Testicular ectopia: A case report

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Abstract. This paper mainly describes three cases of children with ectopic testis, of which two patients with transverse testicular ectopia (TTE) and one with perineal ectopic testis (PET). All patients who underwent orchidopexy at the same pediatric surgical unit in the Affiliated Hospital of Jining Medical University (Jining, China) between June 2010 and February 2021 were retrospectively evaluated (age range, 14-34 months). A total of two patients (67%) was admitted with asymptomatic unilateral inguinal masses and the contralateral testis missing; the first patient was diagnosed with TTE intra-operatively, whereas the other patient was diagnosed with TTE through physical examination and ultrasound preoperatively. The third patient (33%) was admitted with the right testis missing and a left perineal mass, which was confirmed using PET by physical and ultrasound examination before the operation. The first two patients underwent transseptal orchidopexy, whereas the third patient underwent simple orchidopexy. Postoperative complications were not observed (follow-up, 10-24 months). The low incidence and poor understanding of ectopic testis compels us to report our findings and further discuss this particular disease of testicular ectopia, including its pathogenesis, diagnostic and treatment options.

Introduction

The testes occasionally fail to completely descend along the normal path or may deviate from it. The former condition is called undescended testis and is present in ~4.5% of newborns, with its incidence being higher in preterm babies (1), whereas the latter is known as testicular ectopia (TE). TE is a rare anomaly characterized by the testes lying outside the normal descent route (retroperitoneum to the scrotum). There are several variants of TE, including perineal ectopic testis (PET), femoral and penile ectopic testis, transverse TE (TTE) and preperitoneal and anterior abdominal wall ectopic testis (2). PET is the most common variant (3); in this case, the testis is located in the perineal region, with its final position often being further from the groin than a scrotal testis, and is covered by a perineal fat pad rather than being within the low-temperature environment of the scrotum. Although it is the most common type, PET accounts for <1% of abnormal testicular migration cases, and may be as rare as <1/10,000 cases (4). TTE, also known as crossed TE, is an extremely rare anomaly of testicular descent, during which bilateral gonads migrate along the same inguinal canal toward the scrotum, and it is commonly associated with an inguinal hernia. To the best of our knowledge, <100 cases have been reported in the literature worldwide (5). An accurate preoperative diagnosis is challenging because it is easy to diagnose simple undescended testis in the majority of the reported cases. However, with the improvement of medicine, diagnostic advancements continue to be made. Laparoscopy surgery is the main treatment (6). Laparoscopy is commonly used in the diagnosis and treatment of TTE. The early diagnosis and surgical treatment of ectopic testes facilitate normal development of the genitourinary system and prevent complications. Ultrasonography, laparoscopy and magnetic resonance imaging (MRI) have been used for the diagnosis of this condition (2).

Case report

Case 1. A 4-month-old male patient was admitted to the Department of Pediatric Surgery (The Affiliated Hospital of Jining Medical University, Jining, China) presenting with bilateral absence of the testes. The patient was treated with human chorionic gonadotropin (HCG; each ampoule contains 5,000 IU of HCG injection powder; Nanjing Organon Pharmaceutical Co., Ltd.) 1,000 IU via intramuscular injection twice a week for 5 weeks. The right testicle reached the bottom of the scrotum 8 months later, but the left scrotum remained empty. At 1 year and 2 months of age, the parents of the patient identified an oval mass (diameter, 1.0 cm) in the right inguinal region, which was painless, retractable and gradually increased in size. The ultrasound examination (ZS3 Exp; Zonare) showed the echo of two testes; the epididymis was detected at the right scrotum and external inguinal ring,

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and no testicular echo was identified in the left inguinal or scrotum. A transverse incision of the right inguinal region identified a testis in the right inguinal canal. Bilateral spermatic cords and vessels were carefully dissected, and no fused abnormality or remnant of Mullerian structure was identified. Since the left spermatic cord was long enough to reach the left hemiscrotum, the scrotal septum was opened through subcutaneous tunneling and the left testis was fixed at the bottom of the left scrotum (transseptal orchidopexy). Next, the hernia sac was transected and ligated at a high level. No testicular abnormalities were observed following ultrasonography (ZS3 Exp; Zonare) of the scrotum during the 2-year follow-up.

Case 2. A 2-year and 10-month-old male was admitted to Department of Pediatric Surgery in the Affiliated Hospital of Jining Medical University with a left inguinal mass. On physical examination, an egg-sized mass was felt in the left inguinal groin area after the increase in abdominal pressure, which then retracted automatically with the lack of pressure. The left testis was felt at the bottom of the scrotum, while a testicular substance was found at the level of the left external inguinal ring. No testicular substance was felt in the right hypoplastic scrotum and groin area. The ultrasound indicated that the right scrotum was empty, while the echo of two testes and epididymis were detected at the left scrotum and external inguinal ring (Fig. 1A). During laparoscopic exploration, it was observed that the right spermatic cord and vessels travelled upwards above the bladder and entered the left inguinal canal together with the left ones (Fig. 1B). Laparoscopic (German Wolf) exploration was employed to pull the two testicles down to the scrotum to avoid an incision in the inguinal area, identify the laterality of the two testicles directly and avoid confusing the laterality of the testes during fixation. Remnant Mullerian duct structures and other fused anomalies were not discovered (Fig. 1C). Next, the internal ring was ligated at the extraperitoneal area and transseptal orchidopexy was performed (Fig. 1D). No testicular abnormalities were observed in the ultrasonography of the scrotum following the surgery.

Case 3. A male patient aged 1 year and 3 months came to the Department of Pediatric Surgery with the right testis missing and a left perineal mass. On physical examination, the bilateral scrotum was hypoplastic, the right testis was felt in the ipsilateral inguinal region and the left was in the ipsilateral perineum (Fig. 2A). Ultrasound confirmed the results of the aforementioned examination (Fig. 2B). The left testis was easily discovered in the subcutaneous tunnel and limited dissection of the spermatic cord and blood vessels was adequate to perform orchidopexy. No testicular anomalies were observed via ultrasonography of the scrotum during routine follow-ups (10 months). Differences between the three cases are presented in Table I.

Discussion

The descent of the testes is hypothesized to occur in two phases: Intra-abdominal and inguinal migration. The ectopic testis completes inguinal migration normally but is misdirected outside the normal path of descent below the external ring (7). TE can be an abnormality of testicular development and is caused by several factors, such as persistent Müllerian duct syndrome, androgen disorder, true hermaphroditism and mixed gonadal dysgenesis (8). It can also be an abnormality caused by descent and migration processes, including anomalous insertion of the distal gubernaculum testis (9). It has been hypothesized to result from abnormal fixation or movement of the peritoneum during testicular descent (4). There are several variants of TE, including PET, femoral and penile ectopic testis and TTE.PET is the most common variant of TE where the testis was found in the perineal region (10-12). In perineal ectopia, the testis is not truly undescended, but exhibits malformed cryptorchidism as the testis is not located in the scrotum, its final position is often further from the groin and it is covered by a perineal fat pad, where it benefits from the specialized, low temperature environment (3). There are numerous theories for the pathogenesis of PET, including congenital obstruction of the secondary external inguinal ring and the subsequent migration of the testis to the perineal pouch (12), abnormal interplay between androgen and calcitonin gene-related peptide release from the genitofemoral nerve (10) and aberrant gubernacular stabilization caused by an anomaly at the distal extremity of the gubernaculum (11). These factors may also lead to the occurrence of femoral and penile ectopic testis (2,13). The cases reported in the present study were admitted to the hospital due to an abnormal perineal mass and were diagnosed with PET following preoperative examination. However, the specific cause of the disease was not defined because the hormone and gene levels were not determined.

TTE is an extremely rare congenital anomaly characterized by the presence of two testes in the same hemiscrotum or the descent or migration of two testes along the same inguinal canal (14). TTE is frequently associated with congenital inguinal hernia and less frequently with additional genitourinary anomalies, including hypospadias, seminal vesicle cyst, renal agenesis and persistent Müllerian duct (15). The persistence of Müllerian duct structures has been reported in 20-49% of patients with TTE and is hypothesized to be due to inadequate anti-Mullerian hormone or because the target organ is not sensitive to this hormone (16,17). However, the Müllerian duct structures were not found in either of the TTE cases in the present study. At present, the pathogenesis of TTE is not clear; the mainstream views on its pathogenesis are as follows: Both testes being derived from the same germinal ridge; mechanical effect of persistent Müllerian duct structures preventing testicular descent or causing both testicles to descend toward the same hemiscrotum and defective gubernacular formation (3). No single theory explains the pathogenesis. Due to the high cost, the long duration of genetic testing and the refusal of the parents of the children, gene testing and hormone evaluation are not routinely carried out in clinical treatment. Therefore, regardless of the type of ectopic testis, clarifying its specific pathogenesis is difficult.

A careful physical examination reveals an empty, sagging scrotum. For all children with scrotal cavitation, regardless of the type of ectopic testis, careful clinical examination along the descent path and all potential ectopic testis sites is important. These include the perineum, femoral canal, base of the penis, intraperitoneal cavity, contralateral pouch, inguinal canal and deep ring (13). Imaging models can be used for the diagnosis

Table I. Clinical characteristics of the three cases.

Age at diagnosis	Type of ectopic testis	Location of testes	Presentation	Location of testes (US)	Types of operation	Follow-up/ testicular abnormalities
1 Y, 2 M	TTE	R scrotum L inguinal	R inguinal hernia L undescended testis	R scrotum L inguinal	Transseptal orchidopexy	2 Y/N
2 Y, 10 M	TTE	R inguinal L external inguinal ring	Left inguinal hernia Right undescended testis	R external inguinal ring L scrotum	Laparoscopy-assisted transseptal orchidopexy	1 Y, 4 M/N
1 Y, 3 M	Perineum ectopic	R inguinal L perineum	R undescended testis L perineum ectopic	R: inguinal L perineum	Orchidopexy	10 M/N

TTE, transverse testicular ectopia; US, ultrasound; Y, year; N, no; M, month; R, right; L, left.

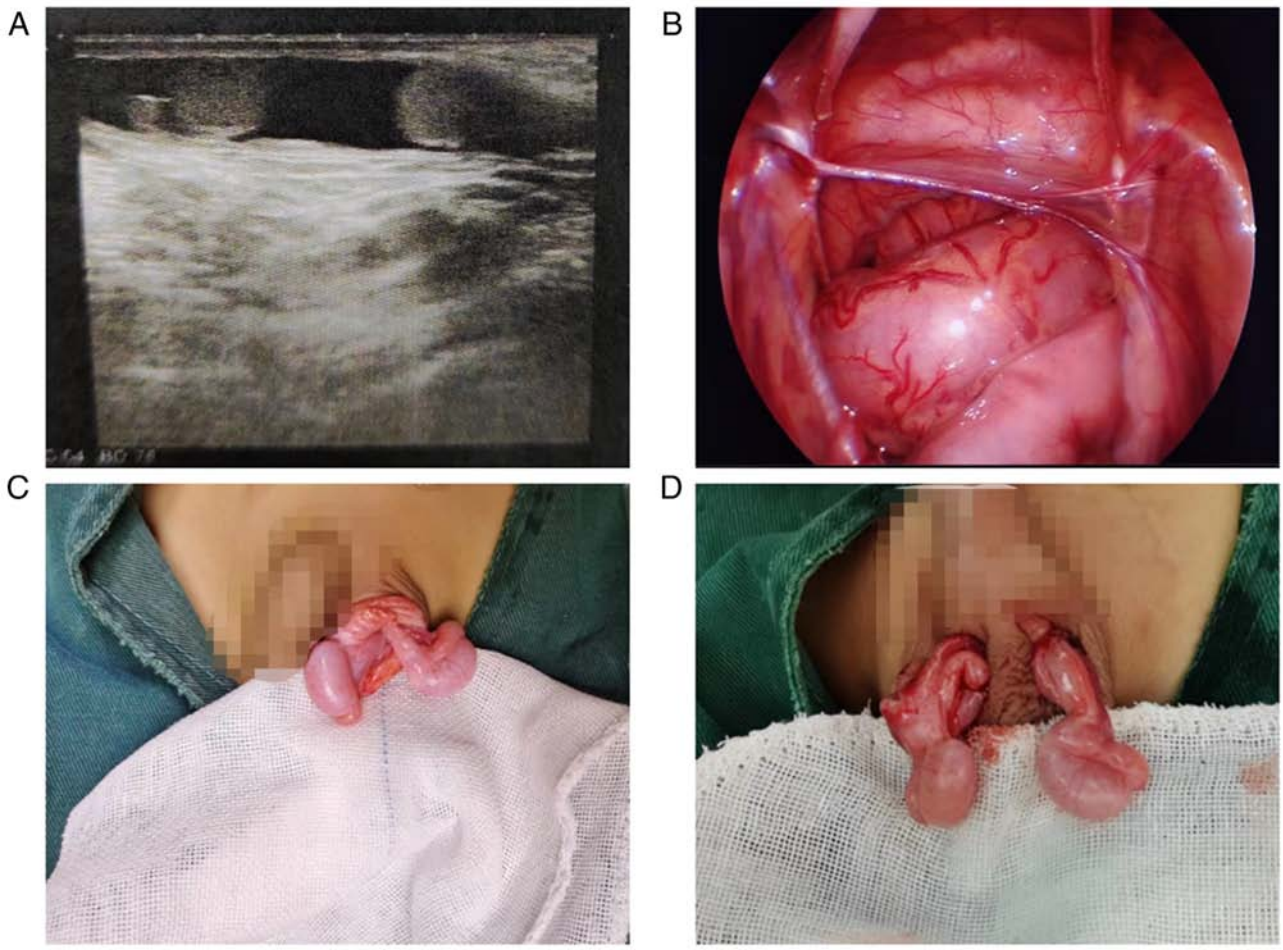

Figure 1. Representative images and echography of case 2. (A) Echography of two testicular echoes at the left scrotum and external inguinal ring. (B) Right spermatic cord and vessels crossed the upper bladder and entered the left inguinal canal. (C) Transverse testicular ectopia. (D) Transseptal orchidopexy.

of TE in cases where physical examination is inconclusive or in children who are uncooperative or who have had a genitourinary surgery. Ultrasound is especially used extensively in diagnostic procedures to avoid ionizing radiation (18). If physical examination and ultrasound cannot confirm this, CT and MRI may be considered to locate the testis before surgery. CT has a relatively high success rate in locating ectopic testes. However, the pediatric population is unique as there is a risk of developing a malignancy due to ionizing radiation; therefore, CT is not routinely used in diagnosis and localization of ectopic testes (19). MRI is an option, as it has an accuracy similar to CT without ionizing radiation exposure, but it is expensive,

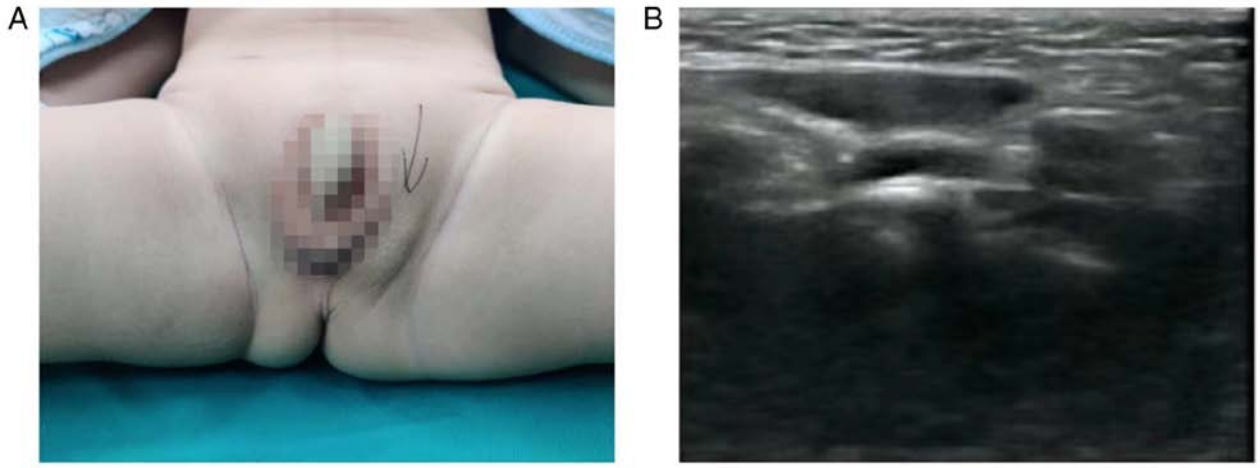

Figure 2. Representative images and echography of case 3. (A) Palpable perineal mass in addition to the left hypoplastic scrotum. (B) Bilateral empty scrotum and perineal echography

may not be readily accessible and typically involves a form of sedation or anesthesia (20). Diagnosing PET based on an empty scrotum and swollen perineum is easier (21). However, the preoperative diagnosis of TTE is difficult; the diagnosis of the first TTE case in the present study was determined intraoperatively. Due to this complexity, TTE is often misdiagnosed as simple cryptorchidism, testicular tumor and single testis deformity. The presence of ectopic masses can be detected by imaging diagnostic techniques, but the specific nature of the masses cannot be clearly defined in most cases (2). Therefore, the diagnosis of TTE is often made intraoperatively during inguinal hernia repair (22). The first case in the present study began with HCG therapy as simple cryptorchidism; although the most recent guidelines recommend the surgical management of a congenital undescended testis, HCG hormone, has been widely used for the treatment of congenital undescended testes since 1930 (23,24), but the treatment of congenital undescended testes with HCG hormone has low success rates; it may, however, be effective in high scrotal testes (25). Since the present case was not a simple cryptorchidism, a complete cure was not achieved.

TE is associated with complications, such as trauma, torsion and bilateral infertility (26). Therefore, surgical treatment is necessary and early surgical intervention is important for long-term prognosis. The aim of treatment is the fixation of the testes into the respective hemiscrotum (27) to restore optimal development environment, improve the later development potential and decrease incidence of malignant transformation. Surgical correction is recommended at 1 year of age, as cryptorchidism may show definite histological changes (28). It is reported that it is not reasonable to expect TTE to have a natural recovery, so it is very important for surgical treatment to be performed as soon as possible following the detection of TTE (29,30). The optimal time of surgery is at 1-2 years of age, and the age of operation tends to decrease continuously. The treatment principles of TTE are to preserve fertility, decrease risk of testicular neoplasia and repair congenital anomalies, hernia and orchidopexy (6,31). Common surgical methods for TTE treatment include transseptal, extraperitoneal orchidopexy, testicular anatomical reduction, Fowler-Stephens primary or Stage descent orchidopexy and *in situ* orchidopexy, testicular transplantation or orchiectomy (32). Both TTE cases in the present study underwent transseptal orchidopexy. For PET, orchiopexy is recommended and surgical intervention should be performed early, regardless of the existence of an inguinal hernia, as there is no clear benefit to delaying the procedure until the child is ≥6 months old (33). Orchidopexy is an option for children <2 years old. The PET cases in the present study were treated using simple orchidopexy; however, if atrophic testes were identified, orchiectomy should be performed, which may effectively reduce the occurrence of long-term testicular malignancies (34). Orchidopexy, which is suitable for PET and femoral and penile ectopic testis, is usually easy to perform, as long as it is the length of the testicular vessels and vas deferens is adequate (35). The traditional method of testicular fixation via scrotal incision is no longer recommended for the treatment of ectopic testes alone due to limitations such as testicular position, spermatic cord length and inability to observe other abdominal malformations (4). The minimally invasive laparoscopic technique achieves great visual assessment of the abnormal testicular development situation, spermatic cord length, vas deferens and ectopic spermatic cord, presence of Müllerian tube residues and deformity of other abdominal organs.

In conclusion, TE is a rare abnormality and its etiology remains unclear. It is characterized by the testes being outside the normal descent path. There are several variants of TE, including PET and TTE. PET is the most common variant and can be detected by physical examination. However, preoperative diagnosis of TTE is difficult and is usually achieved using imaging or intraoperatively. Ectopic testes should be treated surgically at an early stage, often with orchidopexy or transseptal orchidopexy, to ensure good postoperative recovery; however, the overall management of ectopic testes, long-term follow-up and possible complications need further study.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors' contributions

JC and MG collected data and drafted the manuscript. HL, LS, NH and CZ performed data analysis. LS, JS, CH and NH performed minimally invasive surgery. CH, JC, MG and LS confirm the authenticity of all the raw data. All authors have read and approved the final manuscript.

Ethics approval and consent to participate

The present study was approved by the Ethics Committee of the Affiliated Hospital of Jining Medical University (approval no. 2023-01-C015).

Patient consent for publication

The parents or legal guardians of all the children involved in the present study provided written informed consent for publication.

Competing interests

The authors declare that they have no competing interests.

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