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REVIEW ARTICLE

Hernia uterine inguinale as an intraoperative diagnosis of male pseudohermaphroditism: case report

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Abstract

Persistent Müllerian duct syndrome is a very rare form of pseudohermaphroditism characterized by the presence of structures deriving from persistent Müllerian duct in a patient phenotypically and occasionally genotypically expressed as a normal male. Utero Inguinal Hernias are a rare entity, which in the female population reaches 1% of incidence. However, in the male population, it is directly associated with abnormalities of sexual differentiation. We present the exceptional case of a male patient with no previous diagnosis of intersexuality who is scheduled for a right inguinal repair of a hernia. During the operative management, structures compatible with the uterus, fallopian tubes, and ovaries were identified in the hernial sac. Reduction of the hernial content to the peritoneal cavity was performed and the inguinal defect was resolved with placement of a prosthetic mesh by Lichtenstein technique.

Keywords: *Inguinal hernia. Disorders of sexual development 46 XY. Male infertility. Herniorrhaphy.*

Introduction

Uterine inguinale hernias are a rare entity, reaching 1% of incidence in the female population. However, in the male population, it is directly associated with abnormalities of sexual differentiation or intersex conditions such as male pseudohermaphroditism or also called Type I Müllerian duct persistence syndrome, with no more than 150 cases described worldwide. Intersex states usually have an early manifestation that allows multidisciplinary intervention, but cases of silent courses are reported in phenotypically male patients whose only clinical signs are infertility or cryptorchidism and whose diagnosis is made accidentally during an inguinal hernioplasty or an orchidopexy[1,2]. The following is the case of an adult male patient, infertile, with right homolateral cryptorchidism, who was scheduled on an outpatient basis for a right

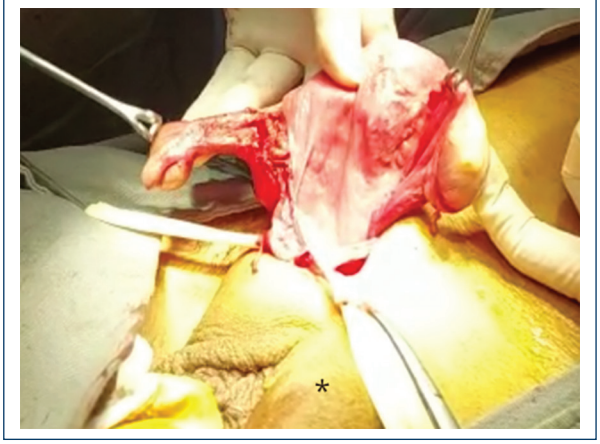

Figure 1. Inguinotomy and content of hernial sac compatible with Müllerian structures (uterus, fallopian tubes, and ovaries). Caudally male reproductive structures, testicles, and penis (*).

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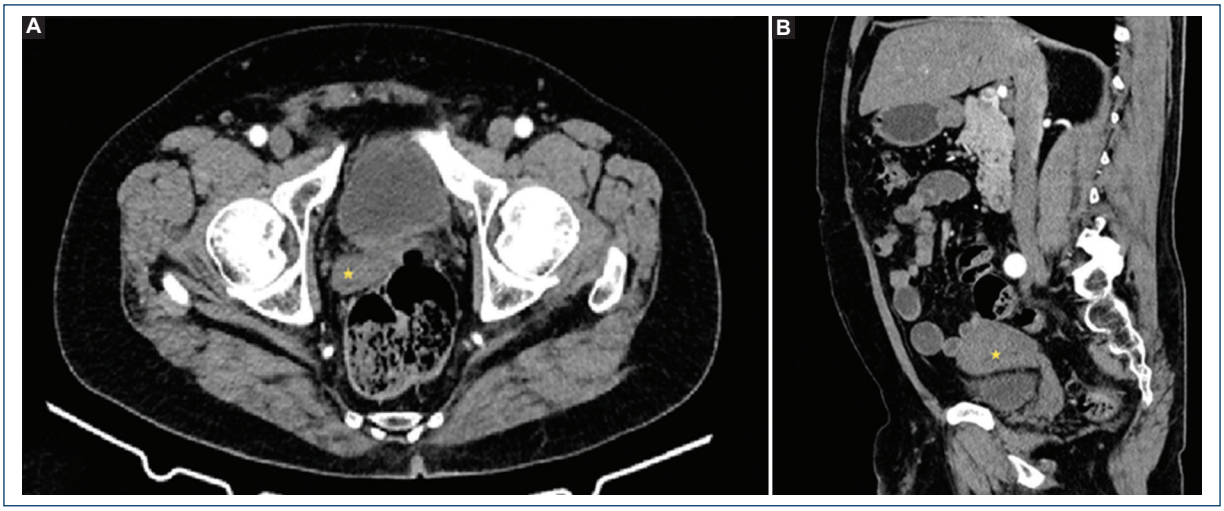

Figure 2. Contrast abdominal computed tomography in arterial phase in axial and sagittal planes
A and **B**: respectively showing evidence of muscular soft tissue density mass (30 HU) in retrovesical space in contact with pelvic structures (yellow star).

inguinal hernioplasty and whose intraoperative findings described the presence of macroscopically Müllerian structures such as uterus, fallopian tube and ovaries running inside the operated inguinal hernial sac.

Case report

We present the case of a 64-year-old male patient, infertile, with a long-standing right inguinoscrotal hernia. He had a soft tissue ultrasound where an inguinal and umbilical hernial defect was documented with no additional findings. The patient also presented a diagnosis of right homolateral cryptorchidism and nodular images in the right testicle with normal Doppler analysis. A right inguinal hernioplasty was scheduled on an outpatient basis. The surgical approach was performed through an oblique inguinotomy incision. An indirect inguinoscrotal hernia with sliding of contents into the hernia sac compatible with structures macroscopically compatible with the uterus, fallopian tubes, and involuted ovaries was observed (Fig. 1).

Reduction of the contents of the inguinal hernial sac and hernioplasty with the placement of prosthetic mesh with the Lichtenstein technique was performed. In the post-operative period, the patient had no complications and underwent an axial tomography of the abdomen and total pelvis which confirmed the intraoperative findings described above (Fig. 2A and B).

Discussion

Persistent Müllerian duct syndrome (PMDS) is a very rare form of pseudohermaphroditism characterized by the presence of structures deriving from persistent Müllerian duct in a patient phenotypically and occasionally genotypically expressed as a normal male[3-5]. It was initially described by Nilson in 1939, and since then approximately 150 related cases have been published[6,7]. To have an appropriate male differentiation, the production of anti-Müllerian Hormone is needed, which induces the regression of the Müllerian structures, allowing the differentiation of the Wolf's ducts and the formation of the vas deferens, epididymis, and seminal vesicles, which together with testosterone allow the adequate organization of the male reproductive system[8,9]. Its etiology is not entirely clarified; however, it is related to a lack of synthesis and release of Müllerian inhibitory factor (MIF) or its receptor (MIFr). MIF is secreted by Sertoli cells in fetal tissue from the 7th gestational week and is responsible for the regression of the Müllerian ducts in male fetuses. These patients are usually of 46 XY karyotype and besides presenting infertility or cryptorchidism, they can be asymptomatic until they are taken for surgical intervention and accidentally realize the recognition of organs macroscopically similar to the uterus and fallopian tubes, exceptionally during an inguinal herniorrhaphy or an orchidopexy[10-12]. The presence of macroscopically uterine-like structures, with an ovary, in an inguinal hernial sac is due to the persistence of the paramesonephric duct in the male also called as PMDS type I[13-15]. In patients with true hermaphroditism, both ovarian and testicular tissue will be found in one or both gonads. In female patients classified as pseudohermaphrodites, the gonads are ovarian, but the reproductive

organ has a masculine tendency. Pseudohermaphroditism may not be identified until puberty and even, as in the current clinical case, in the context of an elderly patient with an active sexual life, sometimes even with preserved reproductive capacities[16]. Currently, there are reports of laparoscopic management of male pseudohermaphroditism in pediatric patients with exeresis of the Müllerian structures, justified by the risk of malignization and infertility[17,18]. In cases of concomitant cryptorchidism, a division of the intra-abdominal uterine tissue attached to the testicle is considered to allow an adequate downward trajectory into the tunica vaginalis and then the remaining Müllerian structures are resected by an anterior inguinotomy[19].

Conclusion

Müllerian duct persistence syndrome Type I is described as an extremely rare pathology that is generally diagnosed intraoperatively when performing orchidopexy or inguinal hernioplasty in patients with phenotypically male characteristics. Given that preoperative findings of physical examination and ultrasonography of the inguinal region, whose ability to discriminate the structures contained in the hernial sac is precarious, it is unlikely that a preoperative identification of the pathology is performed in the patient with silent intersex clinic[20]. In the current case, through outpatient programming of a right inguinal hernioplasty, intraoperative identification of the Müllerian structures, which were not excised in the first surgical stage, was carried out. The defect of the indirect inguinal hernia was repaired by inserting a prosthetic mesh through the Lichtenstein technique without complications and was sent to complement studies with the genetics and endocrinology service, as well as to follow-up controls by the general surgery service. We consider of great importance to take into account that, in spite of being a very rare pathology, it can mean a therapeutic challenge for the multidisciplinary group in charge during the performance of an inguinal hernioplasty in men.

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Conflicts of interest

The authors declare no conflicts of interest.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patients, and received approval from the Ethics Committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript.

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