

Laparoscopically assisted treatment of transverse testicular ectopia with persistent Müllerian duct syndrome: a case report and review of the literature

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Abstract

Background: Transverse testicular ectopia coexisting with persistent Müllerian duct syndrome is a rare malformation and evidence regarding the optimal treatment of these patients is still unclear.

Description of the case: We present the case of a 4-month-old boy in whom laparoscopy was utilized for the surgical correction of transverse testicular ectopia and excision of Müllerian remnants.

Conclusion: Based on current literature and the presented case, we support that laparoscopy is a feasible and safe procedure in patients with transverse testicular ectopia and persistent Müllerian duct syndrome. Hippokratia 2016, 20(1): 88-89

Keywords: Transverse testicular ectopia, crossed testicular ectopia, laparoscopy, persistent Müllerian duct syndrome

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Introduction

Transverse testicular ectopia (TTE) is an uncommon congenital malformation where both testicles descent (or tend to descent) through the same inguinal canal¹. An ipsilateral inguinal hernia coexists in most cases. The simultaneous presence of Müllerian remnants [persistent Müllerian duct syndrome (PMDS)] may further complicate the management of these patients. In the male embryo, Müllerian structures regress under the effect of the anti-Müllerian hormone (AMH) around the seventh week of gestation. In PMDS there is a defect in the secretion of AMH or in the AMH receptors².

Case report

A 4-month-old boy was referred to the pediatric surgery department of Agios Loukas hospital due to impalpable testicles on both sides. On ultrasound examination, one testis was located in the right inguinal canal and the other one near the right internal inguinal ring. Both testicles had normal size, morphology and vascularization. There was also present a mild inguinal hernia. On clinical examination, only the one testis was just palpable in the right inguinal canal, close to the subcutaneous inguinal ring. Laparoscopy was performed using 5 mm, zero degree telescopes and two additional 3 mm ports on either side of the umbilical port. A testicle was identified close to the right internal inguinal ring (Figure 1). Spermatic vessels were entering the inguinal canal. The inguinal testis was brought back into the abdomen. A Müllerian

remnant connecting the two testicles was identified and excised by careful dissection in order not to injure the spermatic vessels or the vas deferens. By laparoscopic dissection, adequate length of the spermatic vessels and spermatic ducts of both testicles was gained. Through a scrotal incision, a Kelly clamp was inserted into the inguinal canal to exteriorize the right testis, which was subsequently placed in a subdartos pouch. The same procedure was followed for the left testicle. No complications occurred in the postoperative period, and the child returned home the following day. Follow-up consisted of clinical and ultrasound examination at one month, six months, and one year postoperatively. Both testicles were normal in size and vascularization one year after the orchiopexy.

On pathologic examination of the Müllerian remnants, there was a presence of a central sinusoid-form duct with focal formation of micropapillary wall projections and microtubular spaces which were covered by one layer of uniform tall cubocylindric or tall cells (Figure 2).

Discussion

The frequency of PMDS has been adequately estimated, yet the coexistence with TTE makes this pathologic condition even rarer³. Laparoscopy has been used in the diagnosis and anatomic clarification of TTE, but only few cases in the literature were treated laparoscopically⁴. Surgical approach depends on whether TTE coexists with PMDS, a patent processus vaginalis or inguinal hernias,

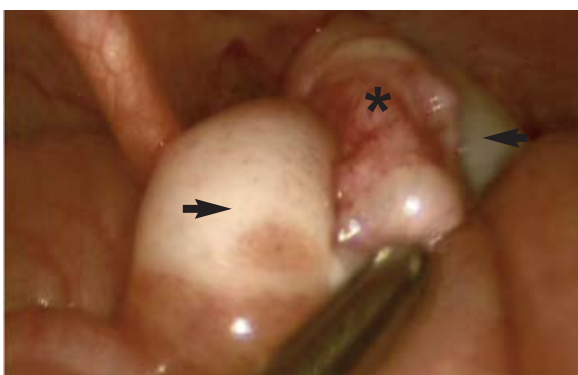


Figure 1: Laparoscopic image showing intra-abdominal presence of both testicles (arrows) and Müllerian remnant (star) connecting the two testicles.

hypospadias or scrotal anomalies⁵. When no Müllerian remnants are recognized during initial laparoscopy, a careful division of the vas deferens and spermatic vessels with orchiopexy can be performed^{6,7}. In the case of PMDS, two different surgical techniques are most commonly applied for TTE. In the first, Müllerian structures are preserved and split in half, while in the second, these structures are removed as extensively as possible. Their difference lies in the belief whether these structures are in danger for future malignant transformation. Bowen et al reported on the successful laparoscopic management of PMDS and intra-abdominal testicles with preservation of the Müllerian remnants⁸. During the recent years, malignancies such as adenocarcinoma or adenosarcoma arising from Müllerian remnants were reported⁹⁻¹¹. Farikullah et al found eleven cases of adenocarcinoma and squamous cell carcinoma in patients with PMDS the last forty years¹². Long-term follow-up should be considered mandatory when Müllerian structures have been preserved or a viable intra-abdominal testis is brought in the scrotum. Fertility should also be assessed in later life in children with PMDS^{13,14}.

It should be emphasized that excision of uterine structures might disrupt testicular blood circulation and also cause an injury to the vas deference. Farikullah et al re-

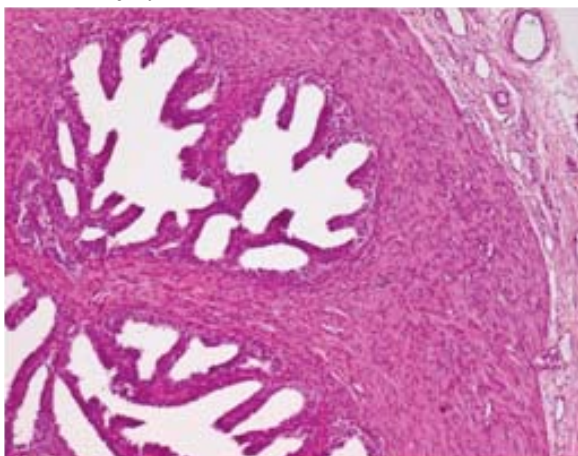


Figure 2: Histologic appearance of the Müllerian remnant. Micropapillary wall projections covered by one layer of tall columnar epithelium is apparent (stain: hematoxylin-eosin, x2000).

ported successful laparoscopic removal of Müllerian remnants in eight patients. Three of these cases were relatives, and the other three had coexistent hypospadias¹². Yamada et al reported on a testicular seminoma in a patient with TTE and PMDS where no operation was previously performed and the testis had been intraabdominal¹⁵.

We believe that excision of Müllerian duct remnants should be performed, when this procedure does not endanger testicular vascularization or integrity of the vas deferens, because of the risk of malignant transformation. Laparoscopy provides excellent magnified operative view, helping in performing the operation meticulously and safely.

Conflict of interest

Authors declare no conflicts of interest.

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