Crossed Testicular Ectopia: A Very Rare Congenital Anomaly

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Abstract

Crossed testicular ectopia or transverse testicular ectopia is a very rare congenital anomaly in which testes deviate from their normal path during descent resulting in the ipsilateral location of both testes usually in the inguinal canal. Associated congenital anomalies include persistent Mullerian duct syndrome, hypospadias, true hermaphroditism, pseudohermaphroditism and scrotal anomalies. Most of the cases are diagnosed incidentally during surgery for inguinal hernia. Treatment options include transseptal or extraperitoneal transposition orchidopexy. We are presenting a case report of crossed testicular ectopia in the one-year-old boy who was operated for right inguinal hernia with right-sided undescended testis and left-sided absent testis but during herniotomy, both testes were found in right inguinal canal.

Keywords: Crossed Testicular Ectopia; Transverse Testicular Ectopia; Undescended Testes; Cryptorchidism; Inguinal Hernia; Ectopic Testes

Introduction

Crossed testicular ectopia or transverse testicular ectopia is a very rare congenital anomaly in which both testes have migration towards the same side of the scrotum [1]. The child usually presents with a symptomatic hernia on the side where testis has migrated and absence of testis on another side. Most of the times ectopic testes are found incidentally during surgery for an inguinal hernia [2].

Case Report

One year old male child, known case of right inguinal hernia with right undescended testis and left absent testis, presented to emergency with right sided inguinal swelling which is now irreducible. Child developed abdominal distension along with multiple episodes of bilious non-projectile vomiting and constipation. One examination there was tense and tender swelling in right inguinal region. Hernia was reduced under sedation with difficulty and child was admitted for herniotomy on next elective operation list. During herniotomy The right testis was present at the right deep inguinal ring. There was another testis at the proximal part of the cord. Diagnosis of Crossed Testicular Ectopia was made preoperatively and the surgical plan changed to separation of cords and fixation of both the testes to their respective hemiscrotum. Both the cords were having a common origin for almost 4 - 5 cm. The left cord was separated from right cord by careful dissection. Finally, the right testis was fixed to the right hemiscrotum. As there was adequate length of cord was available, the left testis was easily brought to left hemiscrotum extra-peritoneally by making suprapubic subcutaneous tunnel. Both the testes were fixed in the sub-dartos pouch. Testicular biopsy was also taken. Post-operative recovery was uneventful and child was discharged on 2nd post-operative day. Histopathology of testicular biopsy showed Testicular tissue showing immature seminiferous tubules.

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Discussion

During the descent, testes may deviate from their normal path and land in an abnormal position. Various ectopic positions have been described in literature such as superficial inguinal, femoral, suprapubic, perineal and penile. Crossed testicular ectopia (CTE) or transverse testicular ectopia (TTE) is a well-documented but a very rare clinical condition in which both testes are present on the same side. It is a congenital anomaly but children usually present with the mean age of 4 years [3].

The first case of crossed testicular ectopia was reported by von Lenhossek in 1886 [4]. Since then nearly 100 cases have been reported in the literature so far [3]. CTE may be associated with other congenital anomalies as well such persistent Mullerian duct syndrome (PMDS),

Figure 1: Both testes present in right inguinal canal (Arrows).

Figure 2: Histopathology shows Testicular tissue having immature seminiferous tubules.

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inguinal hernia, hypospadias, true hermaphroditism, pseudohermaphroditism, seminal vesicle cyst and abnormalities of scrotum [4,5]. Jordan was the first who described the association of persistent mullerian duct syndrome with CTE in 1895 [6]. Some cases of CTE associated with malignancy of testis are also reported in literature [7,8]. CTE is characterized by the absence of testes on one side with the symptomatic inguinal hernia and ectopic gonads on the contralateral side.

The cause of crossed testicular ectopia is unknown. Various theories have been postulated for this abnormal descent. Gubernaculum rapture and dysfunctional genitofemoral nerve have been described to be among the possible causes [9]. According to Gupta and Das, early adherence and fusion of developing Wolffian duct are responsible for CTE, that descent of one testis causes the other one to follow it on the same side. Kimura is of the view that if the ductal system to both testes is fused, it is likely that both testes arise from the single genital ridge and actual crossing occurs only when ductus deferens is separate for each testis [2].

Crossed testicular ectopia is classified into three types according to its associated congenital anomalies. Type I is the most common type (40 - 50%) and is associated with an inguinal hernia alone. Our case report is type I CTE. Type II CTE (30%) is associated with Persistent or rudimentary Mullerian duct structures such as the uterus and fallopian tubes. Type III CTE (20%) has associated genitourinary anomalies other than Mullerian duct remnants such as an inguinal hernia, hypospadias, pseudohermaphroditism and scrotal abnormalities [1,10,11].

In most of the reported cases, CTE was diagnosed incidentally during herniotomy and diagnosis could not be established preoperatively. High index of suspicion is required for diagnosis [12,13]. In cases with absent testes in the contralateral inguinal hernia, CTE should be expected and workup should be done accordingly. Various imaging modalities are used to diagnose CTE preoperatively. These include ultrasonography, CT, Arteriography, Venography and MRI [9]. Ultrasonography is simple, non-invasive and avoids radiation and sedation but has limited sensitivity and specificity. Wolveerson, et al. compared Ultrasonography with CT scan in the diagnosis of CTE. They found 94% sensitivity with CT, 88% sensitivity with Ultrasonography and 100% specificity when both modalities were used [14]. While comparing MRI with MRV, MRV is better than MRI in diagnosing CTE as its sensitivity is 100% as compared to 82.4% sensitivity of MRI. Lam., et al. concluded that sensitivity of MRI (84%) and ultrasound (82 - 88%) are comparable and in the presence of testicular tissue Ultrasound is suitable. MRV is far better than Ultrasound, CT and MRI. They recommended that during the workup of undescended testes Ultrasoundography should be performed first followed by MRI if ultrasonography is negative. MRI should be done if MRI findings are negative [2].

Treatment of CTE is surgical which can be open or laparoscopic. Treatment depends on the type of CTE. The main aim of treatment is to detect associated congenital anomalies and place the testes in anatomical position. In case of type I CTE herniotomy and orchidopexy is done. Ectopic testis can be brought down to respective scrotum either through a transseptal incision or by extraperitoneal transposition orchidopexy if there is an adequate length of spermatic cord available. In transseptal orchidopexy, the ectopic testis is brought down through opposite inguinal canal and fixed in its actual hemiscrotum through the scrotal septum. Transseptal orchidopexy is the treatment of choice. In extraperitoneal transposition orchidopexy, testis is brought to opposite hemiscrotum by a suprapubic subcutaneous tunnel through its respective superficial ring [1,3,4,10,11].

In cases of persistent Mullerian duct syndrome (PMDS), two different surgical techniques are used. In the first technique, Mullerian duct structures are not removed but split in half while in second, these structures are removed as much as possible as they are believed to have a malignant transformation at a later age. Long-term follow up is advised in cases where these structures are preserved. Various malignancies such as adenocarcinoma, squamous cell carcinoma and adenosarcoma have been reported in patients with PMDS [15].

Contribution of Authors

Muhammad Umar Nisar: Case report, write-up of manuscript and literature search.
Khawar Abbas: Editing and Literature search.
Muhammad Amjad Chaudhry: Proofreading and final approval

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Conflict of Interest

Authors declare no conflicts of interest.

Bibliography