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Bilateral ectopic femoral testes: A rare cause of empty scrotum

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ABSTRACT

Empty scrotum is a relatively rare entity, with few cases reported in the literature, so far; if it coexists with bilateral ectopic femoral testes, it then constitutes an extremely rare congenital abnormality. We report a case of empty scrotum revealed at the first physical examination of a neonate. The scrotum appeared empty and no testes could be palpated in the scrotum or the inguinal canal. Two solitary, oval masses were palpable laterally of each hemiscrotum, close to femoral canal and the diagnosis of bilateral femoral ectopic testes was confirmed by the consultant pediatric surgeon.

Introduction

Empty scrotum is a rare congenital abnormality. When such a diagnosis is made, it is crucial to identify if the testes are palpable, as clinical examination still remains the most important diagnostic procedure to locate them. Testes may be found anywhere along their normal descent, resulting in inguinal or suprascrotal testes.[1] This localization and the inability to bring the testis down the scrotum, defines the true undescended testis. If gentle maneuvers can bring them to the scrotum, they are then categorized as retractile testes.[2] Physical examination should also include possible sites of ectopy such as the perineum, the femoral canal, the prepenile area, the opposite scrotum and the superficial inguinal pouch.[2] When no testes can be palpated in any of these areas, one should consider not only impalpable true undescended intra-abdominal or intracanalicular testes, but also an intra-uterine regression of the testes (vanishing testis), bilateral agenesis of the testes, congenital adrenal hyperplasia in a genetically female newborn (46 XX), disorders of sexual development (DSD) and endocrinopathies such as primary or secondary hypogonadism, complete and partial androgen insensitivity, persistent müllerian duct syndrome (PMDS), disorders of antimüllerian hormone [AMH]/receptor, androgen biosynthesis defect etc.[2] Diagnostic investigation in these cases includes imaging studies such as U/S, CT or MRI,[3] diagnostic laparoscopy, hormonal examinations such as hCG stimulation study, measurement of LH, FSH, serum 17-hydroxyprogesterone, urinary 17-ketosteroids, testosterone, müllerian inhibiting substance and karyotype analysis.[2]

Case

A male infant was born by caesarian section for no progress of labor at 40^{+1} week of gestation by a primigravida mother; pregnancy was uneventful and antenatal follow-up was normal. Maternal medical history included high levels of WBC at 12.000 since the age of ten years, with no specific diagnosis. She also had elevated anti-Tg without any biochemical or clinical signs of hypothyroidism. Apgar scores were normal and birthweight was 4180gr. At the first clinical examination the two hemiscrotums appeared empty, poorly developed and with fewer folds than normally anticipated for his gestational age (Fig. 1). No testes could be palpated in the scrotum or in the inguinal canal. Two palpable fixed masses were detected outside of the two hemiscrotums at the femoral canal (Fig. 2). No other genitourinary or congenital stigmata were evident. A pediatric surgeon confirmed by palpation the diagnosis of bilateral femoral testicular ectopia, thus an ultrasound scan wasn't required (Fig. 3).

A follow-up appointment and an ultrasound scan of the testes has been scheduled at the age of six months. Furthermore, the father and his paternal uncle were operated during infancy for reported "unilateral cryptorchidism".

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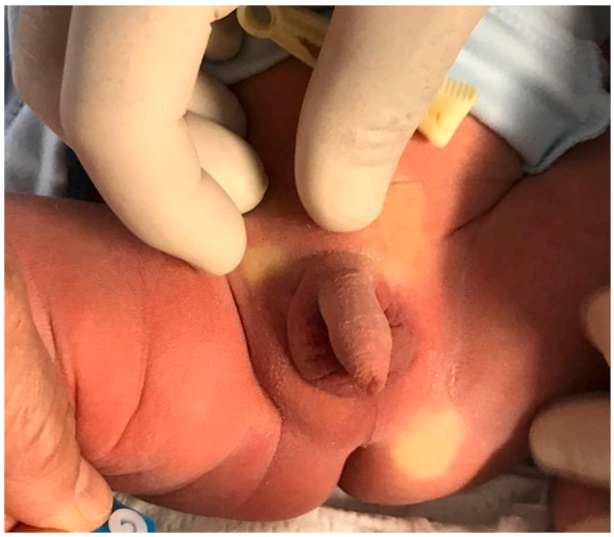

Fig. 1. Bilateral palpable fixed masses lateral to both hemiscrotums.

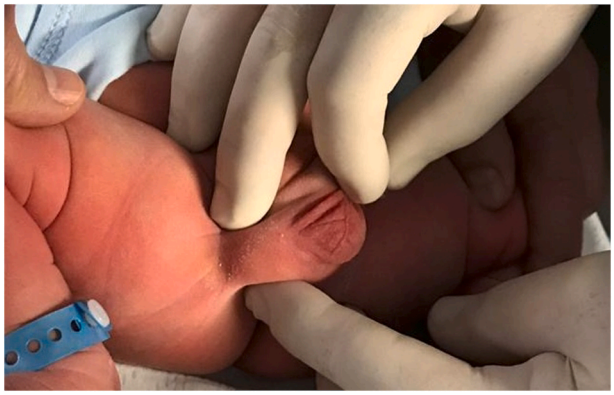

Fig. 2. Right femoral ectopic testis.

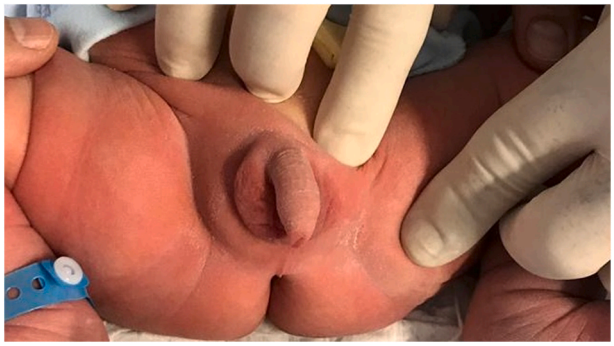

Fig. 3. Left femoral ectopic testis.

Discussion

Development and descent of the testes is a complicated procedure affected and guided by genetic, hormonal, and structural factors. In approximately 5% of the cases of cryptorchidism, the testes are dislocated below the external ring in various sites (ectopic testis), although they descend normally through the inguinal canal (1). The five main sites of ectopic testis are the perineum, the femoral canal, the suprapubic area, the opposite scrotal compartment (transverse testicular ectopia-TTE) and the superficial inguinal pouch. Ectopic testes often remain undiagnosed or are falsely diagnosed as cryptorchidism or anorchia. The femoral testis is located in the femoral canal; it is an extremely rare congenital abnormality with very few reported cases in the literature.[1]

Occasionally, imaging techniques such as ultrasound, CT or MRI may be required for the confirmation of the diagnosis. Ultrasound is generally overused and its capability of confirming or altering an accurate diagnosis and management in case of ectopic testes is fairly questionable with a poor sensitivity of 45% and specificity of 75%.[3] CT and MRI are rarely performed and do not eliminate the need for surgery in ectopic testes; may be indicated when they cannot be palpated (3).

The etiopathogenesis of ectopic testis is controversial (4). Hypotheses include a congenital obstruction of the 'secondary external inguinal ring' with a subsequent dislocation of the testis to an ectopic location, as described by McGregor in 1929. A misplaced gubernacular attach caused by an anomaly at the distal extremity of the gubernaculum, and following attachment of the gubernaculum to an ectopic area is also a possible explanation. As far as the genitofemoral nerve (GFN) is concerned, many theories have been developed, in order to clarify its role in ectopic testes cases. An abnormal interaction between androgen and calcitonin gene-related peptide (CGRP) released from the GFN, misplaced fibers of the GFN, or production of CGRP at the wrong branch of GFN are possible explanations.[4]

Ectopic testes are prone to trauma, testicular torsion, sub- or infertility and malignancy.[2] Similar pathological findings that are found in both ectopic and undescended, such as decreased germ cell numbers, low volume of the testis, persistence of processus vaginalis, anomalies of the epididymis and association with a contralateral cryptorchid testis, suggest that ectopic and undescended testes are variations of the same congenital abnormality.[1]

There is no evidence to delay surgery in ectopic testes though, as they can easily be diagnosed by physical examination in the neonatal period and any spontaneous descent is highly unlikely to occur.[5] Surgery is advised even if there is no hernia present and can be performed between 6 months and 1 year of age. Orchidopexy is the standard procedure as the cord structures are usually of normal length.[5] If the testis is severely atrophic, orchidectomy could also be a choice of approach. Attempts to relocate the ectopic testis into the scrotum with hormonal therapy had no clinical effectiveness.[1] Long term follow-up is vital because of the possibility for malignant transformation and infertility.

Conclusion

Physical examination of neonate males with an empty scrotum should be thorough and must include possible sites of ectopic and undescendent testes. An empty scrotum with no indication of a testis in the inguinal canal and with a palpable fixed lateral mass could be a clinical sign of ectopic testis in the femoral canal. Orchidopexy is the procedure of choice.

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