10,11}

In conclusion, a conservative surgical approach for this rare syndrome in the form of orchidopexy and partial excision of the müllerian duct remnants without risking the vas deferens is recommended. A long follow-up will be needed for assessment of the fertility in these patients.

Acknowledgment

I would like to thank Ms. Cora Rivera for her assistance in typing this manuscript.

References

1. Loeff DS, Imbeaud S, Reyes HM, Meller JL, Rosenthal IM. Surgical and genetic aspects of persistent müllerian duct syndrome. *J Pediatr Surg* 1994;29:65-6.
2. Pappis C, Constantinides C, Chiotis D, Dacou-Doutetakis C. Persistent müllerian duct structures in cryptorchid male infants: surgical dilemmas. *J Pediatr Surg* 1979;14:128-31.
3. Martin EL, Bennett AH, Cromie WJ. Persistent müllerian duct syndrome with transverse testicular ectopia and spermatogenesis. *J Urology* 1992;147:1615-7.
4. Gallady ES, Redman JF. Transverse testicular ectopia. *Urology* 1982;19:181-6.
5. Mouli K, McCathy P, Ray V, Rosanthat IM. Persistent müllerian duct syndrome in a man with transverse testicular ectopia. *J Urology* 1988;139:373-5.
6. Fourcroy JL, Belman AB. Transverse testicular ectopia with persistent müllerian duct. *Urology* 1982;19:536-8.
7. Adamsbaum C, Rolland Y, Josso N, Kalifa G. Radiological findings in three cases of persistent müllerian duct syndrome. *Pediatr Radiology* 1993;23:55-6.
8. Fairfax CA, Skoog SJ. The laparoscopic diagnosis of transverse testicular ectopia. *J Urology* 1995;153:477-8.
9. Sheehan SJ, Tobbia IN, Ismail MA, Kelly DG, Duff FA. Persistent müllerian duct syndrome. Review and report of three cases. *Br J Urology* 1985;57:548-51.
10. Brook CGD, Wagner H, Zachmann M, et al. Familial occurrence of persistent müllerian structures in otherwise normal males. *Br Med J* 1973;1:771-3.
11. Melman A, Leiter E, Perez JM, et al. The influence of neonatal orchidopexy upon the testis in persistent müllerian duct syndrome. *J Urol* 1981;125:856-8.