TRANSVERSE TESTICULAR ECTOPIA: A CASE REPORT AND REVIEW OF LITERATURE.

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Abstract

Transverse testicular ectopia, also called Crossed testicular ectopia, is an uncommon congenital anomaly in which both testes migrate through a single inguinal canal towards the same hemiscrotum. A few more than 100 cases have been reported in the literature. This rare syndrome is commonly associated with abnormalities of genitourinary development, especially inguinal hernia and defective Mullerian regression. A conservative approach is recommended, now performed via laparoscopy. Long-term follow-up is required for assessment of fertility and early detection of testicular malignancy. The authors report a new case of crossed testicular ectopia in a 4-year-old boy operated in King Faisal Hospital Saudi Arabia, who presented with right inguinal hernia and bilateral undescended testes.

Key words: testis crossed testicular ectopia, transverse testicular ectopia, testicular migration, ectopic

Introduction

Transverse testicular ectopia (TTE) in an extremely rare entity in which both testes migrate along the same inguinal canal towards the same hemiscrotum, while the opposite inguinal canal and hemiscrotum are empty. The ectopic testis may be located on the inner inguinal ring, in the inguinal canal, or in the contralateral hemiscrotum [1].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. TTE is usually associated with abnormalities such as persistent Mullerian duct syndrome (PMDS). PMDS is a rare form of male pseudohermafroditism characterized by the persistence of Mullerian duct structures (uterus, fallopian tubes) in phenotypically normal boys [2]. Majority of patients are of a very young age, around 1–2 years old. In 65% cases the exact diagnosis is not determined prior to surgical intervention. Patient often undergoes unsuccessful inguinal exploration and the ectopic testis is usually discovered during the repair of an inguinal hernia [3]. Classically, in TTE the two testes are identical in size and appearance; each has its own epididymis, vas deferens and testicular vessels; the vascular supply and vas deferens of the crossed testis are derived from the appropriate side; the processus vaginalis is patent on the side of the two testes and there is no hernia on the side of the undescended testis [4]. In the pre-laparoscopic era, unilateral TTE was most often diagnosed during herniotomy or following inguinal exploration for contralateral impalpable testis. However, with the increasing use of laparoscopy for the evaluation of impalpable testis, patients with TTE are diagnosed prior to inguinal exploration [5].

In this case, our aim is to raise awareness of this uncommon condition with review of literature on different aspects of management.

Case Report

A 4 years old child was referred to our outpatient department by a general practitioner with the history of bilateral absence of testes in the scrotum since birth and an occasional bulge in the right inguinal canal for a few months, as noticed by the parents. There was no significant family history and other two male children were quite healthy. On physical examination child was found to be in a good state of general health. Scrotum was under developed and cough impule was positive in the right inguinal region. Scrotum was empty and nothing could be palpated in the left inguinal area while testicular tissue and inguinal hernia were clinically appreciated on the right side. So, with the clinical diagnosis of right inguinal hernia with bilateral undescended testes. Ultra sound was requested. Report was consistent with the presence of both testes in the right inguinal region with patent process vaginalis. After necessary blood work up , patient was placed on elective operative list. Under general anesthesia, right side was explored through right inguinal crease incision. On opening the inguinal canal patent process vaginalis was identified. It was traced down and found to be attached with the pubic tubercle through the gubernaculum. Gubernaculum was divided, further exploration revealed both the testes in the hernia sac. Herniotomy was carried out carefully. Both the testes were attached with each other through a small fiberous mesentery. There were separate spermatic vessels and vas deference traced till the deep inguinal ring. The spermatic cord of left testis was longer than the right [Fig.1]. As both were adequately mobilized so, right testis was fixed in the right subdartos pouch while left was fixed in the left subdartos pouch through transseptal approach. Post operative course was uneventful and patient did well on follow up.

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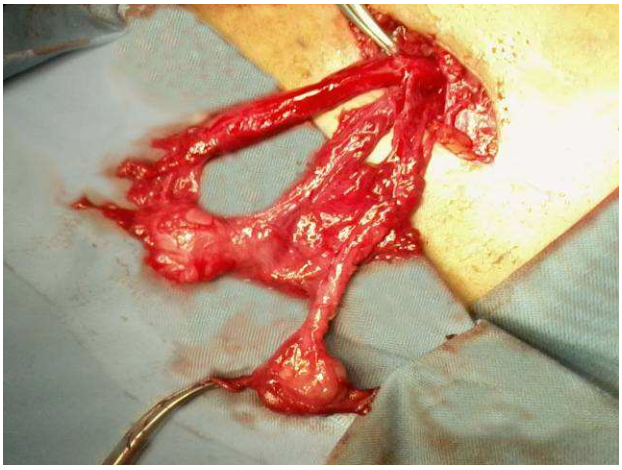

Figure 1. Both testes delivered from the same inguinal canal with separate spermatic cords.

Discussion

In 1886, von Lenhossek described the first case of transverse testicular ectopia and, in 1895, Jordan described the syndrome of transverse testicular ectopia with persistent Mullerian ducts [6]. Transverse testicular ectopia is a rare anomaly in which the testis is seen either in the contralateral inguinal canal or in the hemiscrotum. This anomaly is also called "testicular pseudo duplication", "unilateral double testis" or "transverse aberrant testicular maldescent". An inguinal hernia is invariably present on the side to which the ectopic testis has migrated. About100 well-documented patients with this condition have been reported. Usually the right testis is ectopic, but, as in the present case, the left side has also been reported [7].

On the basis of the presence of various associated anomalies, transverse testicular ectopia has been classified into three types: those associated with inguinal hernia alone (40–50%); those associated with persistent or rudimentary Mullerian duct structures (30%); and those associated with other anomalies without Mullerian remnants, *e.g.* inguinal hernia, hypospadias, pseudohermaphroditism and scrotal abnormalities (20%) [4]. Our case is the most common variety of TTE.

The etiology of this condition is incompletely understood. Various theories have been put forward, such as: (a) Failure of the gubernacular mechanism and, consequently, failure to open the inguinal canal and the descent to the opposite side is due to adhesions to the testes with normal gubernacular mechanism and inguinal rings, (b) Rupture of the gubernaculum and dysfunction of the genitofemoral Nerve, (c) Both testes arising from the same genital ridge,(d) True crossover of the testes,(e) Adhesion and fusion of the Wolfian duct in early embryonic life and the subsequent descent of both testes on the same side,(f) An aberrant ring on the normal side, (g)Both testes lying in the same processus vaginalis prior to descent [8].

There are many theories attempting to explain the etiology of the isolated TTE. The first serious explanation was given by Lockwood through multiple insertion theory [9]. Gupta and Das assumed that merging of the developing Wolf canals is taking place early and that descent of one testis stimulated the other to follow. However, Gray and Skandalakis consider that crossed ectopia occurs later, since the testes have separate sperm canals [9]. Berg states that the real crossed ectopia occurs only if special sperm canals reach each testis [9]. In a patient with TTE and PMDS, it is thought that a MIF does not have a direct role in the descent of the testes. Therefore, it is likely that the mechanical effect of the persistent Mullerian duct structures produce cryptorchidism by preventing normal testicular descent [10]. It seems possible in some cases that this mechanical effect also leads to both testes being located on the same inguinal side. With anatomical point of view, usually there is separate blood supply for both testes and vas deferens is also separate as in our case but anomalies such as common vas deferens with proximal fusion and unilateral absence of vas have been reported in TTE [11].

Literature review shows that most of the cases are diagnosed during surgery for a hernia or exploration for undescended testis. TTE can rarely present with torsion of the testis, while some children present with obstructed inguinal hernia [12]. Although diagnosis of TTE is usually made during an inguinal hernia repair, nonoperative diagnostic modalities, such as ultrasonography, computerized tomography, magnetic resonance imaging, and magnetic resonance venography, have also been used to identify the testis.

The aim of surgical management is fixation of the testes into the scrotum, a search for Mullerian duct remnants and other anomalies, and long-term follow-up due to the risk of malignancy. Fixation is accomplished either by transseptal orchiopexy or extraperitoneal transposition of the testis [13]. Presently, laparoscopy for both the diagnosis and management of this presentation and even a combined approach of an inguinal method assisted by laparoscopy has been described [14]. Till the recent past it was mentioned in the literature that no malignancy arises from the Mullerian structures, hence, surgical management being orchidopexy, leaving the uterus and fallopian tubes in situ. Recently some cases of malignancy have been detected in the Mullerian remnants so the optimal strategy in these patients is to

closely followup these patients if the Mullerian remnants have been left behind[15].

Conclusion

TTE is an uncommon variety of undescended testis. Patients with bilateral undescended testes and symptomatic inguinal hernia must receive due attention and necessary investigations should be carried out. If Mullerian duct remnants are discovered, laparoscopic management should be the first choice and follow up for any remnant structures is mandatory.

References

1. Oludiran OO, Sakpa CL. Crossed ectopic testis: a case report and review of the literature. Pediatr Surg Int 2005;21:672–673.
2. Chandrasekera SK, Barber NJ, Sheriffdeen AH. Persistent Mullerian duct syndrome with transverse testicular ectopia. Urology 2003; 62:1120.
3. Lam WW, Vanle SD, Chan KL, Chan FL, Tam PK. Transverse testicular ectopia detected by MR imging and MR venography. Pediatr Radiol 2002; 32:126–129.
4. De Luna AM, Ortenberg J, Craver RD. Exploration for testicular remnants: implications of residual seminiferous tubules and crossed testicular ectopia. J Urol 2003; 169:1486–1489.
5. Balaji K.C, Diamond D.A. Laparoscopic diagnosis and management of transverse testicular ectopia. Urology 1995; 46 :879-880.
6. Martin EL, Bennett AH, Cromie WJ. Persistent Müllerian duct syndrome with transverse testicular ectopia and spermatogenesis. J Urol 1992;147:1615–7.
7. Malik MA, Iqbal Z, Chaudri KM et al. Crossed testicular ectopia. Urology 2008; 71(5):984
8. ZivanoviT D, SlavkoviT A, MadiT J et al. Transverse testicular ectopia. Srp Arh Celok Lek. 2004; 132:438–440
9. Young SN, Hong KB, Sun JK, Hong KL, Hwon KP. Transverse testicular ectopia found by properative ultrasonography. J Korean Med Sci 1998;13:328–330.
10. Ueno S, Yokoyama S, Isehera HH. A case of transverse testicular ectopia with Mullerian duct remnant. J Pediatr Surg 2001;36(11):1710–1712
11. Chacko JK, Furness PD, Mingin GC. Pre sentation of fused vas deferens. Urology 2006; 67:1085–1085.
12. Chaabane W, Jarboui L, Sahnoun A, Daoud E, Hadjsliman M, Fakhfekh H et al. Persistent Mullerian duct syndrome with torsion of a transverse testicular ectopia: first reported case. Urology 76(1)65–66. doi:10.1016/j.urology 2009; 10.021
13. Naouar S, Maazoun K, Sahnoun L et al. Transverse testicular ectopia: a three-case report and review of the literature. Urology 2008; 71(6):1070–1073.
14. Deshpande AV, La Hei ER. Impact of laparoscopy on the management of transverse testicular ectopia. J Laparoendosc Adv Surg Tech A 2009; 19(3):443–446.
15. Thiel DD, Erhard MJ. Uterine adenosarcoma in a boy with persistent mullerian duct syndrome: first reported case. J Pediatr Surg 2005; 40:e29–e31

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