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Clinical case

Congenital testicular exteriorization by scrotal defect or "scrotoschisis": about 2 cases and review of the literature

Extériorisation testiculaire congénitale par défect scrotal ou « scrotoschisis » :
à propos de 2 cas et revue de la littérature

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Résumé

Le « scrotoschisis » ou extériorisation des testicules par un défect scrotal congénital est une malformation uro-génitale rare. Une vingtaine de cas ont été rapportés dans la littérature depuis sa 1ère description en 1963. A travers ces deux observations nous nous proposons de rapporter notre 1ère expérience sur la prise en charge de cette anomalie rare des organes génitaux externes et de faire une revue de la littérature.

Cas clinique : IL s'agissait pour le 1er patient d'un nouveau-né à H10 de vie, pesant 3200grs, sans antécédent pathologique connu issu d'un mariage consanguin reçu pour malformation scrotale. La mère âgée de 19 ans, est 2e geste- 2e pare et la grossesse menée à terme, était mal suivie avec un accouchement normal, eutocique. La malformation découverte à la naissance était une extériorisation complète des deux testicules à travers un défect scrotal. Après un bilan clinique et paraclinique qui était normal, le traitement fut chirurgical sous anesthésie générale avec des suites simples. Après un recul de 6 mois aucune complication n'était observée.

Quant au 2e patient, il s'agissait d'un nouveau-né à J1 de vie admis pour défect scrotal droit congénital avec extériorisation du testicule. L'enfant était issu d'un mariage consanguin, d'une grossesse mal suivie et d'un accouchement eutocique à domicile assistée par une matrone. Une extériorisation du testicule droit était notée à travers un défect scrotal unique. Le diagnostic d'un scrotoschisis droit était retenu et le bilan malformatif n'a révélé aucune malformation associée. Le traitement était chirurgical avec parage, orchidopexie et suture cutanée. Les suites étaient simples et l'évolution à long terme n'a relevé aucune complication.

Conclusion : Le scrotoschisis est une malformation uro-génitale rare. Le traitement conservateur permet d'obtenir des meilleurs résultats, néanmoins un suivi à long terme est nécessaire afin de déterminer des séquelles fonctionnelles.

Mots-clés : défect scrotal congénital, extériorisation testicules, orchidopexie, Niger.

Abstract

Scrotoschisis, or externalization of the testicles through a congenital scrotal defect, is a rare urogenital malformation. Some twenty cases have been reported in the literature since its 1ère description in 1963. With these two observations, we propose to report our 1ère experience in the management of this rare anomaly of the external genitalia, and to review the literature.

Clinical case: The 1er patient was a newborn at H10 of age, weighing 3200grs, with no known pathological history, born of a consanguineous marriage received for scrotal malformation. The mother, aged 19, is 2e gesture- 2e pare and the pregnancy, carried to term, was poorly monitored with a normal, eutocic delivery. The malformation discovered at birth was a complete externalization of both testicles through a scrotal defect. After a normal clinical and paraclinical work-up, the patient was treated surgically under general anaesthesia, with simple follow-up. After 6 months, no complications were observed.

The 2e patient was a newborn admitted at 1 day of age with a congenital right scrotal defect and externalization of the testicle. The child was the product of a consanguineous marriage, a poorly monitored pregnancy and an eutocique home delivery assisted by a matron. Externalization of the right testicle was noted through a single scrotal defect. The diagnosis of right scrotoschisis was accepted and the malformative work-up revealed no associated malformation. Treatment was surgical with trimming, orchidopexy and skin suture. The postoperative course was straightforward, with no complications noted in the long-term evolution.

Conclusion: Scrotoschisis is a rare urogenital malformation. Conservative treatment offers the best results, but long-term follow-up is necessary to determine functional sequelae.

Keywords: congenital scrotal defect, testicular exteriorization, orchidopexy, Niger.

Introduction

Externalization is the spontaneous emergence of an organ through an acquired or congenital defect. In the urogenital form, the testicles are the most affected. The congenital defect is located in the scrotum, hence the name "scrotoschisis". It is an exceptional condition, with only around twenty cases reported in the literature since its 1ère description by Von Der Leyen in 1963 (1-5). Little is known about its etiology. We report our experience of 2 cases in the pediatric surgery department of the Hôpital National Amirou Boubacar Diallo, Niamey.

Clinical cases

Case n°1:

A male newborn at H10 of age with no known pathological history from a consanguineous marriage was admitted to the emergency department with a scrotal malformation discovered at birth. The patient is the 2e of two siblings, of whom the 1er 2-year- old child is in apparent good health. The mother was a 19-year-old housewife with two pregnancies and 2 parities. The poorly monitored pregnancy was carried to term with 2 antenatal visits. Delivery was eutocic, in a health facility with good adaptation to extrauterine life. Transport was non-medical, with no dressing of lesions. The newborn was in good general condition, apyretic with normal vital parameters. Birth weight was 3200grs, head circumference 32cm, height 46cm, APGAR score 8 then 10 at 5e and 10e minute. Male external genitalia, normal-looking uncircumcised penis, presence of an anterior scrotal defect approximately 3cm in diameter located below the penoscrotal junction. Bilateral exit of the testicles covered with a fibrinous deposit, with the left one completely prolapsed, with a pudgy, inflammatory appearance (fig 1). The hernial orifices were free, and physical examination of the other apparatus revealed no associated abnormalities. Biological tests were normal and abdomino-pelvic ultrasonography for associated malformations

revealed no particularities. The diagnosis of bilateral scrotoschisis was retained and lavage, reintegration and orchidopexy were indicated.

Surgical exploration under general anaesthesia confirmed the 5 x 2cm median scrotal defect with regular margins, the testicles were well stained with no signs of necrosis or perforation, and of normal size and consistency. Orchidopexy with vicry 3/0 after lavage with isotonic saline was performed, followed by skin reintegration and suturing (fig 2 A). Antibiotic therapy with C3G and local care were given for 5 days. The post-operative course was straightforward, and the patient was discharged on postoperative day 5. After 6 months, a follow-up testicular ultrasound showed testicles and epididymides in scrotal position, of homogeneous size and normal echo structure (fig 2 B).

Case n°2

An infant was admitted to the emergency department on D1 for a congenital scrotal wound. The pregnancy had been poorly monitored at term, with 3 antenatal consultations and 2 obstetric ultrasounds. The diagnosis was made at home after an eutoctal vaginal delivery of a male newborn. The parents consulted a health center, which referred her for better care. This was a non-medical transport without patient conditioning. The testicles were not covered with bandages. On admission, the birth weight was 2400 grs. The child was apyretic, conscious, with good mucosal and tegumental coloration, good tone and archaic reflexes. Physical examination revealed male external genitalia with the right testicle fully externalized through a scrotal orifice. The testicle was pudgy and covered with false membranes. The left testicle was intra-scrotal, with a swollen, shiny scrotum (fig. 3A). Examination of the other systems revealed no associated malformations. Biological and biochemical tests were normal. The indication for exploration under general anaesthesia and reintegration was accepted (fig 3B). Abdominal, pelvic and cardiac ultrasound revealed no associated malformations. Parenteral antibiotics were administered for 5 days. Discharge was authorized on day 5 of hospitalization. A follow-up ultrasound at 3e months revealed no abnormalities.

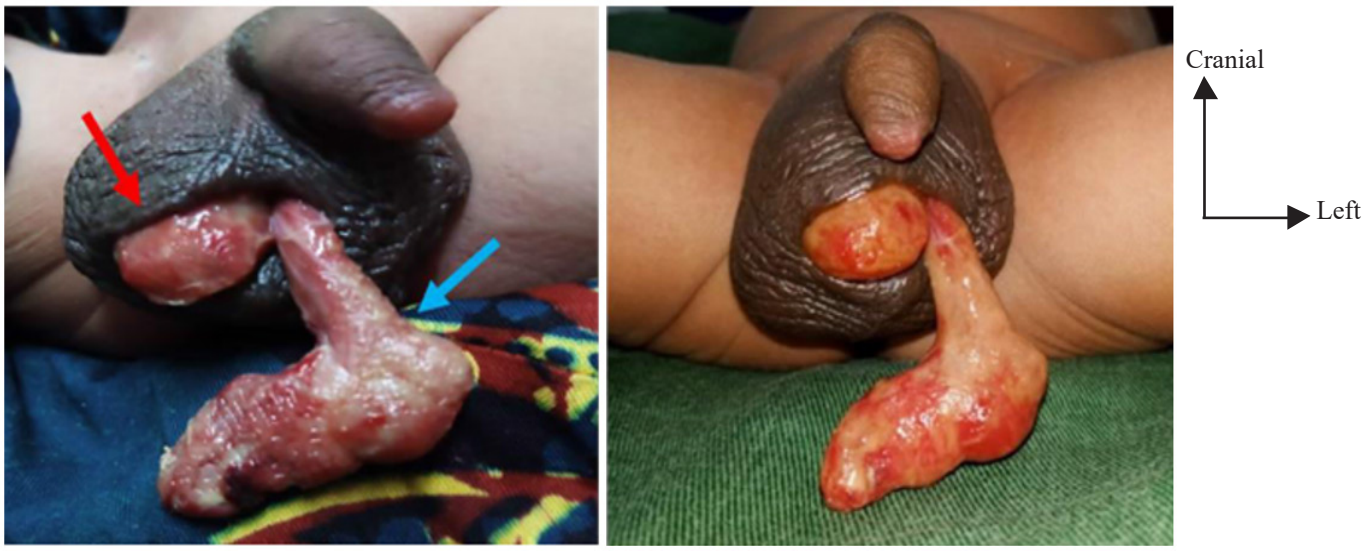

Fig 1: Congenital scrotal defect (red arrow) with bilateral testicular exteriorization. The left testicle and its completely prolapsed cord (blue arrow) *(source: pediatric surgery department/HNABD-Niamey).*

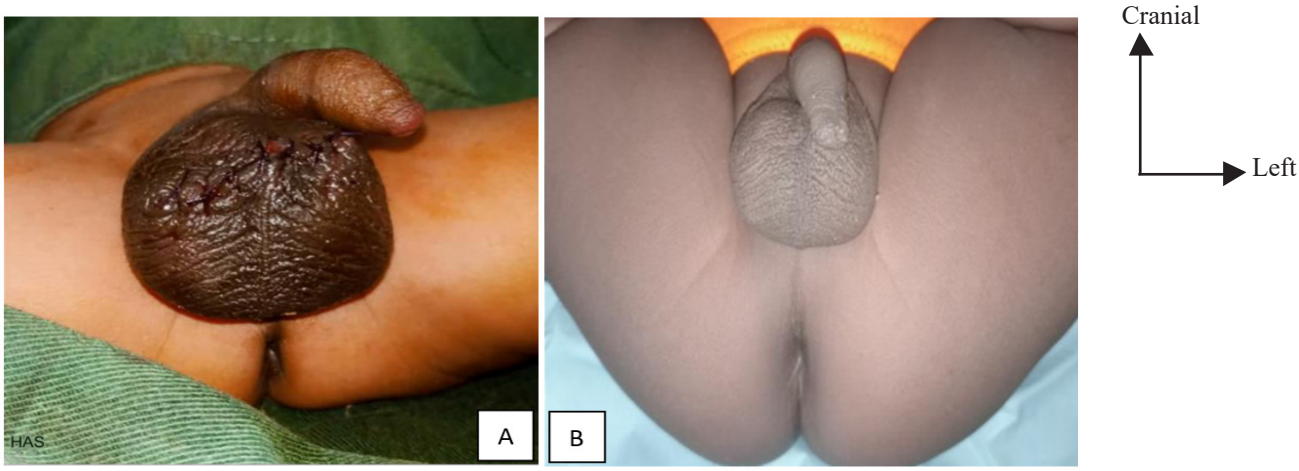

Fig 2: A: immediate post-operative appearance B: physical appearance after 6 months of evolution *(Source: pediatric surgery department/HNABD-Niamey)*

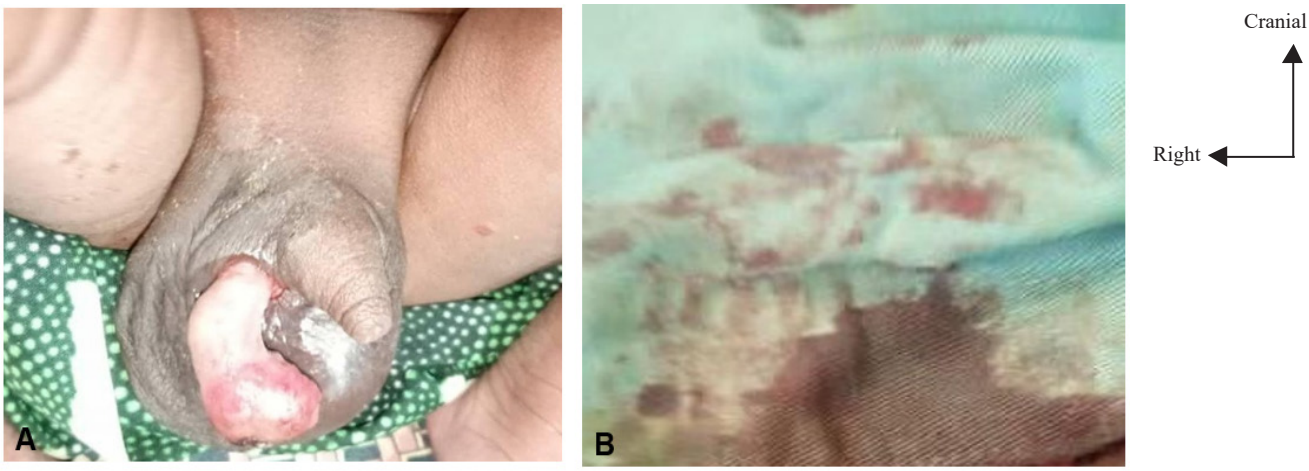

Fig 3: (A): Scrotal defect with complete externalization of the right testicle and cord; (B) immediate post-operative appearance after trimming, reintegration and skin suture. *(Source: Department of Pediatric Surgery/HNABD-Niamey)*

Discussion

Testicular descent anomalies are frequent in pediatric urology, dominated by testicular ectopy and cryptorchidism. (1). However, testicular externalization through a congenital scrotal defect or "scrotoschisis" remains exceptional (6,7). This is a rare malformation of the male external genitalia, the incidence of which is not well known ((2). Fewer than twenty cases have been reported in the literature since its 1ère description by Von Der Leyen (2,3,5,8,9) and these were the first cases described in our practice.

The etiopathogenesis of this condition remains poorly understood, and several hypotheses have been put forward to determine the mode of onset. For some authors, the anomaly is embryological, due to a disorder in phagocytosis of the gubernaculum testis, or to an error in scrotal mesenchymation resulting in an absence of labial fusion. (9). But for many others, meconium periorchitis seems the most likely cause. In fact, this periorchitis could be responsible for an inflammatory reaction that weakens the scrotal skin and leads to the rupture and formation of the scrotal defect, which is then responsible for the secondary externalization of the testis. (9-11). The deposit of a

greenish-yellow liquid on the externalized testicle and the meconium content found in the lateral testis corroborate this theory (3,5). This was not the case for the two patients treated at our facility, whose testicles were covered with fibrin deposits, probably due to the delay in consultation. Other causes such as obstetrical trauma, premature membrane rupture or amniotic flange disease have been reported (1,10). And in some cases, an ischemic cause or peri-natal trauma could also explain the occurrence of the anomaly (9). Structural abnormalities such as hypoplasia of the cremasteric muscle have also been suggested as a cause of the scrotal defect, leading to externalization of the testis, resulting in scrotoschisis. (7). Few authors agree on the existence of a racial predisposition in the occurrence of this malformation. (2).

In all cases, scrotoschisis is discovered in the postnatal period in the presence of externalized gonads. Antenatal ultrasound is not very sensitive in detecting scrotal defects, a clear sign of this rare genital malformation. This may be explained by testicular development and descent occurring late in pregnancy. Nevertheless, indirect signs of meconium periorchitis, the probable cause of this anomaly, can be detected during obstetrical ultrasound scans (10,12). Although inguinal or bubonochisis localizations have been noted, the scrotal location of the congenital defect remains by far the most frequent (4,9,13). In many cases, the anomaly involves a single testicle (2,3,5,6,13) but a bilateral form may also be found (2) as was also the case in one of the patients in this series. In the majority of cases, scrotoschisis remains an isolated malformation. In these two observations, no patient presented with an associated malformation, and the distant malformative work-up was normal. However, a malformative association should be sought, as some authors have reported cases of intestinal atresia, omphalocele or associated Wiedmann Beckwith syndrome (1,11,12).

Management combines antibiotic prophylaxis with a conservative surgical approach following intraoperative lesion assessment. Antibiotic therapy is still recommended to prevent possible infectious complications linked either to the delay in consultation observed, or to exposure of the testicles to the outside environment. (5) . The two patients in this series were seen with an average delay of 17 h, with no notion of initial management. Conservative surgical treatment with trimming, orchidopexy and skin suture after lavage with isotonic saline is recommended (10,13). Post-operative management is straightforward in most series, although rare cases of complications such as testicular necrosis or torsion of the spermatic cord have been reported (4,9). Ultrasound follow-up shows good progress, with homogeneous testicles of normal size. Several authors agree on the benign nature of this malformation (5,9).

Nevertheless, long-term ultrasound follow-up of these patients is recommended (3,9). This will enable us not only to confirm the benign nature of this malformation, but also to assess the functional prognosis and any sequelae that may be observed in adulthood.

Conclusion

Scrotoschisis is a rare urogenital malformation. Conservative surgical treatment gives good results. However, long-term follow-up is recommended to determine any functional sequelae.

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