REVIEW ARTICLE

CROSSED TESTICULAR ECTOPIA – A REVIEW

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ABSTRACT

Crossed testicular ectopia, an extremely rare anomaly, is a deviation of testicular descent resulting in unilateral location of both testis, the etiology of which is not exactly known till yet. In most cases, the patient comes to the hospital because of cryptorchism on one side, and inguinal hernia on the other side, so the patients are usually very young. It is often not diagnosed until surgical exploration. This manuscript illustrates this condition with possible etiology, clinical features, diagnosis and management reported in the literature.

KEY-WORDS: Crossed; Testicular; Ectopia

Introduction

Normally the testes are located in the scrotum at birth. Ectopic testes have been reported in different sites, including suprapubic, femoral area, perineal area, the base of the penis.[1] Crossed testicular ectopia is an extremely rare form of testicular ectopia but well recognized entity in which both gonads migrate towards the same hemiscrotum. The clinical findings are usually symptomatic inguinal hernia on one side to which the ectopic gonad has migrated, and an impalpable testis on the other side. In most reported cases, the correct diagnosis is not made preoperatively, but made on the operation table as the patients are operated for repair of inguinal hernia.[2-4]

Development of Testes

Testis develops on the posterior abdominal wall in the lumber region from the genital part of the urogenital ridge. It is mesodermal in origin.[5]

Descent of Testes

In the embryonic life, testis develops in the posterior abdominal wall in lumber region and descent slowly so that it reaches up to the scrotum at birth. Testis reaches the iliac fossa in the third month, at deep inguinal ring in the seventh month, in the inguinal canal at the eighth month and finally reaches at scrotum by the end of ninth month.[5,6]

Factors responsible for descent are[5]

(1) Differential growth of the posterior abdominal wall.
(2) Formation of inguinal bursa by the entry of the tests.
(3) Shortening of gubernaculum-a band of mesenchyme which connects lower end of testis to the scrotum. It also dilates the inguinal bursa and lays down the path for the descent of the testis.
(4) Intraabdominal pressure and temperature.
(5) Squeezing action of internal oblique.
(6) Hormones secreted by the anterior lobe of hypophysis cerebri.

Processus vaginalis is the diverticulum of the peritoneal cavity growing into the mesenchyme of the gubernaculum, inguinal canal and the scrotum. Testis descents and invaginates the processus vaginalis from behind. Once the descent of the testis is complete, the processus vaginalis gets atrophied.[5]

Delayed testicular descent is seen in 3-4% of term newborns. This rate is about 30% in premature infants.[7]

Ectopic testis

Ectopic testis is defined as the aberrant location of testis in places other than the scrotum during descent[3]. Various sites are[5]

(1) Interstitial type-common type where testis lies external to external oblique aponeurosis in lower part of anterior abdominal wall
(2) Anteromedial aspect of thigh
(3) Near the anterior-superior iliac spine
(4) Root of the penis
(5) In the perineum
(6) Monorchism- one testis is intraabdominal and other is in its normal position

Crossed ectopia of testis- rare type where both the testes are in same hemiscrotum.

Crossed Testicular Ectopia

It is an interesting and rare form of testicular ectopia. It is characterized by the descent of both testicles through the same inguinal canal, toward the same hemiscrotum. It is also called as transverse testicular ectopia, testicular pseudoduplication, unilateral double testis and transverse aberrant testicular maldescent.[8]

First description of crossed testicular ectopia was made by Lenhossek[9] in 1886. He presented the case of a 35 years old man, at whose autopsy both testicles were found in the left hemiscrotum. In 1895, Jordan Max presented the case of 8 years old boy with hypospadias operated for left inguinal hernia, and during surgery he found in the hernia sac the right testis.[10] In 1907, Halstead described another case.[11]

Etiology

The etiology of this anomaly is still unknown. Lenhossek (1886)[9] was the first to suggest that this type of testicular ectopia is caused by an abnormal gubernaculum testis. Frey also noted that defective ipsilateral gubernacular development might predispose to crossed testicular ectopia. The mechanism by which the gubernaculum causes descent remains unclear, but scrotal enlargement secondary to gubernacular growth is one possible explanation.[12] Lockwood (1888) further elaborated this theory, describing the five parts of the gubernaculums and their points of insertion: the bottom of the scrotum, the front of the pubis, the perineum, scarpa's triangle in the thigh, the region of the inguinal ligament just medial to the anterior superior iliac spine.[13]

Berg (1905) suggested that both testicles arose from the same genital ridge.[14]

Gray and Skandalakis thought that, since in most cases, the each testis has his own deferent duct, the crossing over occurred during testicular descent, each testis arising from the ipsilateral genital ridge.[15]

Kimura stated that true crossed testicular ectopia is only if there are two distinct deferent ducts, a common duct suggesting the development of the testis from one genital ridge.[16]

Gupta and Das postulated that adherence and fusion of the developing wolffian ducts takes place early and that descent of one testis causes the other one to follow it toward the same hemiscrotum.[17]

Josso (1977) developed the theory of anatomical anomalies, which includes: abnormal gubernaculum testis, obstruction of the inguinal canal, adherences between the testis and the nearby structures.[18]

Another theory says that in the cases which present persistent Mullerian duct syndrome, the crossed ectopia is caused by the traction exercised on the testis by the Mullerian duct.[3]

Clinical features

The mean age at presentation is four years.[3] It is common on right side than left side.[3] Clinical signs include ipsilateral empty hemiscrotum and contralateral inguinal hernia.[2-4] Cases might also present with incarcerated inguinal hernia that cannot be reduced.[3]

Types

Crossed testicular ectopia is classified in three clinical types according to the presence of additional abnormalities.[3,19]

Type I: It is most common form where only inguinal hernia accompanies (40-50%). Vascular structures and ductus deferens of the testicles might be separated or united.
Type II: It is associated with persistent or rudimentary Mullerian canal remnants (30%).
Type III: It is associated with genitourinary anomalies, other than persistent Mullerian duct (13-20%): hypospadias, pseudohermaphroditism, seminal vesicle cyst, scrotal abnormalities, common deferent duct, and horseshoe kidney.

Diagnosis

In most cases, the correct diagnosis has not been made preoperatively, and the condition is revealed during herniotomy. Recently ultrasonography (USG), computerized tomography (CT) scan and magnetic response imaging (MRI) have been suggested to determine preoperative localization of impalpable testis.[20]

USG helps to avoid the use of ionizing ration and permits evaluation without sedation; however it might be difficult to perform in uncooperative or very young patients. Moreover, it is difficult to use USG to scan a testis cephalic to the internal inguinal ring. The ectopic testis is identified by the presence of characteristic bright signal on T2-W images and a linear low-signal structure which may represent the remnant of the gubernaculums testis on MRI.[20]

The diagnosis of crossed testicular ectopia should be followed by more tests to exclude other genitourinary anomalies. It is important to exclude and associated persistent Mullerian duct syndrome, the two being frequently found together.

Differential Diagnosis

(1) Testicular duplication where both testicles aroused from the same genital ridge and they have a common deferent duct and blood supply.[21]
(2) Hydrocele[22]
(3) Spermatocele[22]
(4) Tumor of the testis[22]
(5) Spleenogonadal fusion[22]
(6) Other causes of ipsilateral empty hemiscrotum such as retractile testis, anarchism, undescended testis and testicular atrophy secondary to testicular torsion.[3]

Management

Little attention has been focused on the treatment. A variety of procedures have been described, including a staged procedure to bring the ectopic testis into its correct canal.[2] In most case a transeptal orchidopexy, the Ombredanne technique is performed.[23] Nother technique brings the ectopic testis in his hemiscrotum using a suprapubic subcutaneous tunnel called as extraperitoneal transposition orchidopexy.[2] Where both testes are found to lie in the scrotum, herniotomy is the only action required.[2]

In 2002, Dean and Shah reported a case successfully treated by laparoscopic approach.[24]

Complications

There have been reported cases of malignant transformation (embryonic carcinoma, seminomas, choriocarcinoma, teratoma) of the ectopic testis. The risk of malignant transformation is of 5%, and can happen bilateral. Because of this risk the patient should followed up for a long period of time. The testicular biopsy and hormonal and genetic assessment are not routinely used. They should be performed only when persistent Mullerian duct syndrome is suspected.[25]

Conclusion

In conclusion, it can be said that crossed testicular ectopia should be a diagnosis considered when unilateral inguinal hernia and concurrent cryptorchism of the contralateral side are present. An appropriate preoperative assessment and careful differential diagnosis to rule out other potential abnormalities are needed. Clinicians should be cautioned that patients with a history of crossed testicular ectopia require long-term follow-up for the development of malignancy.

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