Asian Journal of Case Reports in Surgery

5(3): 10-14, 2020; Article no.AJCRS.61664

Transverse Testicular Ectopia with Persistent Müllerian Duct Syndrome and Inguinal Hernia – A Case Report

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Author's contribution

The sole author designed, analysed, interpreted and prepared the manuscript.

Article Information

Editor(s):
(1) Dr. Fehmi Kaçmaz, Üsküdar University, Turkey.
Reviewers:
(1) Salazar Morales Miguel Fernando, Centro Médico Naval, Mexico.
(2) Ismail Abdul Sattar Burud, International Medical University, Malaysia.
Complete Peer review History: http://www.sdiarticle4.com/review-history/61664

Case Report

Received 25 July 2020
Accepted 01 October 2020
Published 19 October 2020

ABSTRACT

Transverse testicular ectopia (TTE) is a rare congenital anomaly, in which both gonads migrate towards the same hemiscrotum. Other abnormalities such as persistent Müllerian duct syndrome, true hermaphroditism, inguinal hernia, hypospadias, pseudohermaphroditism, and scrotal anomalies may be associated with it. Here we present the case of a 3 year old male child who presented with the complaint of a swelling in the right inguinal region since 3 months. Swelling increased in size on crying and coughing. On examination the child had a right inguinal hernia and the left side of scrotum was empty. Right testis was palpable separate from the hernia. Left testis was not palpable in the left side of scrotum or the left inguinal region. The child was planned for right herniotomy and left inguinal exploration. Intraoperatively, both testes were present in the right inguinal canal along with persistent Müllerian structures. The Müllerian structures were divided in the midline and the testes fixed in the respective scrotal sacs. Transverse testicular ectopia should be suspected in any patient with a nonpalpable undescended testis with contralateral inguinal hernia.

Keywords: Transverse testicular ectopia; persistent Müllerian duct syndrome.

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1. INTRODUCTION

Transverse testicular ectopia (TTE) also named crossed testicular ectopia, testicular pseudoduplication, unilateral double testis or transverse aberrant testicular maldescent, is a rare anomaly in which both testes descend or migrate through a single inguinal canal or hemiscrotum. TTE is usually associated with other abnormalities such as persistent Müllerian duct syndrome, true hermaphroditism, inguinal hernia, hypospadias, pseudohermaphroditism, and scrotal anomalies.

2. PRESENTATION OF CASE

A three year old male child presented with the complaint of a swelling in the right inguinal region since three months. Swelling increased in size on crying and coughing. On examination the child had a right inguinal hernia and the left side of scrotum was empty. Right testis was palpable separate from the hernia. Left testis was not palpable in the left side of scrotum or the left inguinal region (Fig. 1). The child was planned for right herniotomy and left inguinal exploration. At the time of operation, the right inguinal region was explored initially. Two gonads were found in the right inguinal area, both gonads were similar in size and were oriented vertically (Fig. 2). There were 2 vas deferens and vascular structures accompanying each gonad, between them was a tubular structure, resembling an immature uterus, and fallopian tubes. The Müllerian structures were divided in the midline and mucosa removed (Figs. 3 and 4). The gonads were placed in the respective subdartos pouches in each hemiscrotum by the transseptal approach. After the surgery, all hormonal studies done were normal and the karyotype was 46 XY.

3. DISCUSSION

TTE is a rare form of testicular ectopia. It was first reported by Von Lenhossek in 1886 [1]. The mean age of presentation is around 4 years and most of the cases are diagnosed on surgical exploration. Preoperative diagnosis maybe done by USG or MRI. There are various theories postulated in an attempt to explain its etiology. Berg proposed the possibility of the development of both testes from the same genital ridge [2]. Kimura stated that if both vasa deferentia arose from one side, there had been unilateral origin but if there was bilateral origin, one testis had crossed over [3]. Josso suggested various anatomical factors that may result in such an anomaly: defective implantation of the gubernaculum, obstruction of the inguinal ring and development of adhesions between the testes and adjacent structures [4]. Gupta and Das postulated that adherence and fusion of the developing Wolffian ducts took place early, and that descent of one testis caused the second one to follow [5]. Paltii suggested a defective implantation of the gubernaculum testis or an obstruction of the inguinal ring preventing testicular descent on the ipsilateral side. Although many theories have been postulated, no single theory can explain the etiology and its variations and associated syndromes [6]. Thevasthan ascribed TTE to both the testes lying

Fig. 1. Preoperative picture

Fig. 2. Both testes present in right inguinal canal

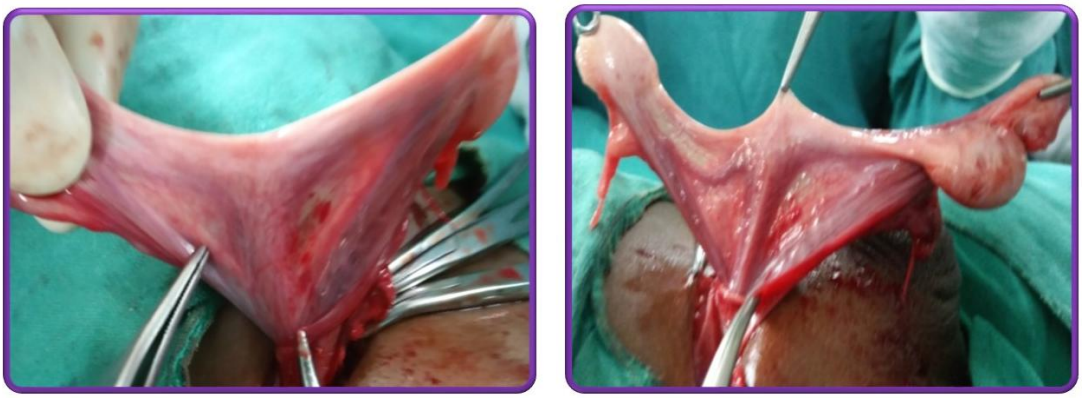

Fig. 3. Both testes and persistent mullerian structures

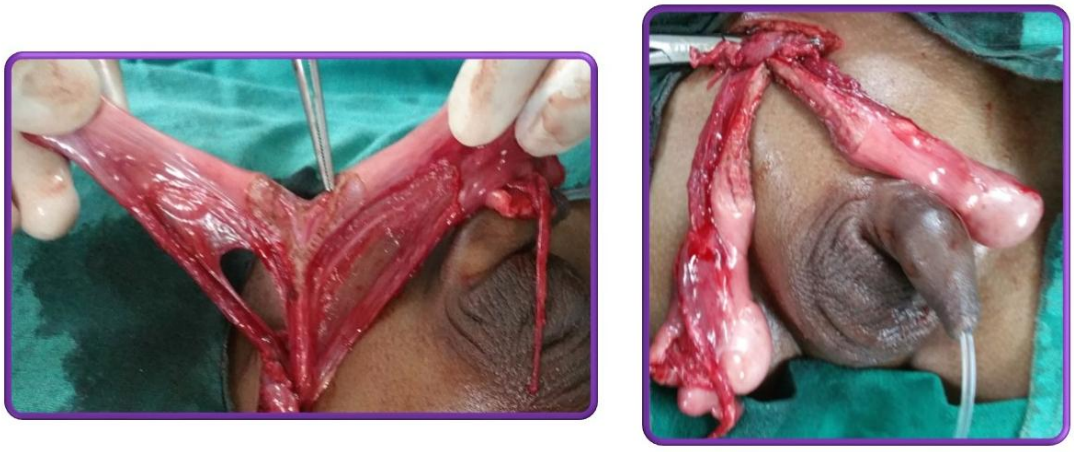

Fig. 4. Division of mullerian structures in the midline

in the same saccus vaginalis before descent. This occurs if the testes or vasa deferentia are bound together or if the vasa are bound to Müllerian structures. This theory explains the persistent Müllerian structures associated with TTE [7]. TTE has been classified into 3 types: Type 1, accompanied only by hernia (40% to 50%); type 2, accompanied by persistent or rudimentary Müllerian duct structures (30%); and type 3, associated with disorders other than persistent Müllerian remnants (inguinal hernia, hypospadias, disorders of sexual differentiation and scrotal abnormalities) (20%) [8]. Persistent Müllerian duct syndrome (PMDS) is a rare form of male disorder of sexual development characterized by the presence of uterus or Fallopian tubes in phenotypically normal 46 XY males. TTE associated with PMDS is a rare syndrome and it was first described by Jordan in 1895 [9]. During embryogenesis, regression of Müllerian structures is mediated by anti-Müllerian hormone(AMH), also called Müllerian inhibiting substance (MIS) produced by fetal Sertoli's cells. PMDS is attributed to deficient AMH activity or to abnormalities in AMH receptors.

PMDS may be suspected on ultrasound or magnetic resonance imaging. It is confirmed by diagnostic laparoscopy, testicular biopsy and chromosomal studies. Patients with TTE are at increased risk of malignant transformation. The overall incidence of malignant transformation of gonads is higher than undescended testis (18%) [10]. Embryonal carcinoma, seminoma, yolk sac tumor and teratoma have been reported in TTE. Though there is a theoretical risk of malignant change, no case of malignancy arising from the Müllerian remnants has been reported; therefore, there is currently no indication for the need to excise the Müllerian remnants which may injure the vas and blood supply to testes. Wuerstle reported a case series of non identical triplets with PMDS with TTE and concluded that the surgical approach of orchiopexy and hernia repair without removing mullerian structures is the optimal surgical management [11]. Vandersteen states that the surgical management of patients with PMDS is controversial due to the potential morbidity associated with both the retention and the removal of the Müllerian structures and there are no absolute indications for removal of the persistent Müllerian duct structures; and that surgical excision of the persistent uterus and tubes has few potential benefits and excision does have significant risks [12]. El-Gohary reported 5 cases of PMDS with TTE managed laparoscopically by splitting the uterus in the midline and then bringing the testis with the vas and attached uterine tissue into the scrotum. They preserved the blood supply to the testicular tissues by bringing them on to a rim of uterine tissues to preserve future fertility [13].

4. CONCLUSION

Transverse testicular ectopia should be suspected in any patient with a nonpalpable undescended testis with contralateral inguinal hernia. Persistent Müllerian Ducts should not be excised as it may damage vas and vessels to the testes and can be divided in the midline. Patients of TTE should be treated adequately and safely and should be followed up for long term considering the increased risk of malignancy in such patients.

CONSENT

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author.

COMPETING INTERESTS

Author has declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/61664