CASE REPORT

A Simplified Management of Transverse Testicular Ectopia in Patients with Persistent Mullerian Duct Syndrome

Massimiliano Silveri[1]*, Antonio Zaccara[1], Marco Cappa[2]

Persistent müllerian duct syndrome (PMDS) in the majority of cases is discovered during surgery for inguinal hernia or cryptorchidism. A transverse testicular ectopia (TTE) with cryptorchidism may be very rarely associated to PMDS. Assuming that müllerian remnants have a very low malignant degeneration potential if compared to the malignancy risk of an undescended and not relocated testis, we describe a simplified surgical technique of orchiopexy that avoids an extensive anatomical dissection, in this way minimizing the risk of losing the deferential blood supply to the testis.

Keywords: radical cystectomy; ileal conduit, cutaneous ureterostomy; orthotopic neobladder; appendix

INTRODUCTION

Persistent müllerian duct syndrome (PMDS) is frequently discovered during surgery for inguinal hernia or cryptorchidism[(1)]. In this disorder of sex development (DSD), the patients show müllerian remnants located at scrotal, inguinal or intraabdominal level. A transverse testicular ectopia (TTE) with cryptorchidism may be rarely associated to PMDS because of defect in regression of fetal müllerian structures and concomitant aberrant testicular descent[(2)]. In these rare cases, a planned surgical approach is advisable confirming the opportunity to perform, as a first step, a laparoscopic diagnostic approach in all cases of unpalpable testis.
We describe a simplified orchiopexy technique performed in one of these rare cases in order to prevent devascularisation or direct damage to the vas.

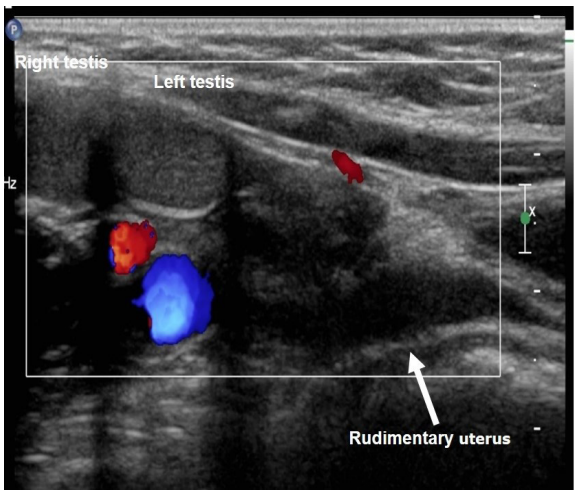

Figure 1. Ultrasound showing two separate gonadal structures both located in the right inguinal channel.

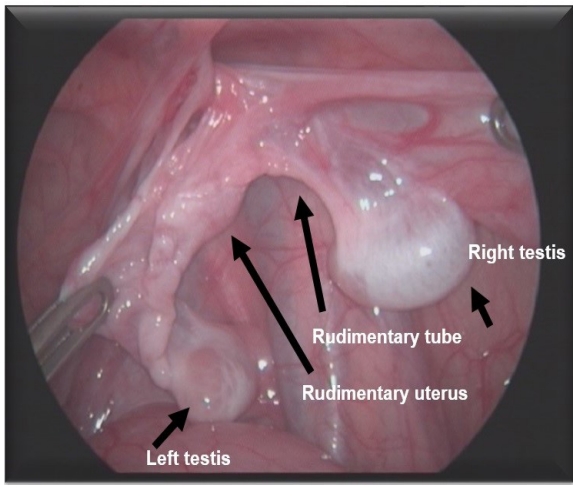

Figure 2. Laparoscopy showing the anatomy at the level of the right internal inguinal ring, with a rudimentary uterus and a rudimentary tube interposed between the two testes.

[1]Department of Surgery, Ospedale Pediatrico Bambino Gesù, Rome, Italy.
[2]Department of Pediatric Endocrinology, Ospedale Pediatrico Bambino Gesù, Rome, Italy.
*Correspondence: Department of Surgery , Ospedale Pediatrico Bambino Gesù, Rome, Italy.
Piazza Sant'Onofrio 4, 00100, Rome - Italy.
Phone: 0039668593358. Fax: 0039668593373. E mail: massimiliano.silveri@opbg.net.
Received October 2019 & Accepted April 2020

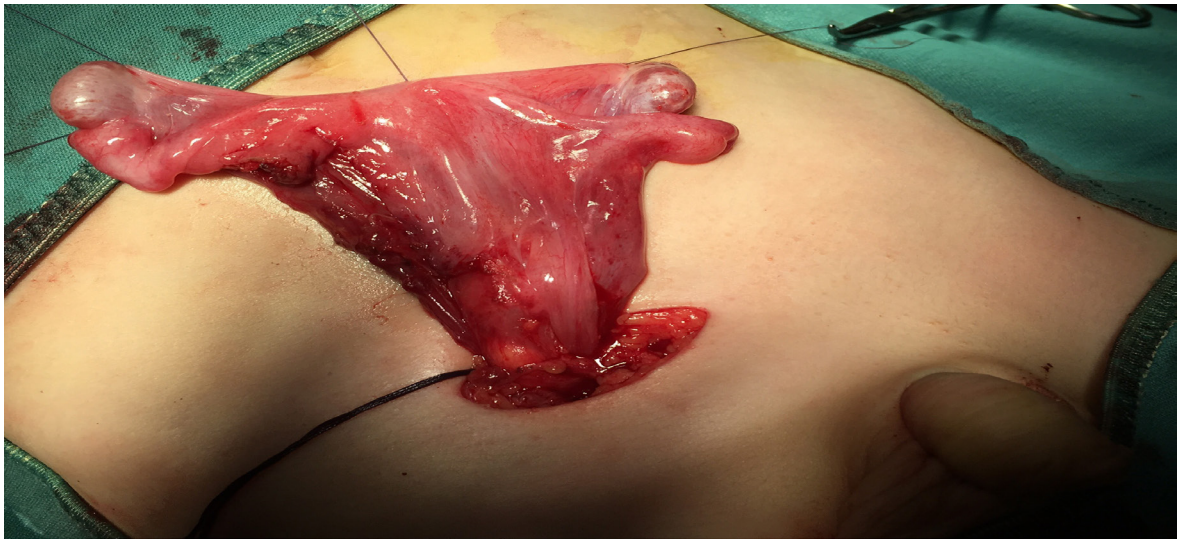

Figure 3. Surgical view of the műllerian structures and both the gonads exteriorized through an inguinal incision.

CASE REPORT

A 2-year-old boy came to our attention for a left impalpable testis. Following a neonatal diagnosis of bilateral cryptorchidism, the baby underwent an ultrasound (US) at 6 months of age showing a right testis in the scrotum and an extremely reduced gonadal structure approximately at the level of the left inguinal ring. However, the evidence of two separate and palpable structures both located in the right inguinal channel (**Figure 1**) alerted our intersex team about the possibility of an underlying DSD. A HCG test for the exploration of Leydigian function was normal and karyotype resulted as male 46, XY.

Technique

The child underwent a laparoscopy (**Figure 2**) showing the two normal testes, both located proximally to the right inguinal ring, with a rudimentary uterus and rudimentary tubes interposed and a not clear anatomy of the left vas. The baby underwent an open procedure through a right inguinal incision (**Figure 3**). Separation of the vas from the remnants was not possible except, most likely, with its sacrifice. Therefore, a simplified pull-through of the complete anatomical package with repositioning of the left testis through a partial violation in the septal scrotum, was successfully attempted (**Figure 4**).

At 3 years follow-up, both testes are correctly located in the scrotum with a progressive catch-up growth highlighted by clinical (twice a year) and US (once a year) controls.

DISCUSSION

This eased surgical approach, in this particular category of patients, can be achievable only with the assumption that müllerian remnants do not have any malignant degeneration potential[3]. Persistent müllerian duct syndrome (PMDS) is a rare form of DSD in which a phenotypically normal male has müllerian structures that fail to regress. This embryological event is upregulated by a glycoprotein produced by Sertoli cells and called Müllerian inhibiting substance (MIS))[4]. Mutations in

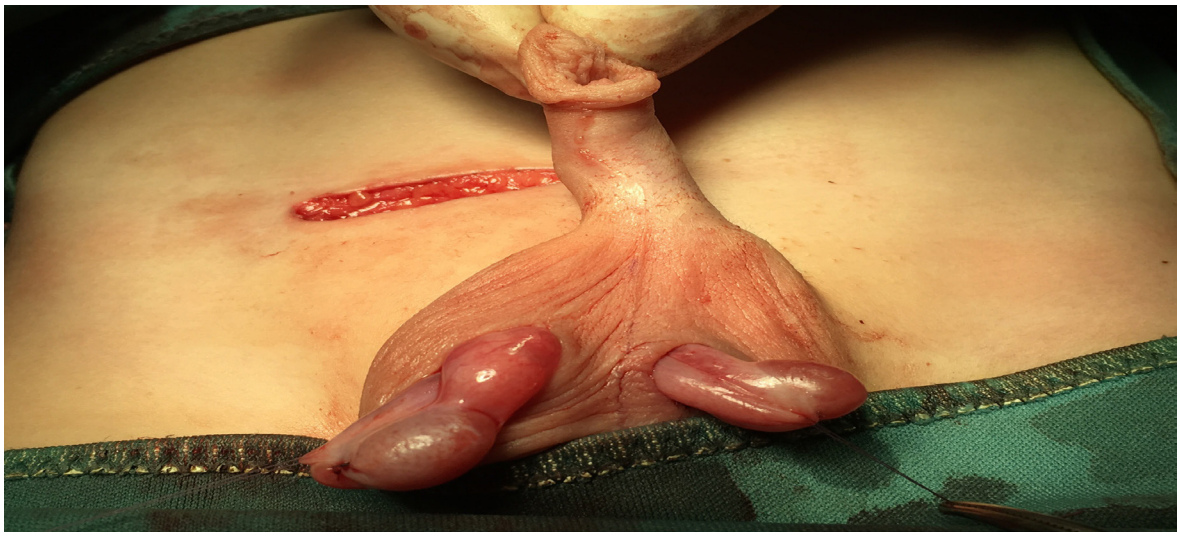

Figure 4. Repositioning of both testes in their respective anatomical sites obtained through a trans-scrotal approach.

MIS and MIS receptor genes, mapped to chromosome 19 (MIS) and chromosome 12 (MIS type II specific receptor) cause lack of MIS secretion or lack of translocation to the surface membrane with inactivity of the MIS receptor[5]. Despite many hypotheses on the relationship between PMDS and TTE[6,7], the simple anatomic close contiguity of the testis to the persistent müllerian ducts is believed to be the cause of cryptorchidism in PMDS. There is a dualism in literature about whether or not to remove müllerian structures in order to prevent a potential malignant degeneration[3,8]. Recent description of gynaecological malignancies in retained müllerian structures[9] is estimated in 3-8%. The overall incidence of malignant transformation in PMDS testes is similar to the rate in abdominal testes in otherwise normal men[9,10] (18%), in this way representing another argument in favour of early orchiopexy.

Starting from these assumptions, we successfully adopted the above-described approach[11] as the gold-standard management in these rare cases. In our opinion, as suggested by the normal pattern of our biopsied gonads, even the previously reported foreclosures about the fertility potential of PMDS testes should be reconsidered assuming that the only real limit is probably cryptorchidism. A close and long term instrumental and clinical follow-up is mandatory above all for that concerning the risk of possible late onset gynaecological malignancies.

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