Persistent Mullarian Duct Syndrome – Hernia Uterine Inguinalis: A Case Report.

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Abstract

Persistent mullerian duct syndrome (PMDS) is an extremely rare form of internal male pseudohermaphroditism in which female internal sex organs are present in a male with normal 46XY karyotype and normal external genitalia. This condition was first reported in 1895. We present a case of right groin swelling of 9 months duration and were clinically diagnosed as complete irreducible inguinal hernia with no palpable testis in the left hemiscrotum, both the testis palpable in the right hemiscrotum. After clinical diagnosis workup was done with routine investigations, ultrasonography and treated with right inguinal herniorraphy.

Introduction

PMDS is a very rare congenital anomaly in which mullerian duct derivatives- uterus and cervix are found in a genotypically and phenotypically normal male. It is usually associated with unilateral or bilateral cryptorchidism and infertility. PMDS is caused by mutation in AMH gene or mutation in AMH receptor.

Case Report:

A 45yr old male married 25yrs back with one live male child presented with a right sided groin swelling of 9 months duration associated with dragging type of pain, poorly developed left hemiscrotum with no testis. Both testes are palpable in right hemiscrotum. The swelling was pyriform in shape and was not reducible. All the investigations are within normal limits. Karyotyping revealed 46XY. After an informed consent the patient was taken up for surgery and the hernial sac contents revealed a mass resembling uterus like structure with a tubular structure on either side resembling fallopian tubes. White firm, irregular mass measuring 7x6x3.5cm and 6.8x5.4x3.2cm were attached to the right and left sides respectively.

On pulling the cord structures normally appearing both testes were delivered from the right hemiscrotum. The spermatic cords were separated. Uterus and fallopian tubes were excised and both the testis were fixed to the right hemiscrotum followed by right inguinal herniorrhaphy. Histopathological examination of the excised specimen revealed the presence of mullerian duct structures- uterus, fallopian tubes and cervix.

Specimen excised was identified as: Uterus, Fallopian tubes and cervix.
CONCLUSION:

PMDS is a very rare congenital anomaly in which mullerian duct derivatives—uterus and cervix—are found in a genotypically and phenotypically normal male. It is usually associated with unilateral or bilateral cryptorchidism and infertility.

PMDS is usually caused by deficiency of fetal anti-mullerian hormone (AMH) effect due to mutations of the gene for AMH or anti-mullerian hormone receptor. Resection of mullerian duct derivatives followed by repair of the hernia is the recommended treatment for Hernia uterine inguinalis.

REFERENCES:


DISCUSSION:

PDMS is a rare syndrome characterized by the presence of mullerian duct structures—uterus, fallopian tubes, and cervix—in a male with normal genotype and phenotype. Only about 200 cases have been reported so far. The present case of PDMS manifested as an inguinal hernia with unilateral cryptorchidism but with no infertility. The patient has an 8-year-old normal male child. This is a rare occurrence in PDMS. Transverse testicular ectopia (TTE) is an anatomical abnormality where both the gonads migrate towards the same hemiscrotum. TTE is the rarest form of testicular ectopia with fewer than 100 cases in the literature.