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RESEARCH ARTICLE

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Müller's Canals Persistence Syndrome: About 2 Cases

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ABSTRACT

The syndrome of persistence of Mullerian derivatives is a rare form of internal male pseudohermaphrodism. It is characterized by the presence of the uterus, tubes and upper vagina in a boy otherwise normally virilized with a karyotype 46 XY. It is the consequence of a deficit in anti-Mullerian hormone or an abnormality of its receptors. We report a case of intraoperative discovery during a diagnostic laparoscopy for non-palpable cryptorchid testes.

ARTICLE HISTORY

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Introduction

Müller's channel persistence syndrome (PMDS) is a rare form of internal male pseudohermaphrodism, in which a development of Mullerian structures occurs in a male phenotype and normal genotype 46 XY [1-3]. It is the consequence of a deficit in anti-Mullerian hormone (AMH) or an abnormality of its receptors [1,3]. Its discovery is often made during a hernia cure or cryptorchidism and exceptionally in front of a gonadal tumor [1,3]. We report 2 cases of laparoscopic discovery for diagnosis of non-palpable cryptorchid testes.

Observation 1

29-year-old single patient, received in consultation in january 2018 for bilateral cryptorchidism. Clinical examination found a male phenotype patient, normally developed penis, empty pouches, bilateral nonpalpable testes.

Abdominal ultrasound does not visualize the testicles

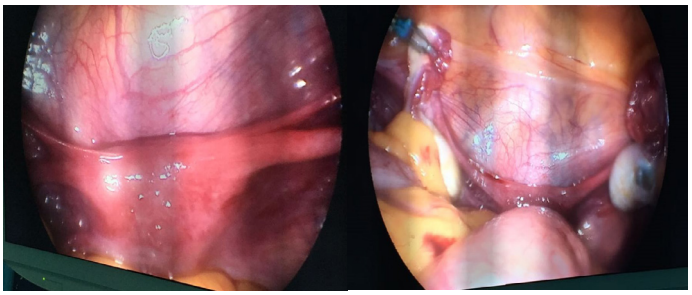
Abdominal CT found a retrovesical mass (contiguous testis or tumor?)

Spermogram: azoospermia

Tumor markers (Beta HCG, Alpha FP, LDH) negative, FSH / LH 3x normal, estradiol / progesterone: normal, Inhibin B 2x normal, normal testosterone.

A diagnostic laparoscopy performed found intraperitoneal and retrovesical female structures: hypotrophic uterus, fallopian tubes, round ligaments, utero-ovarian ligaments and two heterogeneous gonads attached to the pavilions (Figures 1 and 2).

Medial incision under umbilical allowed the release of the testicles up to the level of the iliac vessels (Figure 3) and their fixation in prevesical (facilitating ultrasound surveillance). Biopsy of the bilateral testicular pulp with extemporaneous anatomopathological examination did not reveal the presence of malignant degeneration. The postoperative consequences were simple.



Figures 1 and 2: Coelioscopic view.

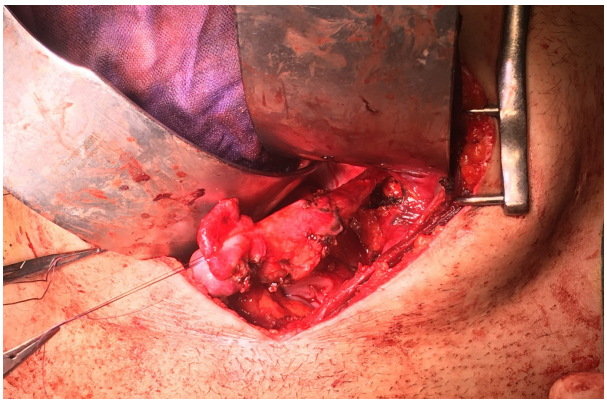


Figure 3: Dissection of the left spermatic cord.

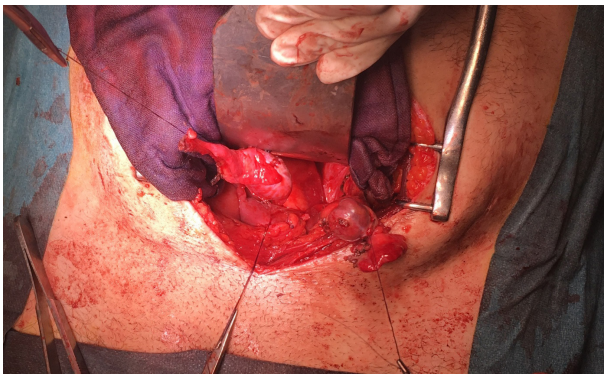


Figure 4: Aspect after dissection of the short spermatic cords, not allowing the externalization of the testicles.

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The genetic study showed a 46 XY karyotype without associated abnormalities

Pathological study: bilateral testicular atrophy without neoplastic lesions.

Observation 2

35-year-old patient, single, seen in September 2020 for bilateral cryptorchidism. The clinical examination found a patient of male phenotype, penis normally developed, bursa empty, bilateral non-palpable testicles

Abdominal ultrasound does not show the testicles

Abdominal CT: absence of visualization of the testicle on this examination, with evidence of female genitalia (vagina and uterus).

Spermogram: azoospermia

Tumor markers (Beta HCG, Alpha FP, LDH), Normal testosteronemia.

A diagnostic laparoscopy performed finds female structures intraperitoneally and retrovesically: hypotrophic uterus, fallopian tubes, round ligaments, utero-ovarian ligaments and 2 heterogeneous gonads hanging from the pavilions.

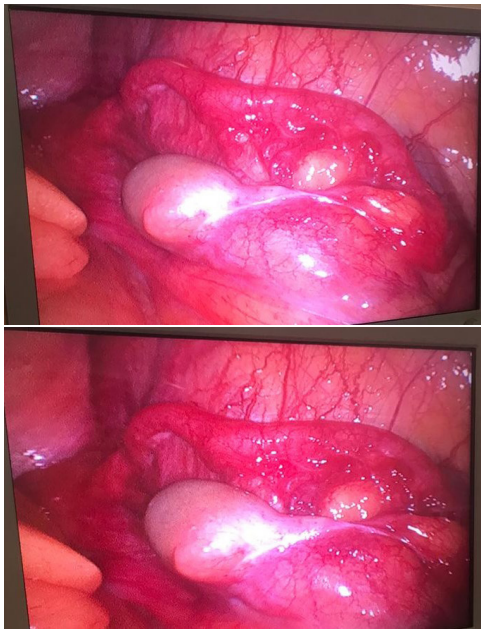
Left inguinal incision allowed the testis to be released to the level of the iliac vessels, showing a nodule at the upper edge of the testis, biopsy and extemporaneous examination performed: pure seminoma an orchiectomy performed (Figures 1 and 2) ; and the patient was send to radiation therapy and chemotherapy.

Right inguinal incision allowed the testis to be released to the level of the iliac vessels and their prevesical fixation (facilitating ultrasound monitoring) (Figure 3).

A biopsy of the testicular pulp with extemporaneous pathological examination did not reveal the presence of malignant degeneration.

The postoperative consequences were simple.

Genetic study showed a 46 XY karyotype with no associated abnormalities



Figures 1 and 2: coelioscopic view witha nodule at the upper edge of the testis

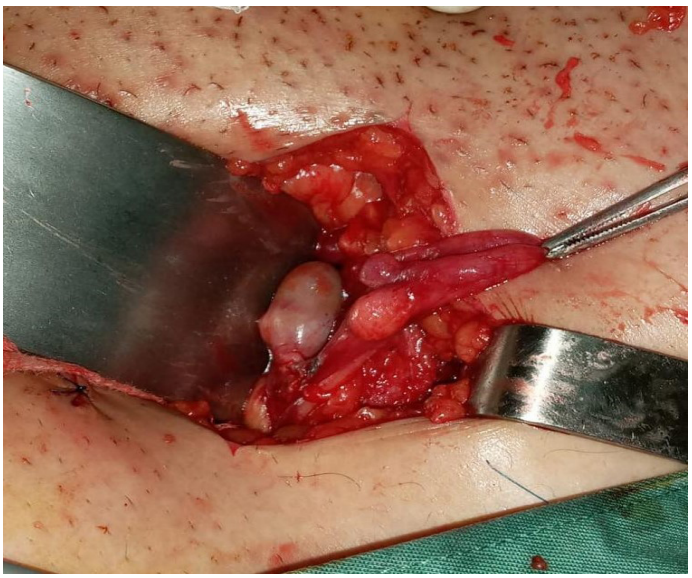


Figure 3: Dissection of the right spermatic cord.s

Discussion

PMDS is a rare form of internal male pseudohermaphrodism, with fewer than 200 cases reported in the literature [2]. It is characterized by the presence in an individual of a uterus, the fallopian tubes and the upper part of the vagina, with, moreover, male-type external genitals, the karyotype is usually 46 XY.

PMDS is a consequence of AMH deficiency or peripheral tissue resistance due to receptor abnormality [1,2].

Indeed, from the eighth week of gestation, the AMH secreted by the sertoli cells of the male fetus causes the regression of Mullerian structures. Only Morgani's hydatid, the rudimentary equivalent prostate tract of the vagina and cervix, and the veran montanum represent the hymen [1].

The persistence and development of Müller's canals and the non-masculinization of gubernaculum testis, which remains as long as a round ligament, would explain the three groups of PMDS [2] (Figure 5).

• group A: both testicles are intra-abdominal (female type);

• group B: a testicle is in the inguinal or scrotal position, the other being in the intra-abdominal position (Male type);

• group C: both testicles and fallopian tubes are in the same hemiscrotum (transverse testicular ectopia).

The diagnosis is often fortuitous during a surgical procedure for cryptorchidism or hernia repair [1,3]. The discovery in front of a testicular tumor is exceptional. In the literature, 40 cases of PMDS associated with gonadal tumors were published. Only four cases had bilateral testicular tumors [3,4].

Berkmen [5] recommends that the diagnosis of PMDS be made radiologically, but neither ultrasound nor CT can help to do so. MRI does not seem more efficient [1].

In combination with a gonadal tumor, the radiological diagnosis becomes more difficult. The Müllerian structures can be confused with the CT tumor, as is the case of our patient, whose diagnosis was certain at laparoscopy.

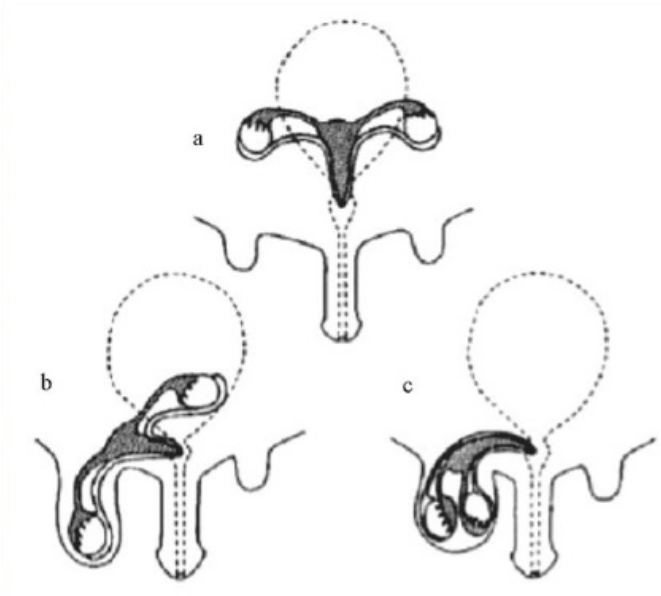


Figure 5: PMDS classification: a: female type; b: male type; c: transverse testicular ectopia.

The biological assessment includes a karyotype to confirm 46 XY masculinity, an AMH and plasma estradiol assay with LH-RH [1].

Surgical treatment of PMDS is made difficult because of two anatomical elements: the close connection between the vas deferens and the lateral walls of the uterus sometimes associated with the brevity of the spermatic pedicle [1,3,5]. It is necessary to obtain at least one unilateral scrotal lowering, for the other testicle two options are possible [1]:

• externalization in the prepubian or inguinal high position;

• Scrotal lowering when taking a vasectomy of necessity.

The uterus must be preserved at least partially since its removal seriously threatens the vasculature of the deferents who walk along its lateral faces.

As a result, only proximal salpingectomy and fundic hysterectomy [1,2] are allowed.

The prognosis of PMDS is related to the high risk of testicular degeneration, which justifies long-term monitoring. The degeneration of Müllerian structures is exceptional, three cases were described in the literature [4]. Infertility is always to be feared in case of bilateral cryptorchidism. However, cases of paternity from men to uterus were reported in the literature [1].

In case of bilateral testicular tumor, hysterectomy is indicated, as long as the patient undergoes bilateral orchiectomy [3,4].

Therapeutic management (chemotherapy, radiotherapy, residual mass surgery) and monitoring should be the same as in other testicular tumors not associated with PMDS [5].

Conclusion

The PMDS should be mentioned in front of any bilateral cryptorchidism. Laparoscopy is of major interest for diagnosis. The surgical treatment must be conservative vis-à-vis the testes and structures Müllériennes who depend on the vascularization of the seminal tract. The increased risk of degeneration of externalised testes warrants rigorous and long-term monitoring.

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