Transverse Testicular Ectopia In An Adult Male; A Rare Form of Persistent Mullerian Duct Syndrome: A Case Report

Persistan Müllerian Kanal Sendromunun Ender Formu; Erişkin Erkekte Transvers Testiküler Ektopi: Olgu Sunumu

Persistan Müllerian Kanal Sendromu / Persistent Mullerian Duct Syndrome

Alp Özgür Akdemir, Utku Lokman, Çetin Volkan Öztekin, Mustafa Sungur, Öztuğ Adsan
Department of Urology II Clinic of Ankara Numune Education and Research Hospital, Ankara, Türkiye.

Özet

Persistan Müllerian Kanal Sendromunun (PMKS) ender formu olan; PMKS ve Transvers Testiküler Ektopi (TTE) birlikteliği, oldukça nadir görülen bir sendromdur. Bu yazıda infertilite nedeniyle polikliniğimize başvuran 24 yaşında bir erkek hastada saptanan PMKS ve TTE birlikteliği tartışılmıştır.

Anahtar Kelimeler

Transvers Testiküler Ektopi; Müllerian Kanal Sendromu; Hernia Uteri Inguinale.

Abstract

The co-existence of persistant mullerian duct syndrome (PMDS) together with transverse testicular ectopia (TTE) is a rare clinical entity. This report includes the discussion regarding a case of PMDS with TTE in a 24 years old male patient, and review of the related literature.

Keywords

Transverse Testicular Ectopia; Mullerian Duct Syndrome; Hernia Uteri Inguinale,

DOI: 10.4328/JCAM.627 Received:20.02.2011 Accepted: 11.02.2011 Publihed Online: 01.03.2013 J Clin Anal Med 2013;4(2):141-143
Corresponding Author: Alp Özgür Akdemir, Department of Urology II Clinic of Ankara Numune Education and Research Hospital, Ankara, Türkiye.
GSM: +905322324024 E-mail: drakdemir@gmail.com, aoakdemir@yahoo.com

Introduction

PMDS is a rare clinical form of sexual differentiation disorder characterized by patients presenting with a male phenotype, 46XY genotype and structures of mullerian duct remnants like uterine tubes, uterus and one third proximal part of the vagina.[1] Presence of a uterus or tubae are usually discovered during an ingunal hernia repair or during an open or laparoscopic exploration for undescended testes.[2] Patients are almost always diagnosed before adolescence. As testes and mullerian duct remnants can be found together in an ingunal hernia sac, TTE was first defined by Nilson as "Hernia uteri inguinalis in men" in 1939.[1] TTE can be observed rarely, as the contralateral testis may be pulled in the hernia sac together with the herniated structures. Although this rare form of PMDS was mostly reported in children in the literature, we hereby report a case of PMDS with TTE in an adult male who had the diagnosis at age 24 and who applied to our institution with infertility as the primary complaint.

Case

Twenty-four years old male patient applied to our institution beacuse of infertility. He had a male phenotype and he was infertile after two years of marriage without applying any forms of contraception. TTE was not diagnosed before, although he underwent a bilateral inguinal hernia repair 1 year ago (Figure1). On physical examinaton, he had male type secondary sex characteristics, and bilateral incision scars due to previous operations. Penis shape and size were normal. The right hemiscrotum included two 3.5x3.5cm lobulated masses which were thought to be the testes, while the left hemiscrotum was atrophic (Figure 1).

Scrotal sonogram reported the lobulated structures in the right



Figure1. Bi-lobulated right hemiscrotal mass. (Note the male phenotype and male type secondary sex characteristics.)

hemsicrotum as testes. Exploration was performed under general anesthesia through a pfannenstiel incision. No intraperitoneal pathologies were found during abdominal exploration. Two tubular structures other than the funicular elements were identified during exploration of the right inguinal canal. These were dissected free until the external inguinal ring was reached. A 3cm scrotal incision was made in order to aid exploration and preserve the testes and cord structures as the testes could not be brought out from the inguinal incision (Figure 2). Another structure which was attached to and covered both testes anteriorly was observed. Further dissection of the structures revealed the presence of a uterus, fallopian tubes with fimbriae and proximal vagina together with both testes in the right hemiscrotum (Figure 3).



Figure2. Scrotal exploration reveals presence of both testes in right hemiscrotum.

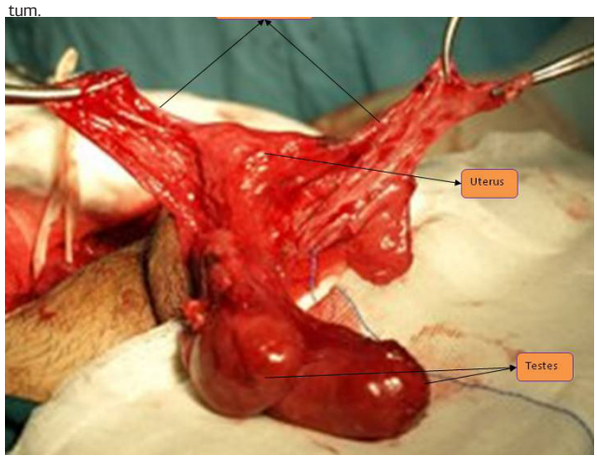


Figure3. Testes, uterus, fallopian tubes with fimbriae

Uterus and the adjacent structures were freed from the testes, completely dissected from the surrounding tissues and excised from the point at which the vagina ended blindly in the posterior pelvis. Spermatic cords were identified, testicular biopsy was performed and the testes were placed in the scrotum and fixed. Testicular biopsy resulted as normal testicular tissue. Pathological examination of the excised specimen was reported as

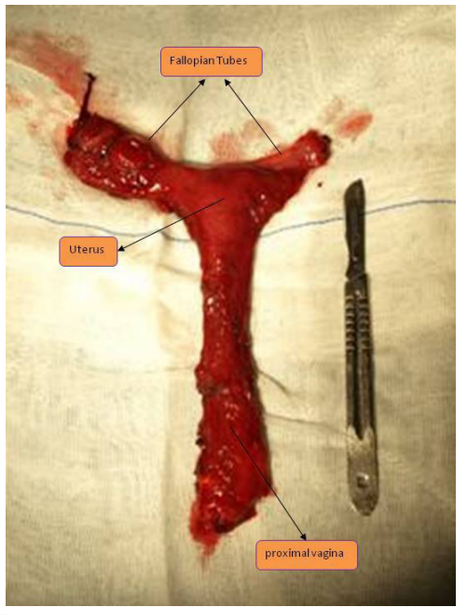


Figure4: Excised specimen including proximal vagina, uterus and fallopian tubes.

uterus and bilateral uterine tubes with histological findings of endometrium, cervix and left and right uterine tubes. Karyotype analysis from peripheral blood resulted as 46XY. Hormonal tests

showed low FSH and free testosterone levels. Sperm analysis was reported as oligoastenoteratozoospermia.

Discussion

Clarnette et al proposed 3 different categories for patients with PMDS.[3] These categories are: 1. Presence of bilateral intraabdominal testes found in analogue positions to ovaries (60-70%), 2. presence of a testis in the hernia sac, or a scrotal testis with a contralateral inguinal hernia (20-30%), and 3. presence of both testes in the same hernia sac together with uterus and uterine tubes, which results in TTE (10%). Similarly, PMDS is seen in 30 to 50% of TTE cases and is believed to play an important role in its etiology.

TTE, which is also named as testicular pseudoduplication is a rare condition with an unclear etiology. It was defined in 1895 by Jorda[4] after it was first reported in 1886 by Von Lenhossek in 1886 . The theories regarding TTE patogenesis include testicular adhesion, a defective inguinal canal, presence of an aberrant gubernaculum, adhesion of the growing testis to the wolfian duct or traction of the testis by Mullerian duct remnants into the contralateral hemiscrotum.[5] PMDS can be present in almost half of the patients with TTE . The reason for this co-existence may be the prevention of testicular descent to the scrotum by the traction caused by the unregressed Mullerian structures or the direction of both testes to the same hemiscrotum. Although the co-existence of TTE with PMDS was reported in more than 100 cases in the literature, most of the cases are children or adolescents. We report our case in order to emphasize the possible delays in diagnosis in these patients and the need for particular attention in patients with abnormal inguinal and scrotal anatomy. While the diagnosis of TTE can be achieved preoperatively by scrotal ultrasound, magnetic resonance (MR) imaging and MR venography are also recommended for preoperative localization of non-palpable testes.[6]

The ideal surgical technique for the correction of PMDS is debateful. While some authors do not recommend testicular biopsy in TTE patients with a male karyotype, others recommend bilateral testicular biopsy as the first, and orchidopexy as the second step of treatment.[7] The dissection of uterus and fallopian tubes from testis and vas deferens requires special attention, as an injury to the deferential artery may occur and testicular blood supply may be compromised.

We removed the mullerian structures as they carry the risk of malignancy. They were removed before testes were placed in the scrotum and fixed. The incidence of testicular tumors is high in patients with PMDS like the patients with undescended testes. Sixteen patients with PMDS and testicular neoplasia were reported[8] in literature.

As a result, it must always be kept in mind that PMDS can also be present in patients with TTE diagnosed either by palpation of both testes in one hemiscrotum or during inguinal exploration. We think that, the primary goals in treatment of PMDS should include the obtaining of biopsies from the intra-operative identified structures, extensive surgical removal of mullerian structures in order to prevent any malignant transformations and placing of the testes in a palpable position in the scrotum with special care to avoid injuries to testes and other adjacent structures.

References

1. Nilson O. Hernis uteris inguinalis beim manne. Acta Chir Scand. 1939; 83: 231-239.
2. Beyribey S, Cetinkaya M, Adsan O, Memis A, Oztürk B. Persistent Müllerian Duct Syndrome (Reports of Two Cases). Scand J Urol Nephrol. 1993;27(4): 563-565.
3. Clarnette TD, Rowe D, Hasthorpe S, Hutson JM. Incomplete disappearance of the processus vaginalis as a cause of ascending testis. J Urol. 1997; 157(5):1889-1891.
4. MacLaughlin DT, Donahoe PK. Sex determination and differentiation . N Engl J Med. 2004; 350(4):367-378.
5. Tiryaki T, Hucumenoglu S, Atayurt H. Transverse testicular ectopia associated with persistent mullerian duct syndrome. A case report. Urol Int. 2005; 74: 190-192.
6. Von Lenhossek MN. Ectopia testis transversa. Anta Anz 1886; 1: 376-381.
7. Jordan M. Ein Fall von einseitigem descensus testiculorum. Beitr Klin Chir. 1895;15:245.
8. Buchholz NP, Biyabani R, Herzig MJ, Ali A, Nazir Z, Sulaiman MN. et al. Persistent Müllerian duct syndrome. Eur Urol. 1998; 34(3):230-232.