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The Sheep in the Wolfs Clothing-A Rare Case Report

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Abstract: ***Isolated Deficiency of Mullerian Inhibiting substance (MIS) is a rare is syndrome that usually does not present in the newborn period as the as genitilia genitilia appear to be those of a male with undescended testes. Its fascinating as the phenotypic findings are those expected in 46XY. The defect lies in complete failure of the testis to produce MIS in males. This report is of a 35 year old with bilateral undescended testis with left hernia diagnosed to be deficiency of MIS***

Keywords: hernia, azoospermia, mullerian inhibiting substance deficiency, pseudohermaphroditism

1. Introduction

MIS is essential for male sexual differentiation. In mammalian males, the fetal testes produce and secrete both MIS, which causes mullerian ducts to regress, and testosterone, which promotes the differentiation of wolffian ducts. Mullerian ducts, in the absence of MIS, continue to develop and differentiate as oviduct, uterus, cervix and upper part of vagina. Wolffian ducts, which give rise to male internal reproductive tract structures-epididymides, vas deferens and seminal vesicles degenerate without testosterone stimulation. Defects in either the gene for MIS or its receptor can result in a form of male pseudohermaphroditism characterized by retained mullerian duct derivates in a phenotypic male.

2. Case

History
A 35 year old male, presented with history of swelling and continuous dull aching pain in the left groin since 2 months, suggestive of left inguinal hernia. Right testis was absent since birth. No history of irreducibility or symptoms of obstruction

Examination
A moderately built and nourished middle aged male with an inguinoscrotal swelling in the left groin, which was reducible. Cough impulse was present, a small left testis was palpable at the superficial inguinal ring and the right testis was undescended with atrophy of right hemiscrotum. Tenderness was present in the left iliac region.

Investigations
Relevant laboratory investigations were unremarkable. Ultrasonography (USG) of abdomen and pelvis revealed a left iliac region mass suspicious of a malignancy (?Teratoma) ,left indirect inguinal hernia with Hypoplastic testis and omentum as its contents and right undescended intraabdominal testis(peeping testis)deep to deep inguinal ring. Semen analysis showed azoospermia.

Treatment
Patient was planned for left inguinal hernioplasty and excision of the left iliac mass with right sided orchidectomy.

3. Procedure

Left iliac incision placed and extended at the lower end transversely. The iliac mass was found to be an Omental mass with adhesions to the surrounding bowel which were released and mass excised. On exploration, the congenital hernia sac had hollow tubular structures apart from the vas deferens, which on tracing medially led to a Hypoplastic uterus. On further exploration mullerian duct derivates namely Hypoplastic uterus, bilateral fallopian tubes with fimbriae, bilateral Hypoplastic ovaries were present. Both the testes were hypoplastic. Hence the mullerian remanants were excised and bilateral orchidectomy with inguinal hernia repair was done.

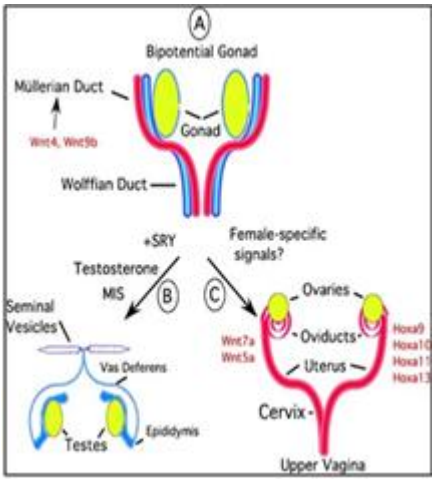


Figure 1: Normal Gonadal development

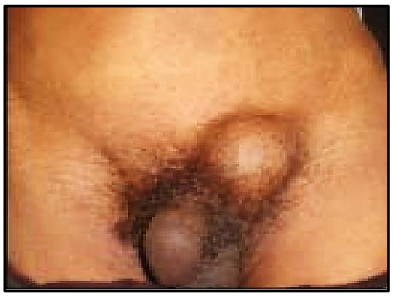


Figure 2: Left reducible inguinal hernia

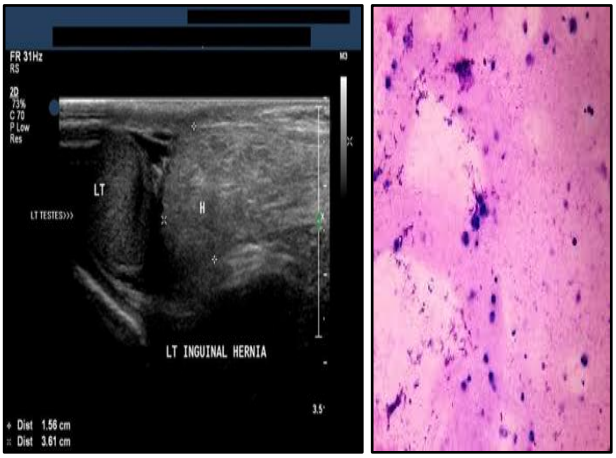


Figure 3: USG-? Teratoma with azospermia

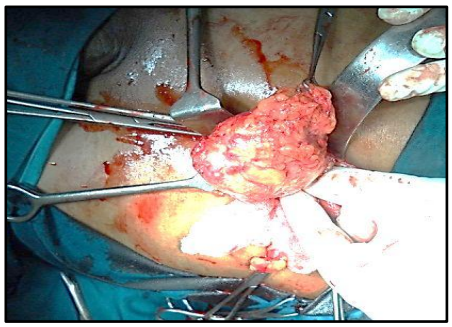


Figure 4: Omental mass

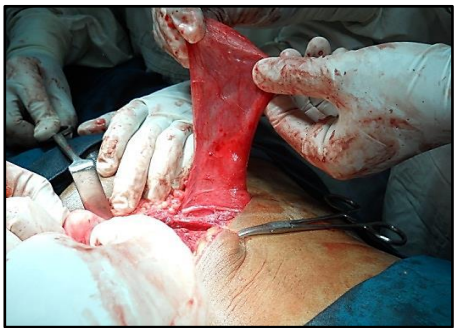


Figure 5: Hernial Sac

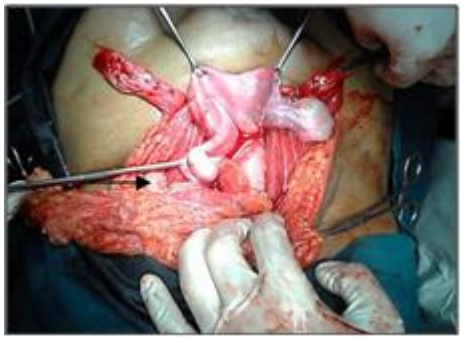


Figure 6: Hypoplastic Uterus

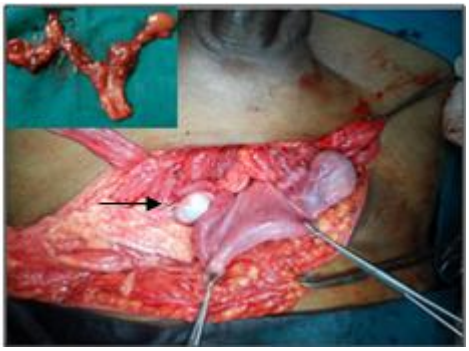
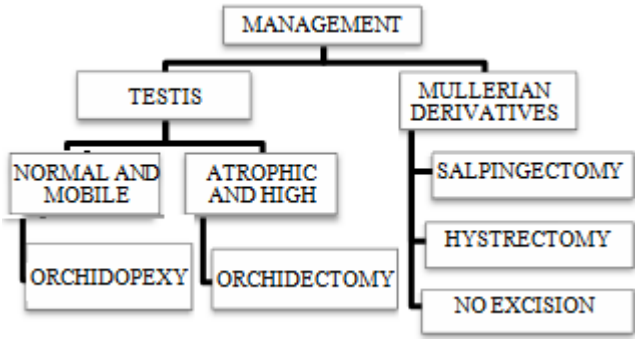


Figure 7: Hypoplastic Ovary and Excised specimen (Inset)

4. Discussion

MIS deficiency causes a rare form of internal male pseudohermaphroditism, in which uterus and fallopian tubes are present in a normal male, karyotype of 46XY. The proposed etiologies for persistent mullerian ducts are deficiency of MIS, abnormality in its receptor or failure to produce MIS before the 8th fetal week. Diagnosing persistent mullerian ducts (PMDS) is based on a combination of anatomic and clinical findings. Imaging features, although classic, are often missed. Ultrasound scan failed to identify the internal structures in our patient too. However, when availability and affordability are not an issue, computed tomography (CT) and magnetic resonance imaging (MRI) are known to show the tubular structures clearly. Ideally, this diagnosis should be complemented by karyotyping, for which we lacked the requisite facility. The surgical management of PMDS is still controversial. If the testes are atrophic or cannot be brought down in orchidopexy, then orchidectomy alone should be performed. The risk of testicular malignancy is similar for PMD and cryptorchidism, at 5-18%. For the PMD few studies advocate bilateral proximal salpingectomies, leaving fimbriae with the epididymides with corporal hysterectomy whereas few however found no absolute indication for the removal of the Mullerian duct structures, other than where they limit testicular mobility during orchidopexy, because no malignant degeneration of the retained Mullerian structures in PMDS has ever been documented.



5. Conclusion

Persistent mullerian duct structures are a rare form of male pseudohermaphroditism often encountered unexpectedly at surgery for cryptorchidism or inguinal hernia. It does not present in the new born period during which the phenotypic findings are of a normal male with genetic being that of a gonadal male with complete failure to produce MIS.As it is seen exclusively in males the inheritance is X-linked recessive or autosomal dominant. It is therefore important to maintain a high index of suspicion of the above and to perform necessary imaging and karyotyping in such cases. Genetic counseling should be offered to parents of affected patients and long-term follow-up is necessary.

References

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