

CASE REPORT

PERSISTANT MULLERIAN DUCT SYNDROME IN AN ADULT MALE DIAGNOSED DURING HERNIORAPHY

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ABSTRACT: Persistent Mullerian duct syndrome (PMDS) is a rare form of internal male pseudohermaphroditism in which Mullerian duct derivatives are seen in a genotypically as well as phenotypically normal male patient. Very few cases have been reported in the literature. PMDS is likely to be encountered during surgery for undescended testis and inguinal hernia. Thus awareness of this disorder and the options of surgical management are necessary. We report a case of PMDS in a 35 year-old-male with left cryptorchidism and right inguinal hernia with uterus as content.

KEYWORDS: cryptorchidism, inguinal hernia, persistent Mullerian duct syndrome, undescended testis.

INTRODUCTION: PMDS classically known as hernia uteri inguinalis, is a rare disorder of sexual development (DSD), first described by Nilson in 1939.^[1] Not many cases have been reported till now. It is an autosomal recessive 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).^[2] There is no ambiguity or malformation of the external genitalia in this condition, and is most likely encountered during laparoscopy or surgery for hernia repair. Knowledge is essential to diagnose this rare entity thus a case report.

CASE REPORT: A 35-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, moustache, beard, pubic and axillary hair. His urethra and penis were fully developed with a poorly developed scrotum. Left testis was absent and right testis was palpable in the right hernial sac. All investigations were unremarkable. USG abdomen showed omental contents in the inguinal sac. He was married with no sexual dysfunction, but had no children. There was history of laparotomy 5 yrs back for abdominal mass that was reported as seminoma based on histology and patient had received radiotherapy for the same. No other significant past or family history was recorded. Patient was posted for elective hernioplasty, an indirect inguinal sac was identified with omentum as content along with two masses, (Figure 1 and 2) one was atrophic testis with two vas deferens and the other appeared as uterus. Though the testis was atrophic, a biopsy was taken and it was placed into the scrotum to sustain hormone secretion, other mass was excised. Hernioplasty was carried out and incision was closed in layers. The histopathologic evaluation of the excised mass revealed it to be uterus and testicular biopsy showed features of atrophy without any evidence of malignancy (Figure 3 and 4). Patient recovered well. The condition along with the problem of

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infertility and need for regular follow up because of the possibility of malignancy was explained to the patient, who is on regular follow up.

DISCUSSION: Embryologically, in a 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, and MIF. Sertoli cells secrete MIF, which leads to regression of the Mullerian ducts. Testosterone has a direct effect on the Wolffian ducts, and promotes their differentiation into the epididymis, vas deferens, and seminal vesicles.^[3] Dihydrotestosterone induces male differentiation of external genitalia. PMDS patients have both Wolffian and Mullerian duct structures. Due to a deficiency of MIF or MMH.

Because both the Wolffian ducts 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 excretory ducts. This can result in infertility, the most serious potential problem caused by this condition. Cryptorchidism occurs suggesting a role of AMH 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.^[4] 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.^[5] 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 coincidentally 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]

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 not recommended as the capacity of the virilization must be maintained and is only 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.

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CONCLUSION: This case is being reported to stress upon the possibility of PMDS-a rare entity in cases of unilateral or bilateral cryptorchidism associated with hernia, so that proper recognition and identification of the condition can be done especially in prepubertal age as fertility can be preserved and future risk of malignancy can be averted. Thus, during inguinal hernia repair, if unusual contents like uterus, ovary or Fallopian tube are encountered, we should keep the possibility of PMDS in the mind.

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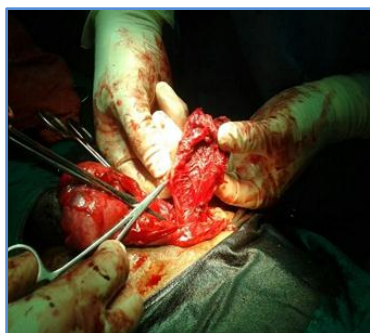


Fig. 1: Showing two masses in the hernia sac. uterine mass being pointed



Fig. 2: Testis with two vas deferens

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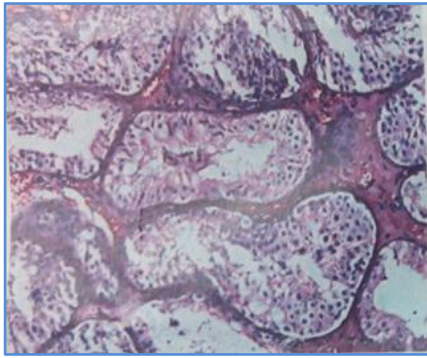


Fig. 3: H.P.E Report from biopsy from right testis showing signs of atrophy

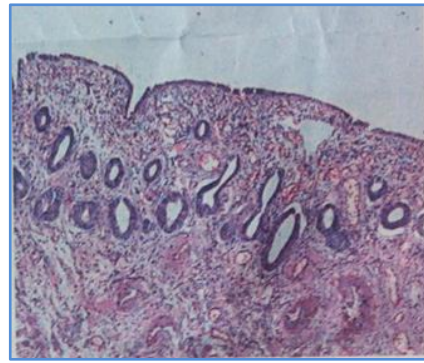


Fig. 4: H.P. E Picture of Uterine Mass

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