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CASE REPORT

Scrotoschisis: A Rare External Genital Anomaly – A Case Report

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Abstract

Background: Scrotoschisis is a rare congenital abnormality of the external genitalia where the testis is exteriorized through an opening in the scrotal wall. Despite unknown aetiology various theories have been proposed for its occurrence. We report a case of a 3-day-old male presenting with an eviscerated left testis, exploring possible aetiologic hypotheses and management considerations. Successful surgical intervention resulted in good outcome.

Aim/objective: To outline the essential steps in the diagnosis and management of this rare congenital anomaly and to explore the possible aetiologic hypotheses in literature.

Case Report: A 3-day old male weighing 3.05 kg delivered vaginally in a rural health centre to a forty-year-old gravida 4 para 3 mother at 39 weeks' gestation was brought to the Sick Baby Unit with an abnormal protrusion of flesh from a left hemiscrotal defect, noticed at birth. Nil significant associated perinatal history. General examination and vital signs reveal essentially normal findings. There was a 2.0 x 1.5 cm defect on the anteromedial surface of the left hemiscrotum, through which the left testis, epididymis and spermatic cord protruded. The exposed testis and spermatic cord were oedematous, covered with granulation tissue but appears viable in colour with good testicular artery pulsations. The contralateral testis was normally descended within the right hemiscrotum. A diagnosis of left Scrotoschisis was made. Patient was admitted, informed consent obtained, prepared and had operative reduction/fixation of the testis with closure of the scrotal defect.

Conclusion: Scrotoschisis is a rare congenital defect of the external genitalia, occurring mostly in term male newborns. Early recognition and prompt surgical treatment are essential in managing scrotoschisis and preventing testicular injury.

Keywords: Scrotoschisis, ectopic, testes, extracorporeal, neonatal, surgery.

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INTRODUCTION

Scrotoschisis, derived from the Latin word *'scortum'* or *'scrautum'* (a bag) and the Greek word *'schisis'* (cleavage), is a rare congenital anomaly where the scrotal wall fails to enclose the testis, resulting in extracorporeal testicular ectopia.[1] It is very rare worldwide, with only three reported cases in Nigeria,[2-5] this anomaly remains poorly documented locally. We present the first documented case of scrotoschisis in Akwa Ibom State, south-south Nigeria, managed at the University of Uyo Teaching Hospital. This report aims to contribute to the literature and highlights management considerations for this rare condition.

CASE PRESENTATION

A 3-day-old male weighing 3.05 kg delivered vaginally at a rural health centre to a forty-year-old gravida 4 para 3 mother at 39 weeks' gestation, was brought to the Sick Baby Unit with an abnormal protrusion of flesh from a left hemiscrotal defect, noticed at birth. The mother had an uneventful pregnancy with routine antenatal care. There was no history of trauma to the baby's external genitalia.

On examination, the neonate was in no obvious distress, pink in room air, afebrile and not pale. There was a 2.0 x 1.5 cm defect on the anteromedial surface of the left hemiscrotum, through which the left testis, epididymis and spermatic cord protruded (Figure 1). The exposed testis and spermatic cord were oedematous, covered with granulation tissue but appears viable with good testicular artery pulsations. The edge of the scrotal defect was adherent to the base of the spermatic cord. The contralateral testis was normally descended within the right hemiscrotum. Plain chest and spine X-rays, Echocardiography, Abdominopelvic and vertebral USS were carried out to rule out associated congenital anomalies. A diagnosis of left Scrotoschisis with extracorporeal testicular ectopia was made.

He was placed on broad spectrum antibiotics (ceftazidime and metronidazole) and mother counselled for surgical intervention. Under General anaesthesia, the edges of the scrotal defect were debrided, and the testis and spermatic cord were freed. The granulation tissue was removed to expose a normal testis and spermatic cord structures. The testis was fixed to the median scrotal septum with 4-0 Polyglactin 910 suture and the scrotal defect was closed vertically to recreate the midline raphe (Figure 2). The child was discharged on the 3rd postoperative day. He was brought by the mother for a single follow up visit 2 weeks after surgery, after which he was lost to follow up.

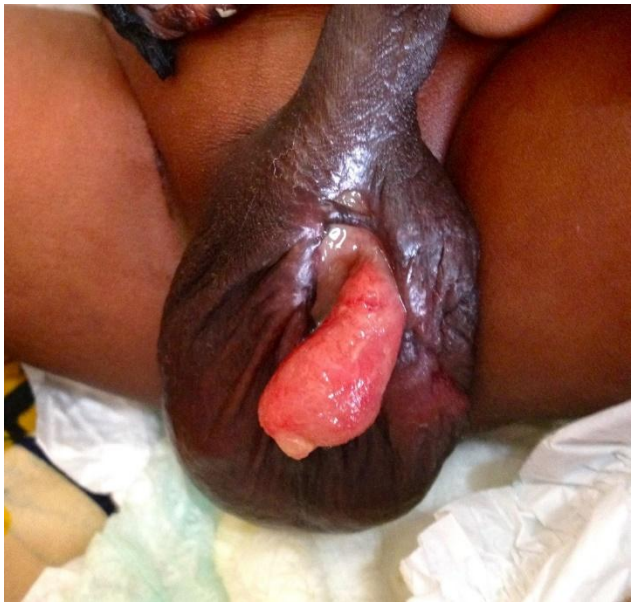

Figure 1: Image of left scrotoschisis, showing the left testis extruding through a high defect on the anteromedial surface of the left hemiscrotum.

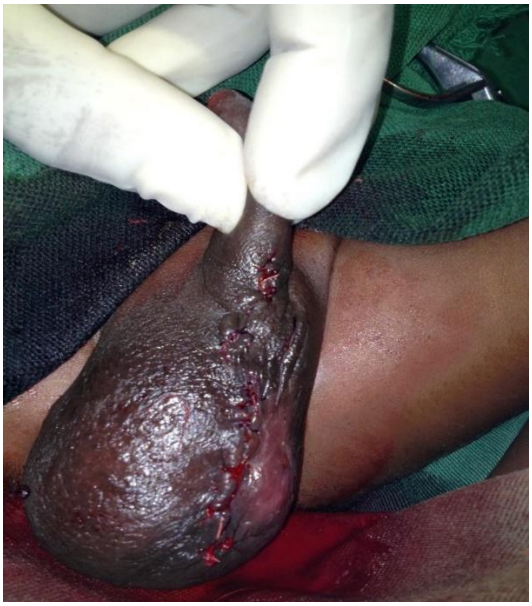

Figure 2: Image of the scrotum after orchidopexy and scrotal skin closure.

DISCUSSION

Scrotoschisis, also referred to as extracorporeal testicular ectopia, testicular exstrophy or congenital rupture of the scrotum is a rare congenital anomaly of the male external genitalia.[6] The first case was reported by Von der Leyen in 1963.[7] Currently, there is no conclusive epidemiological data available on the incidence and prevalence of scrotoschisis, with approximately 30 reported cases globally.[8] We carried out this review by examining other similar cases and individual experiences. Our reported case exhibits clinical presentations that are consistent with those documented in existing literature.

In scrotoschisis, unlike other causes of congenital testicular ectopia, the testes descend normally but protrude through a defect in the anterior wall of the scrotum.[8,9] The aetiology of this scrotal wall defect remains unknown; however, several theories have been proposed. Gongaware et al[1], postulated that a failure of the scrotal mesenchyme to differentiate into the scrotal wall may leave a thin covering of epithelium over the gubernaculum, putting the area at risk of avascular necrosis and rupture. However, studies replicating this theory in animal models have suggested that excision of a part of the scrotal wall early in foetal development would lead to failure of testicular descent. Therefore, this is unlikely to be the mechanism of aetiology of scrotoschisis, as the defect occurs after normal testicular descent.[8,10]

Also, ischaemic necrosis and rupture are unlikely, considering the rich vascularisation of the scrotal wall.[8,11]

Another theory put forward by Lais,[12] who reported scrotoschisis in a patient with arthrogryposis, suggested the possibility that external compression of the skin of the scrotal wall may have played a role in creating a skin defect. However, the most plausible and widely accepted theory holds that meconium periorchitis may occur following spontaneous or accidental perforation of the foetal gastrointestinal tract. This would allow leakage of meconium into the scrotum through the patent processus vaginalis, leading to an inflammatory disruption of the scrotal wall.[6,8,13] The absence of intestinal abnormalities in most affected newborns can be attributed to foetal healing of the intestinal lesion without sequelae.[6,14] This theory of meconium periorchitis associated evisceration is further supported by the common site of the scrotal defect, which is high and medial on the anterior scrotal wall. This site of predilection may be related to the anatomic location of the processus vaginalis, which is anteromedial to the testis.[14] This notwithstanding, only four cases have been reported in the literature occurring in association with peritonitis and/or histologic evidence of meconium periorchitis.[13,15,16,17]

Early evaluation and treatment of scrotoschisis are essential to prevent testicular injury or loss. The evaluation of a child with scrotoschisis begins with a proper perinatal history focusing on maternal illnesses, medications, and exposures to environmental teratogens. There were no significant findings in the perinatal history of this index patient. A physical examination should aim to assess the condition of both testes and identify any congenital abnormalities. While most cases occur in healthy full-term neonates, as in our index patient, exceptions exist, as seen in the case reported by Farinyaro et al[4] and Falqueto et al[18] involving preterm babies. Unilateral presentations are more common, as seen in this index patient, although a few bilateral cases have been documented.[3,8,19] The majority of reports have noted a normal extruded testis; however, Ameh et al[2] reported a rare case of torsion of the affected testis. Testicular torsion is a risk in these infants because of lack of testicular attachments. Furthermore, congenital malformations are uncommon with scrotoschisis, however, Salle et al[15] reported an infant presenting with jejunal atresia, a ruptured omphalocele, and Beckwith-Weideman syndrome. Therefore, it is important that clinicians should conduct a comprehensive examination to rule out the occurrence of other abnormalities.

To avoid desiccation, the eviscerated testis and scrotal wall defect should be wrapped in moist gauze on admission. The definitive treatment for this condition involves scrotal exploration and fixation of the testis in the scrotum. Subsequently, the wound edges are refashioned, and the scrotal wall defect is closed in layers with absorbable sutures. This closure can be achieved either transversally or vertically. In our patient's case, we opted to vertically close the scrotum to reconstruct the median raphe. While the prognosis following surgical repair is expected to be good, long-term follow-up data is currently unavailable.[9,14]

In conclusion, scrotoschisis is a rare genital defect, occurring mostly in term male newborns. Early recognition and prompt surgical treatment are essential in managing scrotoschisis and preventing testicular injury. Surgery typically involves orchidopexy and simple primary closure of the scrotal defect, with a good prognosis. This case report adds to the existing literature on scrotoschisis.

Ethical Considerations: The case report was conducted in compliance with the guidelines of the Helsinki declaration on research in human subjects and we obtained informed consent from the patient's mother for publication.

Conflict of Interest: We declare no conflict of interest.

Authorship: All authors attest to and meet the current ICMJE criteria for authorship.

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