International Journal of Allied Medical Sciences and Clinical Research (IJAMSCR)

ISSN:2347-6567

IJAMSCR |Volume 5 | Issue 4 | Oct - Dec - 2017
www.ijamscr.com

Case Report *Medical research*

An interesting case of inguinal hernia

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ABSTRACT

Persistent Mullerian duct syndrome is a rare form of male pseudo-hermaphroditism characterized by the presence of Mullerian duct structures in a 46XY phenotypic male. We report a case of a 40 year old male patient clinically presenting with left sided inguinal hernia. During herniorraphy, the hernial sac was found to contain uterus and fallopian tube.

INTRODUCTION

Persistent Mullerian Duct Syndrome (PMDS) is a rare form of internal male pseudo-hermaphroditism. The first case was described by Nilson in 1939 [1]. Around 150 cases have been reported in literature till date [2]. It is characterized by the presence of Mullerian duct structures in a 46XY phenotypic male [3]. It is thought to result from the defect of the synthesis or release of Mullerian inhibiting factor (MIF) or from a MIF receptor defect [2]. The persistence of a large uterus-like paramesonephric duct in a man is in itself clinically unusual, but when it forms a part of the contents of a hernial sac, it must be considered a rarity [4]. Hernia uteri inguinalis is one of the rare causes of male pseudo-hermaphroditism [5]. We report a case of a 40 year old male patient with left sided inguinal hernia containing a uterus and fallopian tube. It was detected coincidentally during herniorraphy.

CASE REPORT

A 40 year old male patient presented with a history of left inguino-scrotal swelling since 10 years. The patient was married and had one child. He had average muscular built and well developed secondary sexual characteristics. On local examination, a non-reducible, tense swelling was present in the left inguino-scrotal region approximately 9x7 cm in size. A cough impulse was absent. . The left testis was found to be of normal size in the left scrotum. The right scrotum was empty. Other routine investigations were within normal limits.

On surgical exploration, inguinal canal revealed an indirect inguinal hernia containing uterus with a fallopian tube and adherent right testis. Total excision of the uterus with fallopian tube and right testis was performed and the operation was completed with left inguinal herniorraphy.

On gross examination, the uterus measured 6.5 x 3.8 x 3 cm. The adherent testis measured 3 x 1.5 x 1.5 cm and the fallopian tube measured 4.5 x 0.5 cm (Figure 1).

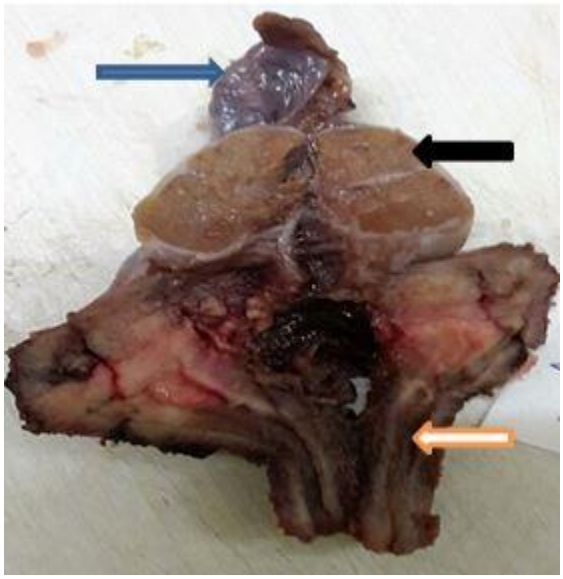

Figure 1: White arrow shows uterus, black arrow shows right testis and blue arrow shows fallopian tube.

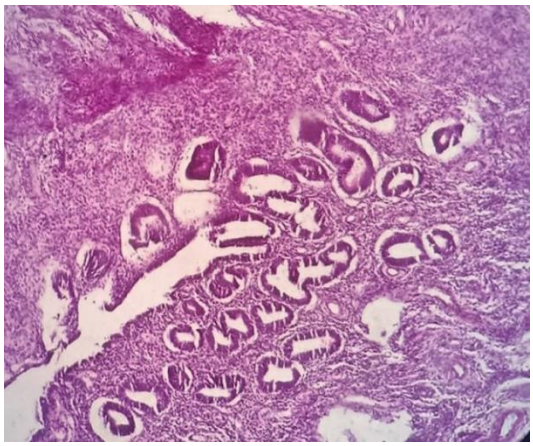

Figure 2: Photomicrograph showing endometrial glands and stroma. (H & E, X40)

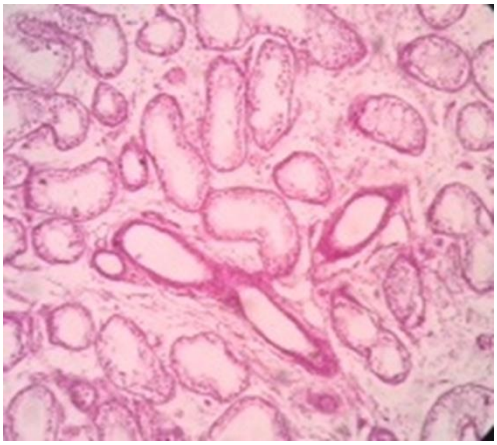

Figure 3: Photomicrograph showing seminiferous tubules with arrest of spermatogenesis. (H & E, X40)

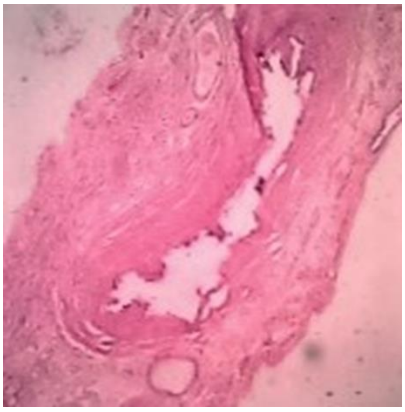

Figure 4: Photomicrograph showing fallopian tube histology (H & E, X10)

Microscopic examination: sections from the uterus revealed thinned out endometrium composed of endometrial glands and stroma. Glands were round to oval and lined by pseudo stratified columnar epithelium. Stroma was compact and composed of spindle cells (Figure 2). Adjacent myometrium showed many thick walled congested blood vessels. Sections from the testis revealed seminiferous tubules with thickened basement membrane. Spermatogenesis was absent. Few sertoli cells and interstitial leydig cells were present (Figure 3). Sections from the fallopian tube revealed mucosal plicae thrown into folds. Many congested and dilated blood vessels were also seen (Figure 4).

The final histopathological diagnosis of the hernial sac contents was consistent with that of a uterus with fallopian tube with the testis showing complete arrest of spermatogenesis.

Post operative ultrasonography of scrotum and pelvis showed an empty scrotum on right side, normal sized testis on left side. There was no evidence of ovaries.

DISCUSSION

Persistent Mullerian Duct Syndrome (PMDS) is a rare form of internal male pseudo-hermaphroditism. The first case was described by Nilson in 1939 [1]. It is characterized by the presence of mullerian duct structures in 46 XY phenotypic males. It is thought to result from a defect of the synthesis or release of Mullerian inhibiting factor (MIF), or from defects in the MIF receptor. MIF, also known as anti- Mullerian hormone, is released by the sertoli cells at around seven weeks of gestation. The gene for MIF is located on chromosome 19. MIF gene defects lead to the persistence of a uterus and fallopian tube in males. The remnant Mullerian structures might lead to cryptorchidism thwarting the normal testicular descent mechanism [2].

Patients with PMDS usually have normal development of external genitalia and secondary sexual characteristics [5]. In our case, patient had masculine external genitalia with fully developed uterus.

There are two anatomic variants of PMDS: male and female form. The male form, which is seen in 80-90% cases, is further classified as hernia uteri inguinalis and crossed testicular ectopia. Hernia uteri inguinalis is more common and characterized by one descended testis and herniation of the ipsilateral corner of uterus and fallopian tube into the inguinal canal. The crossed testicular ectopia, is characterized by herniation of both testes and the entire uterus with both fallopian tubes [5]. The female form is seen in 10% to 20% of cases. It is characterized by bilateral cryptorchidism.

The clinical presentation of PMDS can be either in the form of cryptorchidism or inguinal hernia depending on whether or not Mullerian derivatives can be mobilized during testicular descent [3]. During testicular descent, the uterus and fallopian tube may descend into the inguinal canal if they are mobile. The relative immobility of the Mullerian structures may hinder the testicular descent [10, 11].

PMDS is usually diagnosed incidentally during surgical exploration for cryptorchidism or herniorraphy (as in our case) as the mullerian remnants are not palpable on abdominal, rectal or

scrotal examination. Intraoperative gonadal biopsy can be performed to rule out mixed gonadal dysgenesis and developing malignancy [6, 7]. Ultrasonography, computerized tomography and MRI prior to surgery help in diagnosis of PMDS [3].

Patients with PMDS are at an increased risk for malignant transformation of gonads, similar to other undescended testes. Embryonal carcinoma, seminoma, yolk sac tumor, and teratomas have been reported in patients having PMDS [2]. The overall incidence of malignant change is 15% [8].

PMDS is important to diagnose as there is potential for fertility and malignant transformation can be prevented. Surgical management for preserving fertility is orchiopexy for repositioning of testis into the scrotum, herniorrhaphy with hysterectomy, and bilateral salpingectomy. Vas deferens is retained to preserve fertility. The main indications for orchidectomy are malignant transformation and testes that cannot be mobilised [9]. Routine orchidectomy is not recommended as it leads to loss of virilization [2]. The risk of developing malignancy is greater in an abdominal localization than in an inguinal testis [3]. Therefore, in our patient's case, the right testis was removed and the left testis was protected to maintain virilization.

CONCLUSION

Even though inguinal hernias are common clinical presentations, they may sometimes have interesting contents on surgical exploration as was seen in our case of Persistent Mullerian Duct Syndrome.

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How to cite this article: Dr. Shubhangi Kachore, Dr. Anne Wilkinson, Dr. Sadhana Mahore, Dr. Vidula Gowardhan. An interesting case of inguinal hernia. Int J of Allied Med Sci and Clin Res 2017; 5(4): 883-886.
Source of Support: Nil. **Conflict of Interest:** None declared.