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Abstract : A 50 year-old male presenting with left sided obstructed inguinal hernia was found to have uterine tissue extending through the inguinal canal, warranting a diagnosis of persistent Mullerian duct syndrome (PMDS). PMDS is an extremely rare form of internal male pseudo-hermaphroditism in which female internal sex organs, including the uterus, fallopian tube, persist in a 46XY male with normal external genitalia. The condition results from a congenital insensitivity to anti-Mullerian hormone, or lack of anti-Mullerian hormone, leading to persistence of the female internal sex organs in a male. Patient taken up for emergency surgery.On exploration of scrotum revealed small bowel along with it rudimentary uterus and fallopian tube without ovary was found. It was excised , the operation was completed by left inguinal herniorraphy and its contents confirmed by histopathology report. Opposite scrotum and testis was found to be normal.

Keyword : Obstructed inguinal hernia, Hernia Uterus inguinalis, Mullerian duct syndrome

INTRODUCTION: Persistent Mullerian duct syndrome (PMDS) was first described by Nilson in 1939. Subsequently, only 150 cases have been reported in the worldwide literature. 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 male; The term “hernia uteri inguinalis” is used when the uterus is found in the hernia sac. We report the case of a 50-year-old man with left sided obstructed inguinal hernia containing uterus and fallopian tube (that is, hernia uteri inguinalis; type I male form of persistent Mullerian duct syndrome) detected during an operation for an obstructed left inguinal hernia.

CASE REPORT

50 year male patient presented with abdomen pain of 3 days duration. He gives history of swelling in the groin for one year, became irreducible since one month, pain in the abdomen and scrotum for three days, constipation since three days, vomiting since one day. Our patient had well developed masculine features and had three children. On examination patient was moderately built, and nourished, hydration poor, pulse 88/ min, BP was 110/70 mmHg. Local physical examination revealed a left-sided irreducible inguinal hernia. X-ray abdomen showed multiple air fluid levels. Patient was diagnosed as obstructed inguinal hernia and taken up for emergency surgery.

FIGURE: 1 XRAY ABDOMEN ERECT

Exploration of the inguinal canal and scrotum revealed small bowel, testis and rudimentary uterus with fallopian tube. Bowel found to be viable. Excision of the uterus with fallopian tube and left testis was performed. Herniorrhaphy done.

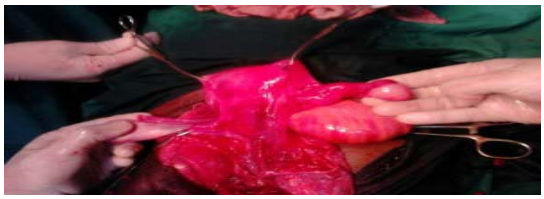

FIGURE:2 Intra-operative photograph showing redundent uterus with fallopian tubes, vas differens ,tesis, small bowel

FIGURE: 3 GROSS PICTURE OF SPCEIMEN

Histopathological examination of the excised specimen was consistent with that of a uterus and fallopian tubes without any evidence of ovaries. Testis was found to be atrophic. On histopathological examination, section from the solid area show fallopian tube and primitive uterine body with unicornuate. The lining epitheliam show tall columnar lining epitheliam. Uterine tissue lined by atrophic endometrial glands surrounded by edematous stroma. The cervical canal show poorly developed endo cervical glands. Testes showed atrophic semineferous tubules with focal hyperplastic leidig cell adjoining another area show thickened congested blood vessels with compact fibros stroma and epididymis and there is no evidence of malignancy.

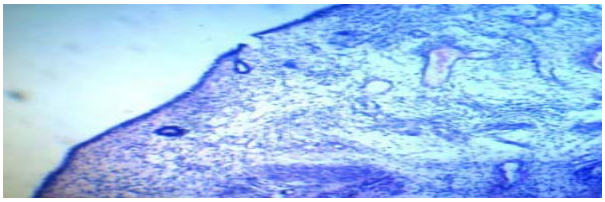

FIGURE:4 Photomicrograph showing uterine tissue lined by atrophic endometrial glands surrounded by edematous stroma

DISCUSSION:

Nelson first reported pmds in 1939 with 150 cases reported until now [1] .Inherited as an autosomal recessive condition. Incidence: 0 -1 per 100,000 live births. The exact cause of PMDS is not known. 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 7thgestational 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[2]. Dihydrotestosterone induces male differentiation of external genitalia. PMDS patients have both Wolffian and Mullerian duct structures due to a deficiency of MIF. Defects in the MIF gene lead to the persistence of a uterus and fallopian tube otherwise normally virilized XY male. According to Nelson's hypothesis, foetal testoterone is normal and the affected males are completely virilised.There is how ever a deficiency of antimullerian hormone with persistence of Mullerian ducts in the male fetus. Two anatomic variants of PMDS have been described: male and female. The most common variant is the male form is encountered 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 [3]. 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. 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 . 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[4]. PMDS is usually coincidently detected during surgical operation, as in our patient's case. However pre-operative ultrasonography, computerized tomography and MRI allow possible preoperative diagnosis[5] .

The prognosis depends upon the integrity of the testicular tissue and successful correction of cryptorchidism, which is often complicated by the close anatomical relationship between the vas deferens and the Mullerian derivatives .The risk of malignancy in an ectopic testis in a case of PMDS is similar to that in a healthy male,with the incidence being 15%[6]. There have been case reports of embryonal carcinoma, seminoma,yolk sac tumor and teratoma in patients with PMDS, whereas tumors of the Mullerian duct derivatives are very rare. Infertility is common, with an absence of spermatozoa observed during semen analysis [7] . The main therapeutic considerations are the potential for fertility and prevention of malignant change. Surgical management is geared towards preserving fertility, and orchiopexy, which is performed to retrieve the testis and position it in the scrotum, should be performed early to maintain fertility with care taken not to damage the vas deferens during the operation. The uterusis usually removed and attempts are made to dissect away Mullerian tissue from the vas deferens[8].

CONCLUSION:

PMDS is a rare form of male pseudo-hermaphroditism characterized by the presence of Mullerianduct structures in an otherwise phenotypically, as well as genotypically, normal male. The patient with PMDS has unilateral or bilateral cryptorchidism and is usually assigned to the male sex at birth without hesitation. Since patients are phenotypically male, the diagnosis is usually not suspected until surgery is performed for cryptorchidism or hernia repair. Our patent had type I male form of persistent Mullerian duct syndrome) coincidentally detected during an operation for an obstructed left inguinal hernia. 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

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