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Case Report

Persistant Mullerian Duct Syndrome with Transverse Testicular Ectopia Presenting as Left Inguinal Hernia with Cryptorchidism, A Rare Presentation

Authors
Dr Javid Iqbal* MS, FMAS, FIAGES[1], Dr Yasmeen Kousar MBBS, DGO[2], Dr Sonam Gupta MBBS[3]
[1]Lecturer, Dept of General Surgery, Govt Medical College Jammu, Jammu and Kashmir, India 180005
[2]Assistant Surgeon, J & k Health Services, Jammu, Jammu and Kashmir, India
[3]Resident, Department of Surgery, Government Medical College Jammu, Jammu and Kashmir, India
*Corresponding Author
Dr Javid Iqbal
Lecturer, Dept of General Surgery, Govt Medical College Jammu, Jammu and Kashmir, India 180005
Phone: 9596620409 Email: *javidiqbal123@gmail.com*

Abstract
Inguinal hernia and undescended testes in a patient can present with transverse testicular ectopia (TTE) and very rarely, with residual Mullerian duct structures. This is called persistent Mullerian duct syndrome (PMDS). PMDS is mostly discovered during surgery for inguinal hernia or cryptorchidism. TTE is a rare congenital anomaly in which both testes descend through a single inguinal canal. Patients with TTE present with symptoms of cryptorchidism and inguinal hernia. Herein, we reported one TTE case with PMDS. For patients with inguinal hernia and cryptorchidism associated with TTE, PMDS should be kept in mind and radiologic evaluation with ultrasonography or magnetic resonance imaging of the genitourinary system and karyotyping should be done.
Keywords: *Orchiopexy, persistent Mullerian duct syndrome, transverse testicular ectopia, inguinal hernia.*

Introduction

Persistent Mullerian duct syndrome (PMDS) is a rare autosomal recessive disorder which could present as male pseudohermaphroditism in which Mullerian duct derivatives are seen in a male patient. This syndrome is characterized by the persistence of Mullerian duct derivatives (i.e. uterus, cervix, fallopian tubes and upper two thirds of vagina) in a phenotypically and karyotypically male patient. This syndrome is caused either by an insufficient amount of Mullerian inhibiting factor (MIF) or due to insensitivity of the target organ to MIF.

Case Report

A two year male child presented with left inguinal swelling. On examination there was a swelling in the left inguinal region, cough impulse was

positive, bilateral hemiscrotum was empty but well developed and phallus was also well developed. There was no other clinical abnormality. With the provisional diagnosis of left inguinal hernia with bilateral undecended testes, patient underwent Usg abdomen and scrotum : left testis was found in inguinal canal , right testis was found just cephaloid to left deep inguinal ring, both the testes was comparable in size and eco texture. Patient was adviced MRI abdomen but could not be done due to patient financial problem, rest of his hematological investigations were within normal limit. Patient was taken up for surgery, intra operatively it was noticed that the hernia sac contains an underdeveloped uterus , bilateral fallopian tubes with fimbria like structures and there were two gonadal structures, one along each fallopian tube, which appeared ovoid with smooth surface like testes [figure 1] . Bilateral vas deferentia were located on either side of rudimentary uterus. Bilateral fallopian tubes and uterus was dissected [figure 2] upto a point where bilateral vas deferentia were in close proximity to each other. Herniatomy and bilateral orchiopexy was done [figure 3]. Biopsy revealed fallopian tubes [figure 4] and rudimentary uterus. Karyotyping was done which revealed 46XY chromosomes, Post operative serum testosterone level was normal.

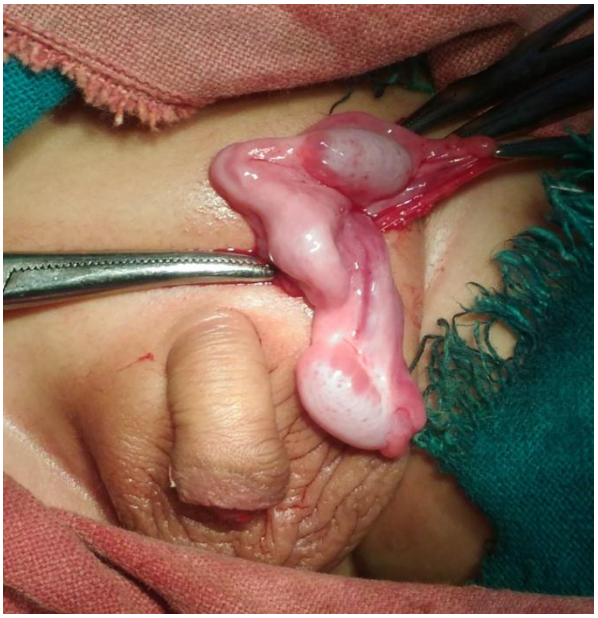

Figure 1. Intraoperative photograph showing rudementary uterus, fallopian tubes and testis.

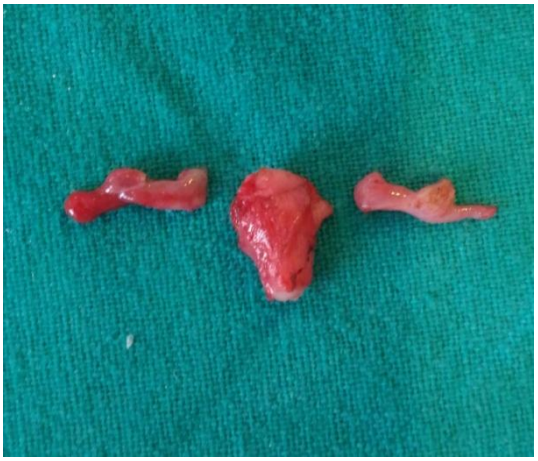

Figure 2. Resected speciman showing rudemantary uterus and fallopian tubes

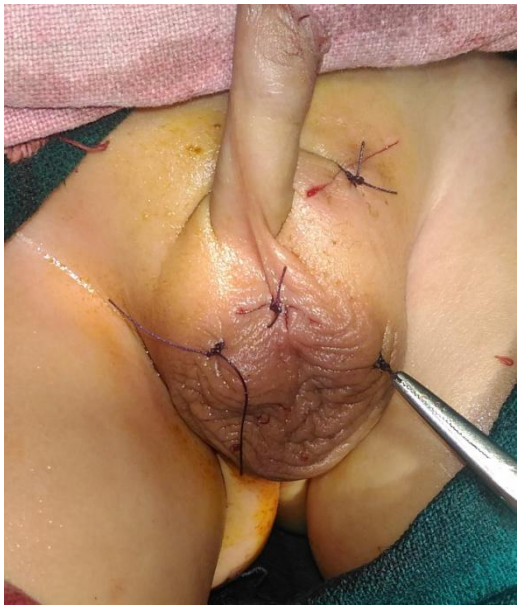

Figure 3. Post operative photograph of patient

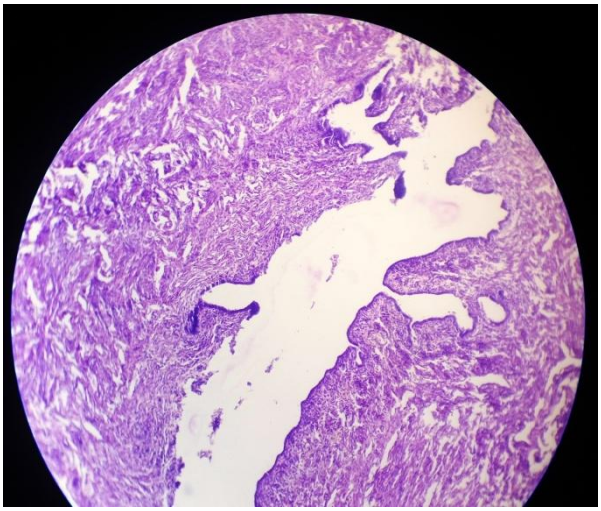

Figure 4. Photomicrograph of histology speciman showing fallopian tube.

Discussion

Nilson reported first PMDS in 1939.[1] Mullerian inhibiting substance (MIS), secreted by the Sertoli cells from seven weeks of gestation, causes the regression of the Mullerian duct in the male fetus.[2] PMDS which is caused by defects in the synthesis or action of Mullerian inhibiting factor and is characterized by karyotypically normal males with retained MD derivatives. Three groups of PMDS have been described. Group 1 (60-70%) female type: Bilateral intra-abdominal testes in a position analogous to ovaries. Group 2 (20-30%) male type: One testis is found in a hernia sac or scrotum in association with a contra lateral inguinal hernia with the uterus and tubes (hernia uteri inguinale). Group 3 (10%) male type: Both the testes are located in the same hernial sac along with the Mullerian structures (transverse or crossed testicular ectopia).[3,4,5,6] Our patient belongs to the rarest group, transverse testicular ectopia associated with PMDS, presenting with left inguinal hernia and bilateral undescended testes. Upon exploration bilateral testes along with uterus and fallopian tubes were present as contents of left sliding inguinal hernia. Transverse testicular ectopia (TTE) or crossed testicular ectopia is a rare form of testicular ectopia. It was first reported by Von Lenhossek in 1886.[2] TTE, also named testicular pseudo-duplication, is characterized by both testes descending through a single inguinal canal. The precise etiology of TTE is still unclear. Various anatomic factors (defective implantation, rupture or tearing of the gubernaculum, obstruction of the internal inguinal ring, development of adhesions between the testis and adjacent structures, late closure of the umbilical ring, etc.) are suggested as causative or inducible factors in failure of testicular descent.[7] Some of TTE cases are very rarely associated with PMDS as in our case. PMDS is mostly discovered during surgery for inguinal hernia or for cryptorchidism.[1] These testes are usually histologically normal in patients with PMDS with an overall incidence of malignant transformation around 18%, similar to the rate in abdominal testes in otherwise healthy men.[8] Since patients are phenotypically male, diagnosis is not usually suspected until surgery for cryptorchidism or hernia repair.[9]

The treatment of PMDS is relatively straight forward, all patients are phenotypic males who require orchiopexy by open or laparoscopy. Early orchiopexy with the removal of Mullerian structures is the preferred approach. Vasa deferentia lie near the uterus and proximal vagina. Therefore, preservation of necessary Mullerian structures to avoid injury to vasa deferentia is recommended to preserve fertility. Parents should be made aware of the risk of testicular malignancy and infertility.

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