Crossed testicular ectopia: what should be specified?

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Crossed testicular ectopia, also named transverse testicular ectopia, 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 Müllerian duct syndrome, inguinal hernia, hypospadias, pseudohermaphrodism, and scrotal anomalies. We report two cases of crossed testicular ectopia; a persistent Müllerian duct syndrome was associated in one case. We also performed a literature search for other reports of crossed testicular ectopia. *Ann Pediatr Surg* 13:110–112 © 2017 Annals of Pediatric Surgery.

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Introduction

Testicular ectopia is an anomaly of testicular descent characterized by localization of the testis out of its normal migration pathway from the retroperitoneum to the scrotum.

In crossed testicular ectopia (CTE), the ectopic testis is found in the opposite groin or hemiscrotum, beside the other testis. CTE is a very rare congenital anomaly, which is characterized by a symptomatic inguinal hernia, with a nonpalpable testis in the controlateral side. Persistant Müllerian duct syndrome (PMDS) is also a rare anomaly that may be associated with CTE. PMDS is an inherited form of intersex disorder characterized by the presence of Müllerian derivatives in otherwise phenotypically normal males [1].

We report two cases of CTE, in which diagnosis was made preoperatively.

Case 1

A 1-year-old boy presented with a left inguinal hernia and right nonpalpable testis. A crossed testicular ectopia was suspected clinically, and ultrasonography examination was performed. It showed crossed testicular ectopia on the left side with hydrocele; no other anomalies were detected. Left inguinal herniotomy showed a PMDS: two left testes, two fallopian tubes, and Müllerian cavities (Figs 1 and 2). Müllerian remnants were resected. Each testis had separate vessels, and both testes had passed through the left inguinal ring after ligation of the peritoneal sac. Fixation of the right testis was performed in the right hemiscrotum after trans-septal passage (Fig. 3). His karyotype was 46 XY.

Case 2

A 2-year-old boy presented with a right inguinal hernia and nonpalpable left testis. An ultrasound was performed for nonpalpable testis, and it showed the absence of the left testis in his normal pathway but the presence of both testes through the right inguinal canal.

Exploration through the right inguinal incision showed two testes: one in the right scrotum and the second in the hernia sac. Both of them had a gubernaculum. The right testis had a gubernaculum fixation on the iliopubic branch. The lowest one had gubernaculum fixation on the right hemiscrotal cavity. No Müllerian remnants had been detected. Each testis had its own vas deferens, but the spermatic cord fused together proximally. After ligature of the hernia sac, the left testis was fixed by a trans-septal way after its passage through the right inguinal ring. The patient was discharged 24 h later without any complication.

Discussion

Crossed testicular ectopia is also called transverse testicular ectopia, or testicular pseudoduplication. Since the first case was reported by Von Lenhossek in 1886, about 170 cases were reported in the literature [2].

Clinically, the most usual presentation is an inguinal hernia with ipsilateral palpable testis and nonpalpable testis on the opposite side. The diagnosis is often made incidentally during herniotomy or during exploration for undescended testis [3]. However, preoperative diagnosis could have been made in some cases [4]. In our cases, diagnosis is made preoperatively by clinical examination and confirmed by ultrasonography.

Gauderer and colleagues has described a classification system for crossed testicular ectopia based on the presence of associated abnormalities: type 1 (40–50%) associated with inguinal hernia alone, type 2 (30%) associated with persistent or rudimentary Müllerian duct structures, and type 3 (20%) associated with other genitourinary abnormalities without Müllerian remnants [2].

The etiology of CTE is unknown. Several theories have been advanced: Berg is the first to explain that both testes develop from the same genital ridge [5]. Kimura proposed that if both vas deferens arose from one side,

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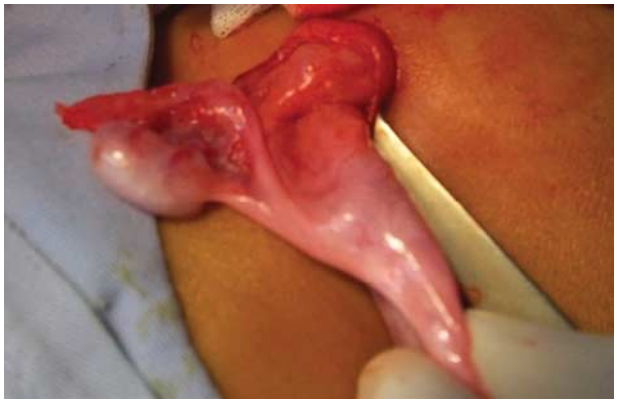
Fig. 1



Persistent Müllerian duct syndrome: right testis, fallopian tubes, left testis, spermatic cord, uterus.

there had been unilateral origin; however, if there was bilateral origin, one testis had crossed over [3,6].

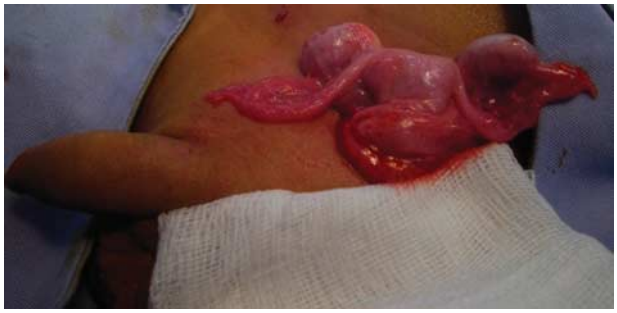
Many authors propose that abnormal or absent gubernaculum development results from an adherent developing wolf ducts. Most authors also agree, that later one testis crosses to the opposite side; they believe that mechanical causes are important for this pathology. The association of CTE with Müllerian duct abnormalities has led some to hypothesize a mechanical interruption of testicular descent by persistant Müllerian duct [4,7].

The treatment of CTE is focused on the detection of associated congenital abnormalities and placement of ectopic testicles into anatomical positions. Trans-septal orchiopexy via controlateral inguinal incision is the treatment of choice if adequate length of the spermatic cord is present [7].

Similarly, rodent studies show that transection of the gubernaculum can lead to accidental transverse ectopia, but in our second case gubernaculum of the controlateral testis was fixed in the opposite iliopubic branch [8]. In this case, the adherence of developing wolffian ducts was the most interesting theory.

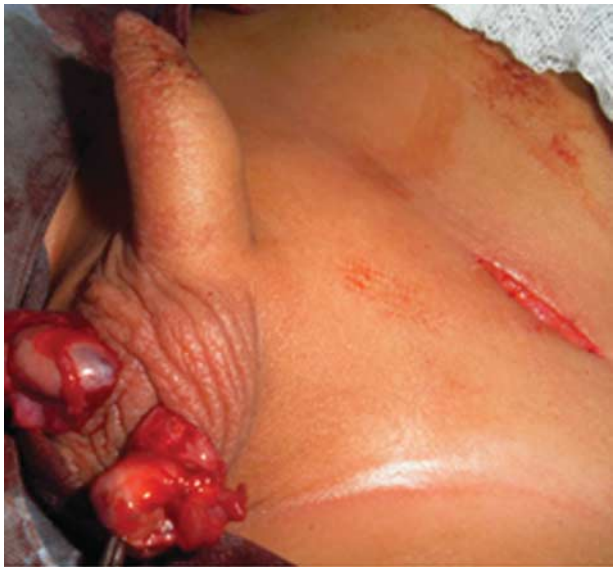
The management of PMDS associated with CTE is still controversial. Many authors prefer testicular biopsies;

Fig. 2



Fallopian tube and testis in lateral view.

Fig. 3



Both testis were placed in separate hemiscrotums, keeping their blood supply intact.

reintroducing Müllerian cavity, testes, and fallopian tube to the abdominal cavity; herniorrhaphy; and reoperating after confirming the sexual orientation (karyotype, age, social). In the case of male orientation, Müllerian cavity will be resected (to prevent mass syndrome and pain at puberty) and both testes will be fixed in dartos. Management of testicular ectopia is either by trans-septal or transperitoneal transposition orchidopexy [9]. An orchidectomy is indicated for testis that cannot be mobilized to a palpable location [7].

Recently, with the experience of laparoscopic surgery for impalpable testis, both diagnosis and management of CTE are possible, as well as the associated anomalies [10,11].

Patients with CTE are at an increased risk of malignant transformation. In fact, the overall incidence of malignant transformation of gonads is 18%. There are no reports of malignancy arising from the Müllerian remnants [4,9,12].

Conclusion

It is important to specify the location of the gubernaculums. We must distinguish between simple CTE and PMDS. Management is still controversial in the form with PMDS most author prefer a conservative approach to this rare syndrome without risking the vas deferens, but we prefer to resect Müllerian cavity to prevent pain and mass syndrome later [13]. Trans-septal fixation is a safe option for contralateral orchidopexy.

Long-term follow-up is needed for the assessment of fertility in these patients.

Conflicts of interest

There are no conflicts of interest.

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