Hernia Uteri Inguinale: Rare Incidental Finding in an Adult Male: Case Report

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Abstract: Hernia uteri inguinale a type 1 persistent mullerian duct syndrome (PMDS) usually an accidental finding either during orchiopexy or during routine inguinal hernia repair in male patients presenting with mal-descended testes. Persistent Mullerian duct syndrome (PMDS) is a rare form of internal male pseudohermaphroditism in which Mullerian duct derivatives are seen in phenotypically normal male patient. It is caused by a defect in the Mullerian Inhibiting Substance (MIS) system. We are presenting a rare case report of a male aged 68 year presented with left cryptorchidism and right inguinal hernia. Very few cases have been reported in the literature. In India, owing to neglect and lack of facilities, we encounter this condition in adult males. Though rare, every surgeon operating upon inguinal hernia or cryptorchidism needs to know about the presence of the uterus in a phenotypic male patient at any age. High degree of suspicion and awareness is needed to diagnose this condition. Early treatment is needed to maintain fertility and to prevent the occurrence of malignancy in remnant müllerian structures.

Keywords: hernia uteri inguinale, cryptorchidism, persistent mullerian duct syndrome

1. Introduction

Intersex management is a medico-legal & social issue. Management in these cases is for functional goals or to prevent degenerative and malignant changes in vestigial remnants of gonads or internal primitive ducts[PMDS 3]. Inguinal hernia repair is one of the commonest operation in surgical practice. However, when a surgeon encounters unusual content it could pose a difficulty in its management [1,2]. The often encountered unusual contents are appendix, ovary, fallopian tubes and urinary bladder [1,2]. In this study we aim to present our experience of unusual content & approach to its management. Hernia uteri inguinalis is first described by Nilson in 1939[3]. It is an autosomal recessive (AR) disorder seen in genetically and phenotypically male subjects who develop female internal organs (uterus and fallopian tubes) due to a deficiency in the anti-Müllerian hormone (AMH) produced by Sertoli cells, or its type II receptor (AMHR-II)[4]. There is no ambiguity or malformation of the external genitalia in this condition. Familial cases have been reported with a probability of sex-limited autosomal recessive or X-linked recessive inheritance. An incidence of PMDS in identical twins has also been reported. Knowledge is essential to diagnose this rare entity thus a case report.

2. Case Report

A 68-year-old man presented to us with a right-sided inguinal hernia of six months duration with no other significant complaints. On examination his secondary sexual characters were well developed. The patient had normal masculine features like moustache, beard, pubic and axillary hair. His urethra and penis were fully developed with a well developed scrotum. Left testis was absent and right testis was palpable in the right hernial sac. He was married with no sexual dysfunction, but had no children. Patient was posted for elective hernioplasty. Subsequent under spinal anaesthesia right inguinal incision was taken, when the inguinal canal was opened and the scrotal content was retracted out, to our surprise we found uterus with fallopian tubes and testis. All mullerian duct structure removed with right sided orchidectomy. Hernioplasty was carried out and incision was closed in layers. Left sided orchidopexy done.

Figure 1A: Hernial content as uterus, fallopian Tube, small intestine and testis.

Figure 1B: Showing uterus and cervix? Vas deference clamped.
Surgical specimen sent for pathological examination. Intraoperative blood was drawn for hormonal assay. Patient recovered well. There is need for regular follow up because of the possibility of malignancy was explained to the patient, who is on regular follow up examination.

Histopathology Findings:
Gross findings: Specimen consists of uterus measuring 6×6×4.5 cm. On opening endometrium was 0.1 cm & myometrium was 2.5 cm seen. An oval firm structure resembling testis is seen at corner of uterus with tube like structure of 5 cm long. Cut surface is tan yellow like testis. A vaginal / cervix like tubular structure was 4 cm long seen. Entire structure is covered in gray membranous like tissue. Representative tissue is submitted for processing.

Microscopic examination:
Section A shows Testis: Seminiferous tubules show interstitial fibroses, and marked with Leydig cell hyperplasia. No evidence of intraepithelial germ cell neoplasm. (ref: Fig 2A)

Section B shows epididymis.

Section C, D, E, G shows fibromuscular tissue.

Section F shows endometrium: with cystically dilated glands and myometrium (Fig 2B)

3. Discussion

In human fetus the Mullerian and Wolffian ducts are both present at 7 weeks of gestation. In a male fetus, the testis differentiates by the end of the 7th gestational week. Normal sex differentiation is controlled by testosterone, dihydrotestosterone (DHT) and MIF (Mullerian inhibitory factor). Sertoli cells secrete MIF, which leads to regression of the Mullerian ducts. Testosterone has a direct effect on differentiation of the Wolffian ducts, and promotes their differentiation into the epididymis, vas deferens, and seminal vesicles[5]. DHT induces male differentiation of external genitalia. PMDS patients have both Wolffian and Mullerian duct structures due to deficiency of MIF. Because both the Wolffian and Mullerian ducts begin to develop, the tissues are often intertwined, resulting in obstruction or non-patency of the vas deferens or other parts of the reproductive excretery ducts [5]. This can result in infertility, the most serious potential problem caused by this condition. Cryptorchidism occurs suggesting a role of MIF in transabdominal testicular descent, perhaps by facilitating contraction of the gubernaculum. The typical patient with PMDS has unilateral or bilateral cryptorchidism and is assigned to the male sex at birth, as they have normal male genotypes and phenotypes [6]. Two anatomic variants of PMDS have been described: male and female. The male form is encountered in 80% to 90% of cases, characterized by unilateral cryptorchidism with contralateral inguinal hernia, and can be one of two types: the first type is hernia uteri inguinalis, which is characterized by one descended testis and herniation of the ipsilateral corner of uterus and fallopian tube into the inguinal canal. The second type is crossed testicular ectopia, which is characterized by herniation of both testes and the entire uterus with both fallopian tubes[7]. The female form, seen in 10% to 20% of cases, is characterized by bilateral cryptorchidism. The gonads are fixed within the pelvis, with the testes fixed within the round ligament in the ovarian position with respect to the uterus[5]. Clinically, the persistence of a uterus and fallopian tubes leads to either cryptorchidism or inguinal hernia depending on whether or not Mullerian derivatives can be mobilized during testicular descent[6]. PMDS is usually coincidently detected during surgical operation, as in our case. However pre-operative ultrasonography, computerized tomography and MRI allows for possible preoperative diagnosis [6]. The risk of malignancy in an ectopic testis in a case of PMDS is similar to that in a cryptorchid testis in a healthy male [7]. The overall incidence of malignant transformation in these testes being 18%[8]. Germ cell tumors have been reported in the testis, whereas tumors of the Mullerian duct derivatives are very rare[9]. Infertility is common, spermatozoa being absent on semen analysis [5]. Another disorders of sex development with male phenotype includes mixed gonadal dysgenesis (MGD), dysgenetic male pseudohermaphrodite (DMP) & ovo-testicular syndrome (true hermaphrodite). MGD have variable Mullerian duct structure along with one streak and other dysgenetic testicle, small size phallus with
hypospadias. DMP is a short stature mental retarded, aniridic male with abnormal genitalia having combination of immature hypoplastic testicular tubules & ovarian like stroma without ovarian follicles. The true hermaphrodite can have male or female phenotype, internal genitalia being ovary and or testis with mullerian duct structure showing ambiguous external genitalia. The main therapeutic considerations are the potential for fertility and prevention of malignant change. Surgical management is aimed towards preserving fertility. Early Orchidopexy with removal of mullerian structures is the preferred mode of management. Routine orchiectomy is indicated for testes that cannot be mobilized to a palpable position[10]. Despite the risk of malignancy and no chance of fertility, we preserved the right testis to maintain virilization. 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.

4. Conflict of interest

There is no conflict of interest.

References