Perineal Ectopic Testis- A Rarity In Pediatric Surgical Practice

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Abstract
Perineal ectopic testis (PET) is a rarity in pediatric surgical practice seen in less than 10% cases of maldescended testis. An eighteen months old boy presented with empty right scrotum and a palpable soft mass in the perineum which on preoperative ultrasonography was consistent with testicular morphology. Orchidopexy was performed by standard operative technique and testis was placed in Dartos Pouch. The case is reported to highlight the importance of examination of ectopic sites in a cryptorchid child and the need for long term follow-up of patient.

Key Words
Undescended Testis, Ectopic Testis, Perineal Ectopic Testis, Orchidopexy

Introduction
An ectopic testis (ET) is a testis located in an aberrant position out of the path of normal descent. It completes normal transinguinal descent but is misdirected outside the path of descent below external ring. The first case was reported by John Hunter in 1786(1). ET occurs in only about 5% of the cases of empty scrotum (2). The most common sites in descending order are the superficial inguinal pouch (Denis Brown Pouch), perineum, femoral canal, contralateral scrotum and prepenile region (2). Though it is the commonest form of true testicular ectopia excluding the superficial inguinal pouch, PET is a rare anomaly seen in approximately 1% of all cases of testicular maldescend (3). PET is more prone to trauma, torsion, infertility and malignancy. Early surgery is therefore recommended (4). The present case is reported to highlight the importance of thorough physical examination of ectopic sites including perineum in cases of testicular maldescend, relatively easiness of the operative procedure and need for long term follow up of such patients to look for complications.

Case Report
An eighteen months old boy was brought to the surgical outdoor with history of absent right testis from scrotum since birth. Examination revealed a healthy looking child with poorly developed and empty right scrotum. The left testis was present in the scrotum and was of normal size. There was no bulge or history suggestive of hernia in the right inguinoscrotal area and spermatic cord structures were not discernible. An oval shaped soft mass identical to testis, measuring 15mm x 10mm was palpable just below and lateral to right hemiscrotum corresponding to the clinical examination. Ultrasound revealed left testis measuring 15mmx8mm lying in left hemiscrotum and right testis measuring 15mmx9mm located outside the scrotum in the perineum corresponding to the clinical examination. Hence, a diagnosis of right PET...
was made. All preoperative investigations were normal and patient was explored through standard inguinal crease incision. Spermatic cord structures were mobilized, gubernaculum attachments with the perineal tissue were divided and testis was delivered into the wound. It was of normal size with an associated hernial sac and adequate length of the vas and vessels (Fig. 3). Herniotomy was performed and testis was placed in the ipsilateral hemiscrotum creating a standard Dartos Pouch. Postoperative period was uneventful. Follow up after 3 months revealed rights testis present in the scrotum.

Discussion

Testicular development and descent from the abdomen to scrotum is a complex and multistage process which starts from 7th week and is completed by 35th week of gestation (5). Testicular descent can be described in two phases: transabdominal and inguinocrural. During the inguinocrural phase, the testis may deviate from the normal path of descent and migrate to an abnormal position, described as ET. Different hypothesis have been proposed regarding etiologies of ET. It is presumed to be either the result of hormone imbalance between androgen and calcitonin gene related peptide (CGPR), or result of aberrant gubernaculum stabilization due to anomaly at its distal end or possibly caused by local mechanical obstacle blocking the normal descent (5). Lookwood postulated that gubernaculum has several inferior insertions and scrotal insertion is dominant in normal descent. If one of the other insertions is stronger and larger, the testis will be pulled in that direction and become estopic (6). Middleton el all suggested that increased intra-abdominal pressure facilitates normal descent and gubernaculum has an important guiding role which if inadequate, could lead to testicular ectopia (7). Five major sites of ectopia are perineum, femoral canal, superficial inguinal pouch, suprapubic area and contralateral pouch. In addition to these sites, preperitoneal, extracorporeal and anterior abdominal wall ET have also been reported (8). PET is seen very rarely and approximately 175 cases have been reported in the literature and 80% of these are unilateral (4). Bilateral perineal ectopia is even scarcer (9). An empty scrotum with soft perineal mass on ipsilateral side is very suggestive of PET, as were the findings in the child under report which was also substantiated by preoperative ultrasound findings. Some cases of ET diagnosed on ultrasound antenatally at 38 weeks and confirmed in postnatal period by clinical examination have also been reported (10). ET are thought to be at greater risk of trauma, torsion, infertility and malignancy. But due to the low incidence of the anomaly, there is not enough evidence available in literature showing such risk in long
term follow up of PET (2). Surgical correction of undescended testis is generally done at about 6 months of age to allow for spontaneous descent to occur. However, in case of coexisting inguinal hernia, early intervention has been advised to prevent incarceration. It has been suggested that there is no need to delay surgery in ET even if it is not associated with hernia because possible descent as seen in undescended testis will not occur (3) and attempts to move ET into the scrotum with hormone therapy has been found to be ineffective (5). PET is usually explored through the standard inguinal approach as was performed in the present case. The testis can be placed in the scrotum easily because spermatic cord and vessels are sufficiently long as was observed in our patient and reported by other authors (3). The functional outcome is difficult to define but has been found to be similar to other forms of maldescended testis (11). Most authorities agree that testicular cancer is more common in misplaced testis than in normally descended testis. Currently, however there are no satisfactory reports on whether the risk of testicular cancer in ET is higher than that in cryptorchidism (12). Thus education about testicular self examination and long term follow up of these patients is just as important as the operative procedure.

Conclusion

PET is a rare anomaly in pediatric surgical practice and is easy to diagnose. The correction is not difficult, as in other cases of maldescended testis, due to long length of vascular pedicle. Timing of surgery can be individualized without unnecessary delay and long term follow-up is advised to look for possible complications in such patients which can contribute further to the literature on the subject.