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Transverse testicular ectopia: A surgical surprise

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Abstract

Crossed Testicular Ectopia / Transverse Testicular Ectopia is a rare but well known congenital anomaly in which both gonads migrate towards same Hemiscrotum. Most of the cases were found associated with other abnormality such as Persistent Mullerian Duct Syndrome, Inguinal hernia, Hermaphroditism and other scrotal abnormality. Most of the cases reported in literature are children with very few reports in adults. About 100 cases of Transverse Testicular Ectopia were reported in literature till now. We report a case of Transverse Testicular Ectopia in a 42 year old gentleman presented with partially reducible Left Inguinal Hernia with non-palpable Right Testis. On exploration, one testis with rudimentary Uterus and bilateral rudimentary Fallopian tubes were present in Left Inguinal Canal while another testis in Left Hemiscrotum. Castration with left hernioplasty done as the patients is adult.

Keywords: TTE (transverse testicular ectopia), PMDS (persistent Mullerian duct syndrome), hermaphroditism, crossed testicular ectopia

Introduction

Transverse Testicular Ectopia otherwise called as Crossed Testicular Ectopia/Testicular Pseudoduplication /unilateral double testis is a rare but a well-known entity where both the testis migrates towards the same hemiscrotum^[1]. In literature more than 100 cases were reported till date. Often diagnosis is made on surgical exploration. There are five known types of testicular ectopia: superficial inguinal (interstitial), femoral (crural), perineal, pubopenile, and crossed (transverse)^[2]. Transverse Testicular Ectopia can be suspected if there is ipsilateral inguinal hernia with contralateral undescended testis^[3]. The inguinal hernia invariably presents on the side where the ectopic testis has migrated^[4]. Associated abnormalities may include Persistent Mullerian Duct Syndrome (PMDS), true hermaphroditism, inguinal hernia, hypospadias, pseudohermaphroditism, defects of abdominal wall, and scrotal anomalies^[5]. The lack of awareness leads to delay in diagnosis and accidental detection during surgery for inguinal hernia. We report a case of Transverse Testicular Ectopia presented as left inguinal hernia.

Case Report

42 year old gentleman presented with left inguinal swelling of 6months duration. Swelling increases in size while lifting heavy objects. Patient is married for 15 years with primary infertility with normal sexual function. On examination, there is a globular swelling of size 6X5cm in the left inguinal region; swelling was partially reducible and expansile impulse on cough-present. There was a palpable testis in left hemiscrotum with absent testes in right hemiscrotum. The scrotum was normally developed. Secondary sexual characters were normal. Basic investigations were normal. Seminal Analysis showed azoospermia. Ultrasonography of scrotum showed both testicle on left side, one in the left inguinal canal and another in the left hemiscrotum. Ultrasonogram of abdomen showed no significant abnormality in abdomen and KUB area. A provisional diagnosis of Transverse Testicular Ectopia was made. On exploration, rudimentary uterus with bilateral rudimentary fallopian tubes and atrophic testis was present as hernia content. A diagnosis of Persistent Mullerian Duct Syndrome with Transverse Testicular Ectopia was made. B/L Orchidectomy was done and biopsy taken from mullerian remnant. Deep Ring was closed, Hernioplasty was done. Post-operative periods uneventful.



Figure description 1 and 2: showing rudimentary uterus with b/l rudimentary fallopian tube and b/l atrophied testis as hernial sac content

Discussion

Transverse Testicular Ectopia (TTE) is a rare form of testicular ectopia. TTE was first described by von Lenhossek in 1886 in a cadaver. [5] Holsted published the first case of testicular ectopia in English literature followed by 100 cases and more. Several theories regarding the embryogenesis of TTE have been postulated namely adhesion and fusion of developing Wolffian canals, aberrant gubernaculum, testicular adhesions, defective formation of the internal inguinal ring, and traction on a testis by persistent Mullerian structures [5]. Berg proposed that there is a possibility of the development of both testes from the same genital ridge. Gupta and Das [6] proposed that early adherence and fusion of the developing Wolffian ducts, the descent of one testis has caused the second one to follow [6]. Persistent Mullerian Duct Syndrome develops due to the result of failure of synthesis or release of Mullerian duct inhibiting factors or due to failure of end organs to respond or defect in timing of release of Mullerian inhibiting factors. [5] An inguinal hernia invariably presents in the side where the ectopic testis has migrated. [6] On the basis of presence of various associated anomalies Transverse Testicular Ectopia has been classified into three types:

Type 1: Associated with only hernia it contributes about 40-50%.

Type 2: Accompanied with Persistent or rudimentary Mullerian structures.

Type 3: Associated with hypospadiasis, scrotal abnormality and pseudo hermaphrodite [4].

According to this classification, our case comes under Type 2.

Mean age of presentation of Transverse Testicular Ectopia is 4 years. Clinically it presents as inguinal hernia in one side with contralateral cryptorchidism. Diagnosis of TTE is made mostly during surgical exploration for inguinal hernia repair due to unawareness of the condition. So every surgeon operating on inguinal hernia should be well aware of this condition.

Malignant transformation of gonads is the major risk of patients with TTE. Incidence of malignant transformation of gonads is 18% as per studies includes embryonal carcinoma, seminoma, yolk sac tumour, and teratoma. [7] Walsh *et al.* [8] conducted a study and concluded that the incidence of testicular cancer increased to 6 times if the orchidopexy has been delayed until after the age of 10-11 years of age. Wood *et al.* [9] conducted a study and concluded that risk of malignancy in undescended testicles decreased if their orchidopexy performed before ages 10 to 12 years. In almost 97% of patients with Crossed Testicular Ectopia have disorders associated with upper and lower urinary tract system. There is no report of malignancy arising from the retained Mullerian structures, and the absence of MIS does not appear to increase the relative risk of testicular malignancy [10, 11]

Hysterectomy is, hence, not recommended routinely in patients who have obvious uterus and fallopian tubes [11]. Extensive dissection of vas deferens and excision of persistent Mullerian duct structures should be avoided in order to prevent the injury [11].

Once the diagnosis of TTE is made, surgical treatment for fertility preservation and placement of testis in hemiscrotum is the best method in younger age group. In our case the patient is 42 yrs old and chances of malignant transformation are high so bilateral orchidectomy done. In younger individual, diagnostic laparoscopy should be performed to look for Mullerian remnant and to access the cord length, vascularity and any other proximal attachment can be made out. According to intra-abdominal finding, various surgical treatments can be given. If there is adequate length of the vas deferens and cord then transeptal orchidopexy is the choice. Another method is transeptal contralateral orchidopexy or transabdominal orchidopexy if the length of the cord with vascularity is limited [11].

Conclusion

Transverse testicular ectopia is a rare congenital disorder with unclear pathology. Diagnosis of transverse testicular ectopia should be suspected if there is unilateral inguinal hernia with contralateral cryptorchidism. Ectopic testis can lie in superficial ring, inguinal canal, deep ring or intra-abdominal cavity. In suspected case diagnostic laparoscopy and ultra-sonogram or CT can be considered as investigation of choice. Laparoscopy at present is useful for both diagnostic and therapeutic management of Transverse Testicular Ectopia and associated anomalies. Bilateral orchidectomy can be considered if the patient's age is more than 12 yrs.

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