CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 5 (2014) 1285–1287

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

# Hysterectomy in a male? A rare case report

Afak Yusuf Sherwani [a,*], Abdul Qayoom Shah [a], Abdul Majeed Wani [a], Ahmad Chalkoo Bashir [b], Ahmad Khan Bashir [b], Farooq Ahmad Sofi [a], Ashfaq Amin Wani [a], Wasim Lone [a], Ab Hamid Sherwani [b], Mehmood Rashid Sheikh [a], Raj Reshi Sharma [a]

[a] Department of General surgery, District Hospital Baramulla, Kashmir, India
[b] Department of Anaesthesiology, District Hospital Baramulla, Kashmir, India

ARTICLE INFO

*Article history:*
Received 28 January 2014
Received in revised form 6 July 2014
Accepted 7 October 2014
Available online 30 October 2014

*Keywords:*
Hysterectomy
Inguinal hernia
Male

ABSTRACT

*INTRODUCTION:* Persistent Mullerian duct syndrome is a rare form of male pseudo-hermaphroditism characterized by the presence of Mullerian duct structures in an otherwise phenotypically, as well as genotypically, normal man; only a few cases have been reported in the worldwide literature. A great variety of organs have been found in indirect inguinal hernial sacs.
*PRESENTATION OF CASE:* We report a case of 70 year old man, father of 4 children with unilateral cryptorchidism on the right side and left-sided obstructed inguinal hernia containing uterus and fallopian tube (that is, hernia uteri inguinalis; type I male form of persistent Mullerian duct syndrome) coincidentally detected during an operation for an obstructed left inguinal hernia.
*DISCUSSION:* PMDS is usually coincidently detected during surgical operation, as was in our case. However pre-operative ultrasonography, computerized tomography and MRI allow possible pre-operative diagnosis.[3]
*CONCLUSION:* In cases of unilateral or bilateral cryptorchidism associated with hernia, as in our patient's case, the possibility of PMDS should be kept in mind.


## 1. Introduction

Persistent Mullerian duct syndrome (PMDS) was first described by Nilson in 1939.[1] Subsequently, approximately 150 cases have been reported in the literature.[2]

PMDS is a rare form of male pseudo-hermaphroditism characterized by the presence of Mullerian duct structures in an otherwise phenotypically, as well as genotypically, normal man.[3] It is characterized by the persistence of the uterus, fallopian tubes and upper vagina in otherwise normally virilized boys. Despite the normal male genotype (46 XY) and the subsequent normal development of fetal testes, müllerian structures do not regress either due to absence of Müllerian Inhibiting Substance (MIS) or lack of response to it. 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] We report the case of a 70 year old man with unilateral cryptorchidism on the right side and left obstructed inguinal hernia containing uterus and fallopian tube (that is, hernia uteri inguinalis; type I male form of PMDS) coincidentally detected during an operation for an obstructed left inguinal hernia with right cryptorchidism.

## 2. Case report

A 70 year old man presented to our hospital with a painful left-sided inguinal swelling of one day duration. The patient gave history of asymptomatic left inguinal swelling from past 20 years and absence of the right testis since birth.

The patient was phenotypically male with normal secondary sexual characters. He had been married for fifty years and was having 4 children, the youngest one being 38 year old female. General physical examination revealed a man of sub-average built with well developed secondary sexual characters. His urethra and penis were fully developed with a poorly developed right hemi-scrotum and no palpable right testis in the scrotum or inguinal canal. The left hemiscrotum was well developed and left testis was palpable in scrotum.

There was a non-reducible, tender swelling measuring approximately $10 \times 8$ cm in the left inguinal region with absent cough impulse. Baseline investigations were normal. Patient was prepared for surgery after obtaining a written consent. Exploration of the inguinal canal revealed an indirect inguinal hernia containing a globular structure resembling uterus, fallopian tubes with an

* Corresponding author. Tel.: +91 01952234369.
*E-mail address:* afaksherwani@gmail.com (A.Y. Sherwani).

http://dx.doi.org/10.1016/j.ijscr.2014.10.020

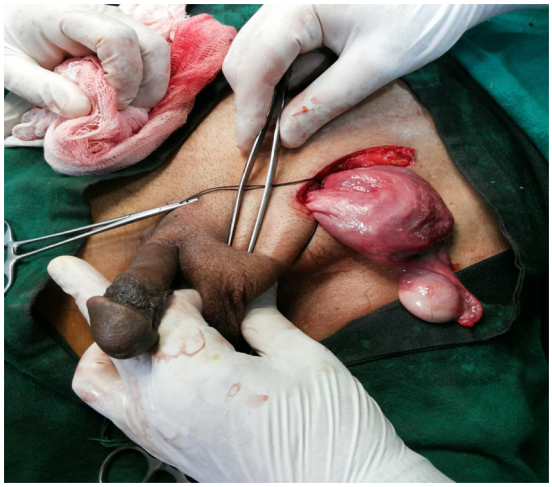

**Fig. 1.** Intra operative photograph showing uterus, fallopian tube and right testis in the broad ligament as contents of the inguinal hernia. Also seen in the picture are the left intra-scrotal testis phallus.

atrophic right testis embedded in the broad ligament and attached to pelvis with a thick fibrous band [Figs. 1 and 2]. Total excision of the uterus with fallopian tubes and atrophic right testis was performed and the operation was completed with left inguinal hernioplasty [Figs. 3 and 4]. Our patient had an uneventful post-operative period. Grossly, the specimens removed were identified as a uterus with patent endometrial and endocervical linings and two fallopian tubes. The right testis measuring $2 \times 1 \times 1$ cm, was atrophic and embedded in the right broad ligament. The specimen was sent for histopathological examination which revealed uterine muscular tissue with its cavity lined by endometrial tissue and congested fallopian tubes. No ovarian tissue was seen. Sections from right testes showed atrophic semineferous tubules, Sertoli cells and Leydig cells. No evidence of malignancy was seen in tissue samples from testicle. Post-operative karyotype analyses of the patient revealed 46, XY.

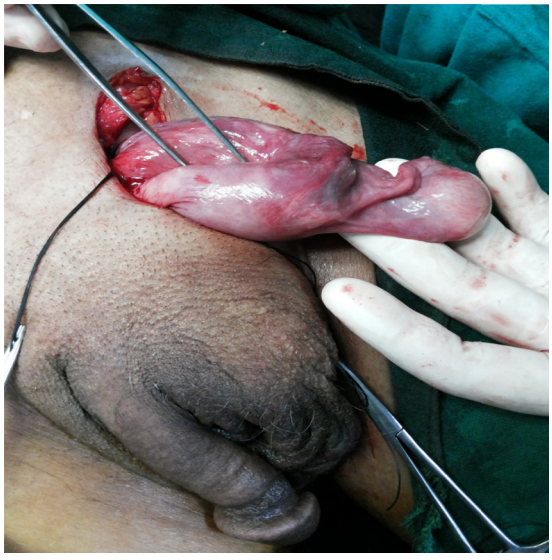

**Fig. 2.** Picture showing right testis in the broad ligament, fallopian tube and uterus extending toward midline.

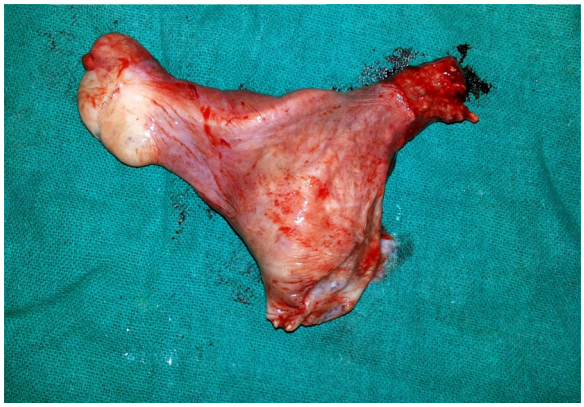

**Fig. 3.**

## 3. Discussion

Male pseudo-hermaphroditism is a condition in which the gonads are testes but the internal genitalia are not completely virilized. It is possible for pseudo-hermaphroditism to be undetected until puberty.[6] PMDS is a rare form of internal male pseudo-hermaphroditism in which Mullerian duct derivatives are seen in men. It was first described by Nilson in 1939.[1] Subsequently, approximately 150 cases have been reported. A familial association has been found in some cases.[2] The exact cause of PMDS is not known, however it is thought to result from a defect of the synthesis or release of MIF, or from defects in the MIF receptor. Defects in the MIF gene lead to the persistence of a uterus and fallopian tube in males. It is likely that remnant Mullerian structures lead to cryptorchidism by hindering the normal testicular descent mechanism.[2]

Patients with PMDS usually have normal development of external genitalia and secondary sexual characteristics.[5] The typical patient with PMDS has unilateral or bilateral cryptorchidism and is assigned to the male sex at birth without hesitation, as they have normal male genotypes and phenotypes.[2] Two anatomic variants of PMDS have been described: male and female. The male form is encountered in 80–90% of cases, characterized by unilateral cryptorchidism with contralateral inguinal hernia, and can be one of the 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.[5]

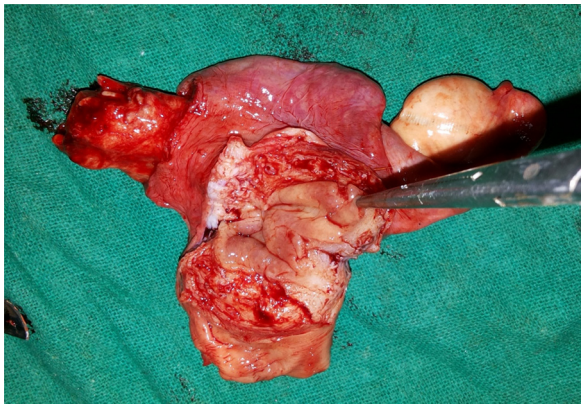

**Fig. 4.** Photograph showing the removed structures. Also seen is the opened uterine cavity with endometrial lining.

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.[3] If the uterus and fallopian tube are mobile, they may descend into the inguinal canal during testicular descent. However, if the Mullerian structures are relatively immobile testicular descent may be impeded.[5,7,8]

PMDS is usually coincidently detected during surgical operation, as in our patient's case. However pre-operative ultrasonography, computerized tomography and MRI allow possible pre-operative diagnosis.[3] Management of PMDS is controversial. An element of deciding on intervention is largely based on reducing risk of malignancy, while maintaining maximum reproductive function. In PMDS patients with undescended testes, the rate of testicular cancer is about 12% which is comparable to the rate seen in undescended testes not associated with PMDS.

Several authors have asserted that the mullerian structures should not be removed as there is minimal risk associated with their retention, and excision of the mullerian structures risks damaging primary blood supply to the PMDS testis via the internal spermatic and deferential arteries.[9] However most recent reports have demonstrated rare malignancies including adenocarcinomas of the mullerian duct associated with retained mullerian structures.[10,11]

## 4. Conclusion

PMDS is a rare form of male pseudo-hermaphroditism characterized by the presence of Mullerian duct structures in an otherwise phenotypically, as well as genotypically, normal man.

Since patients are phenotypically male, the diagnosis is usually not suspected until surgery is performed for cryptorchidism or hernia repair. Hernia uteri inguinalis is type I of the male form of PMDS, characterized by one descended testis and the herniation of the ipsilateral corner of the uterus and fallopian tube into the inguinal canal. In order to prevent further complications such as infertility and malignant change, the surgeon should be aware of PMDS while dealing with patients who present with unilateral or bilateral cryptorchidism.

In summary, in cases of unilateral or bilateral cryptorchidism associated with hernia, as in our patient's case, the possibility of PMDS should be kept in mind.

## Conflict of interest

No conflicts of interest.

## Funding

No source of funding. Purely a part of academic exercise in the hospital.

## Ethical approval

Ethical approval given by the Academic Committee District Hospital Baramulla. A written informed consent was obtained from the patient for publication of the case report and various images which may be used with the same. The patient being a major gave a written consent of the same after assuring him that name and hospital MRD no of the patient will not appear in the case report.

## Author contributions

All the authors have contributed either as being part of surgical and anaesthesia team or helping in the photography, paper writing, submission etc. Afak Yusuf Sherwani, Abdul Qayoom Shah, Abdul Majeed Wani, Farooq Ahmad Sofi and Ashfaq Amin Wani were in the surgical team. Bashir a chalkoo, Bashir a Khan and Ab Hamid Sherwani in the anaesthesia team. Wasim Lone, Mehmood a Sheikh and Raj Reshi Sharma did photography and paper writing.

## References

1. Nilson O. Hernia uteri inguinalis beim Manne. *Acta Chir Scand* 1939;**83**:231.
2. Prakash N, Khurana A, Narula B. Persistent Mullerian duct syndrome. *Indian J Pathol Microbiol* 2009;**52**:546–8.
3. Yuskel B, Saygun O, Hengirmen S. Persistent Mullerian duct syndrome associated with irreducible inguinal hernia, bilateral cryptorchidism and testicular neoplasia: a case report. *Acta Chir Belg* 2006;**106**:119–20.
4. Kane GJ. Inguinal hernia containing a paramesonephric (Mullerian) duct in an adult male: a case report. *S Afr Med J* 1978;**53**:639.
5. Renu D, Rao BG, Ranganath K. Namitha persistent Mullerian duct syndrome. *Indian J Radiol Imaging* 2010;**20**:72–4.
6. Michiels I, Peperstraete L, De Wever I, Gruwez JA. Inguinal hernia repair leading to the diagnosis of internal male pseudohermaphroditism. *Acta Chir Belg* 1984;**84**:255–8.
7. Dekker HM, de Jong IJ, Sanders J, Wolf RF. Persistent Mullerian duct syndrome. *Radiographics* 2003;**23**:309–13.
8. Clemente A, Macchi V, Berretta M, Morra A. Female form of persistent Mullerian duct syndrome – MDCT findings. *Clin Imaging* 2008;**32**:314–7.
9. Vandersteen DR, Chaumeton AK, Ireland K, et al. Surgical management of persistent mullerian duct syndrome. *Urology* 1997;**49**:941–5.
10. Elnemr A, Yonemura Y, Shinbo M, et al. Primary retroperitoneal Mullerian adenocarcinoma. *Rare Tumors* 2010;**2**(e6):16–9.
11. Warmann SW, Vogel M, Wehrmann M, et al. Gaint Mullerian duct cyst with malignant transformation in 15 year old boy. *Urology* 2006;**67**:424.