

Transverse testicular Ectopia: a rare anomaly

Abstract

Crossed Testicular Ectopia (CTE)/Transverse Testicular Ectopia (TTE) is a rare but well known congenital anomaly, in which both gonads migrate toward the same hemiscrotum. It is usually associated with other abnormalities such as persistent Mullerian duct syndrome, True Hermaphroditism, Inguinal Hernia, Hypospadias, Pseudohermaphroditism, and scrotal anomalies. We report a case of 10month old male child with left inguinal hernia with suspicious left inguinal swelling and empty right scrotum. Diagnosis was confirmed preoperatively by ultrasound followed by diagnostic laparoscopy and open inguinal exploration for orchiopexy.

Keywords: transverse testicular ectopia, orchiopexy

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Abbreviations: TTE, transverse testicular Ectopia; PMDS, persistent mullerian duct structures; MIF, mullerian duct inhibitory factor

Introduction

Transverse Testicular Ectopia (TTE) is a rare congenital anomaly and is also referred to as testicular pseudoduplication, unilateral double testis, and transverse aberrant testicular with mal-descent. Associated abnormalities may include persistent mullerian duct syndrome, true hermaphroditism, inguinal hernia, hypospadias, pseudohermaphroditism, and scrotal anomalies.^{1,2} We report a case of TTE diagnosed pre-operatively due to prompt surgical suspicion and confirmed by exploration and repair.

Case presentation

A 10 month old male child presented with complaints of left inguinal hernia and absent testes on right side. General examination and blood investigations were normal. On local genital examination, the right hemiscrotum was well developed but empty. A well developed left hemiscrotum with testes was seen. A single left inguinal swelling of 1x1 centimeter (cm), with testicular sensation and cough impulse was present (Figure 1). Ultrasound confirmed the presence of the testes with one demonstrated in the left hemiscrotum and other at the left superficial inguinal ring (Figure 2). Right hemiscrotum was empty and no mullerian structures were seen on abdominal ultrasound. Diagnostic laparoscopy confirmed these findings. The right vas deferens was visualized crossing the midline entering the left deep inguinal ring along with left cord structures. Open inguinal exploration showed left testis in left hemiscrotum and a small right testis with an accompanying fluid hernia in the left inguinal canal. Both testes had a common meso-orchium proximally and were separated by a distance of 4cm (Figure 3). Herniotomy was performed and the lower left testis was placed in the right subdartos pouch by trans-septal approach and right testes in left subdartos pouch.

Discussion

TTE is an uncommon anatomical abnormality where both the gonads migrate towards the same hemiscrotum. Approximately 100 cases have been reported in literature¹ and it was first described by Von Lennhosek.²

Embryologically, several theories regarding the origin of TTE have been suggested including 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. Persistent mullerian duct structures (PMDS) may result from the failure of synthesis or release of mullerian duct inhibitory factor (MIF), the failure of end organs to respond to MIF, or defect in the timing of the release of MIF. It seems possible that the mechanical effect of the persistent mullerian duct structures prevents the testicular descent or leads to both testicles descending toward the same hemiscrotum producing TTE.³ The association with cryptorchidism is accompanied by an increase in malignancy potential of crossed ectopic testes.



Figure 1 Clinical photo showing both the testes on the left side.



Figure 2 Ultrasonogram confirming testes in left hemiscrotum and other in left inguinal canal.



Figure 3 Intraoperative pathology with both testes on same side sharing a common mesorchium.

The ectopic testis may lie in opposite hemiscrotum, in the inguinal canal or at the deep inguinal ring. An inguinal hernia is invariably present on the side to which the ectopic testis is migrated. On the basis of the presence of various associated anomalies, TTE has been classified into 3 types:⁴

Type 1– accompanied only by hernia (40% to 50%)

Type 2– accompanied by persistent or rudimentary mullerian duct structures (30%)

Type 3– associated with disorders other than persistent mullerian remnants (hypospadias, pseudohermaphroditism, and scrotal abnormalities) (20%).

The mean age at presentation is 4 years and the clinical presentation generally includes an inguinal hernia on one side and a contra lateral or sometimes a bilateral cryptorchidism.^{5,6} Usually, the correct diagnosis cannot be made before surgical exploration. The diagnosis of TTE can be made preoperatively by close clinical examination and use of ultrasonography by an experienced sonographer.⁷ Patients with TTE are at increased risk of malignant transformation with the overall incidence of malignant transformation of gonads approximately 18%.⁸ There have also been reports of associated embryonal carcinoma,⁹ seminoma, yolk sac tumor,¹⁰ and teratoma.⁸ Walsh et al.¹¹ concluded that testicular cancer was nearly 6 times more likely to develop in cryptorchid cases where operations were delayed until after 10 to 11 years of age. Wood et al.¹² showed that risk of malignancy in undescended testicles decreased if orchiopexy performed before ages 10 to 12 years. In patients with TTE, disorders of urinary tract system have also been reported.¹³ Once diagnosis of TTE is made, orchiopexy is recommended for the preservation of fertility.⁶ Laparoscopy is useful for both diagnosis and treatment of TTE and associated anomalies.¹⁴ Management for testicular ectopia is either trans-septal or extra-peritoneal transposition orchiopexy,^{15,16} a search for mullerian remnants or other anomalies, and a long-term postoperative follow-up. In the extra-peritoneal technique the testis is brought to the contra-lateral hemiscrotum crossing the root of penis. In the trans-septal technique the testis should traverse the scrotal mediastinum (septum) to be fixed in it. TTE may also be misdiagnosed as an inguinal hernia, intersex,¹⁴ or present as an irreducible hernia requiring urgent surgery.¹⁷

TTE associated with fused vas deferens is extremely rare. This condition may hinder the testis from being placed into the scrotum

during orchiopexy.¹⁸ In cases of fused vas deferens, a trans-septal orchiopexy is recommended.

Conclusion

TTE is a rare anomaly whose pathogenesis remains unclear. The diagnosis should be considered when unilateral hernia and concurrent cryptorchidism of the contralateral side are present. In suspected cases, ultrasonographic evaluation and laparoscopy may be helpful in diagnosing this condition before surgery. Transseptal orchiopexy is recommended to manage TTE. Laparoscopy may be useful for both diagnosis and management of TTE and associated anomalies.

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Conflicts of interest

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