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Ectopic Perineal Testis With Contralateral Undescended Testis With Bilateral Scrotal Hypoplasia: Rare Case Report

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Abstract

Undescended testis is a very common presentation in male pediatric population but testis at ectopic sites is a very rare pathology. One of the ectopic sites is perineum and it accounts for only 1% of all cases of undescended testis. Perineum should especially be examined in a case where testicle could not be palpated scrotally. Diagnosis is usually straightforward through a physical examination of the ectopic regions and an empty scrotum on the same side. it is predominantly identified during childhood, with its occurrence being even rarer in adults and young boys. The recommended and widely accepted treatment for perineal ectopic testis is open orchiopexy.1
We came across an 8 months old child with bilateral empty scrotum. On physical examination right sided testis found in inguinal region while left testis found in perineal region. Patient underwent surgical exploration bilaterally through inguinal incision and bilateral orchidopexy was done.

Keywords: Ectopic testis; Orchidopexy; Perineal ectopic testis; Undescended testis

Introduction

Testicular maldescent is the most common anomaly of the genitalia seen in 0.2–1% of all men. Most of these testes get arrested along its normal pathway, resulting in abdominal, inguinal or high scrotal testis. About 5% of undescended testis are ectopic. The most common ectopic sites in descending order are superficial inguinal pouch, perineum, root of penis, femoral triangle/upper thigh and contralateral scrotum[2]. Perineal ectopic testis is a rare encounter in pediatric surgery accounting for about 1% of all cases of undescended testis.[3]

Relocation of testis in an atypical site after going through the inguinal canal and leaving it through the external ring is named ectopic testis. The most widely recognized aberrant locations are superficial inguinal pouch perineum, femoral canal, contralateral scrotum and prepenile area. Ectopic testis happens in just 5% of instances of empty scrotum. This paper aims to highlight the significance of doing perfect clinical examination in every case of testicular maldescent or empty scrotum looking for ectopic sites of testicular positioning.[4]

Case Report:

A 2 month old term male presented to the surgery clinic for evaluation of a bilateral empty scrotum. The patient's prenatal history was unremarkable. The

parents reported that they had never been able to feel the testicles in scrotum. On physical exam, the right testicle was palpated in the right inguinal region (Fig 2) and the left testicle was palpated in the perineum (Fig. 3). The patient was advised to wait and watch for spontaneous descent up to six months which was not successful. Then subsequently underwent an open bilateral orchidopexy at 8 months of age, which was performed through an inguinal approach. On right side testis found in right inguinal canal. Once the testicle was delivered through the incision, the patient was found to have an open processus vaginalis on right side. Herniotomy was performed and after adequate mobilization of the cord structures testis placed in right scrotum and fixed in dartos pouch. While on left side the gubernaculum was noted to be attached to the perineum and not the scrotum. After adequate mobilization of the cord structures through inguinal incision, the testicle was delivered and was then placed in left scrotum within a dartos pouch. The patient recovered appropriately from the procedure without complication.

Discussion:

Cryptorchidism is defined as the absence of one or both testes in the scrotum. Spontaneous descent may occur during the first 6 months of life and only 0.8% of boys are affected by 12 months of age. In most cases of cryptorchidism, the testes have incompletely traversed the normal pathway of descent. However, they can also migrate to an ectopic position or have vanished as a result of torsion or a vascular accident.

The indifferent gonad develops adjacent to the rudimentary kidney and descends trans abdominally beginning at 5 weeks gestation. By 10 weeks, it is in close proximity to the internal inguinal ring, and by 12 weeks the gubernaculum is visible as it begins its swelling phase. The swelling of the gubernaculum, in addition to development of the cremaster muscle and migration of the processus vaginalis, are thought to produce widening of the inguinal canal necessary for testicular passage. Between 20 and 28 weeks the testis passes through the inguinal canal, with further caudal descent into the scrotum occurring up until birth. This is accompanied by regression of the gubernaculum.

The testicle becomes ectopic when it deviates from the normal pathway of descent. Possible ectopic locations include the superficial inguinal pouch, perineum, penis, lateral to the scrotum, pubic region, thigh or contralateral scrotum. However, perineal ectopic testes are rare and have been estimated to comprise only 1% of all cases of undescended testes. There is no statistically significant difference in the rates of patent processus vaginalis and epididymal abnormalities in ectopic testes compared to undescended testes irrespective of location. The testicular histopathology of these two groups has also been shown to be similar. These findings suggest they may have a similar etiology, which is thought to be multifactorial but has not been fully elucidated. Risks associated with both ectopic and undescended testes include torsion, as well as an increased incidence of infertility, trauma and malignancy. Associated anomalies include testis-epididymis dissociation and inguinal hernias.

The assessment of a neonate with an undescended testis should begin with a thorough history, including risk factors such as prematurity and low birth weight, followed by a physical exam. Findings on exam, particularly unilateral versus bilateral undescended testes and whether an undescended testis is palpable or non-palpable, guide any additional workup and treatment. The treatment for ectopic testes is surgical and, as opposed to testes located in the normal pathway of descent, there is no need to wait for descent. Open orchidopexy is performed through an inguinal incision to allow for concomitant treatment of an inguinal hernia if present.[5] This remains the optimal choice for management, even in the setting of a delayed diagnosis. Patients and their parents should be counselled regarding the increased risk of subfertility and malignancy and taught to perform self-examinations after puberty.[6]

Conclusion:

Similar pathological findings in ectopic and undescended testes as well as the association of ectopic testis with a contralateral undescended testis suggest that ectopic and undescended testes are variants of the same congenital anomaly. Thus, boys with ectopic testis may have an increased incidence of subfertility and testicular malignancy. Surgical intervention should be the gold standard treatment for perineal ectopic testis cases.

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Figure 1 showing preoperative bilateral empty scrotum

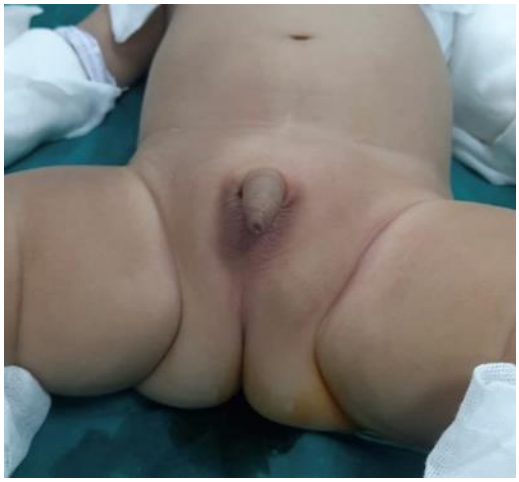

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Figure 2 showing right undescended testis in inguinal canal

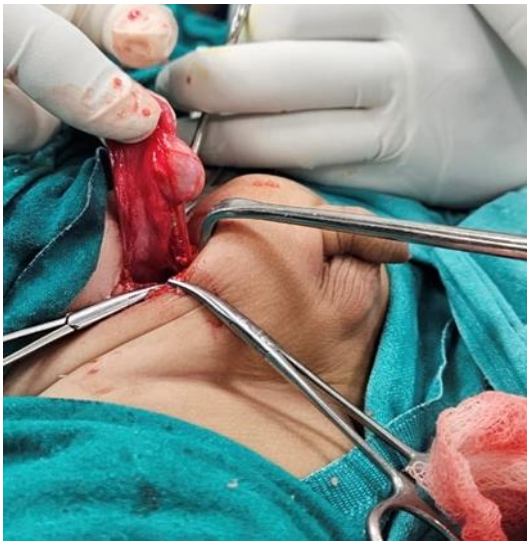

Figure 3 showing left testis attached in perineal region with right testis fixed in dartos pouch in right scrotum

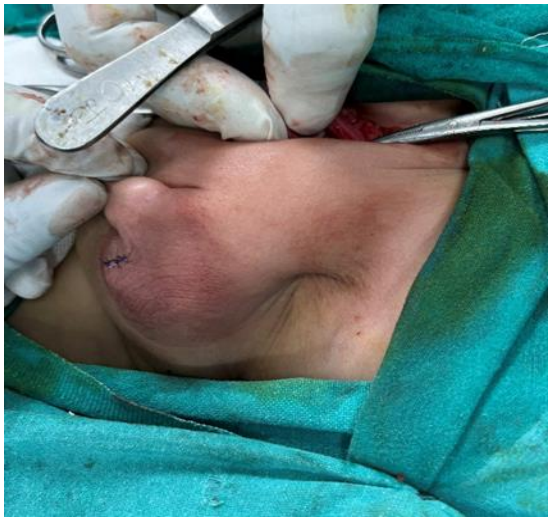

Figure 4 showing left perineal testis after mobilization of cord structures

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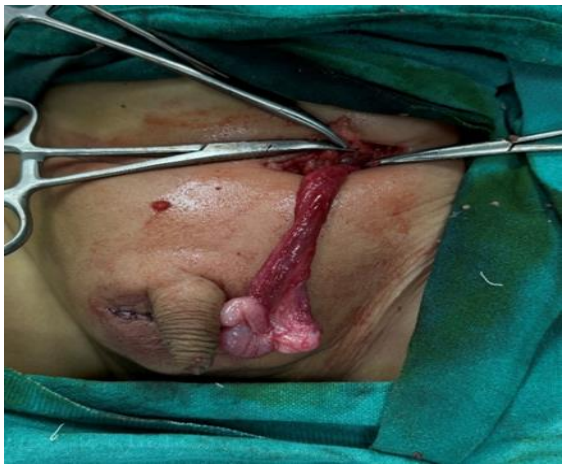

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