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Scrotoschisis: Review in clinical and embryological perspective of a rare anomaly

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ABSTRACT

Scrotoschisis is a rare congenital anomaly affecting the scrotum and testes, characterized clinically by a defect in the scrotal wall and extrusion of testicular tissue. This literature review is based on reports gathered from PubMed, Science Direct, and Google Scholar using relevant keywords to explore the underlying basic science of this rare condition. The etiology and mechanism of scrotoschisis development remain largely unknown, with only theoretical explanations and a lack of empirical evidence regarding its pathogenesis, which is thought to be linked to embryological abnormalities during scrotal formation. Diagnosing scrotoschisis typically involves a straightforward visual examination of the extruded testes immediately after birth, although clinical presentations may vary, being either unilateral or bilateral. Surgical intervention, primarily through orchidopexy, is generally beneficial for repositioning the testes, closing defects, and preventing potential complications. The prognosis for individuals with this anomaly is generally favorable, although long-term testicular function has been inadequately documented. Further research, including additional clinical reports and investigations involving animal models or embryological studies, is warranted to enhance our understanding of this condition.

INTRODUCTION

Scrotoschisis, also known as extracorporeal testicular ectopia, is a congenital defect of scrotal development characterized by one or both testes being extruded outside the scrotal cavity due to a defect in the scrotal wall [1]. Its etiology is idiopathic, and its prevalence remains unknown due to the rarity of anomalies in scrotal development [2]. This condition is infrequent and has been only sporadically reported worldwide [2]. In a previous study by the author, only one case of scrotoschisis was identified in Indonesia [3]. Therefore, a comprehensive understanding of its precise mechanism is necessary to distinguish scrotoschisis from other scrotal anomalies and to provide appropriate management and reconstruction [4,5]. This narrative review article describes the literature review of this exceedingly rare anomaly.

METHODS

This literature review was conducted by collecting, reviewing, and citing relevant articles obtained from search engines such as PubMed, Science Direct, and Google Scholar. The keywords utilized were (Scrotoschisis) OR (Extracorporeal Testicular Ectopia) OR (Eviscerated Testis). Articles included in the review encompassed freely available full-texts, including case reports and reviews, accessible to any of the authors. Additionally, studies elucidating the basic science of scrotal anomalies, including scrotoschisis, were incorporated. A total of 32 published manuscripts were obtained and reviewed. These reports were assessed based on clinical presentations of scrotoschisis, treatment modalities employed, complications encountered, and patient outcomes, as outlined in the respective reports.

OVERVIEW

Scrotoschisis, also known as extracorporeal testicular ectopia, eviscerated testicle, testicular exstrophy, congenital rupture of the scrotum, and bubonoschisis, is a congenital malformation of the scrotal wall wherein the testes protrude outside the scrotal cavity due to a defect in the scrotal wall [1]. The first documented case was reported in 1963 by Von der Leyen, with approximately 30 cases reported globally [2,6]. The etiology and prevalence of scrotoschisis remain elusive due to the rarity of anomalies in scrotal development [4]. Furthermore,

the exact birth prevalence of scrotoschisis remains undisclosed. The common site of the defect is situated on the high anterior wall, proximal to the median raphe [5]. Typically unilateral, scrotoschisis primarily affects male newborns. The prognosis for this condition is generally favorable, although long-term outcomes are not readily available [7].

Various theories and hypotheses regarding the pathophysiology of scrotoschisis have emerged in the past decade, including trauma, abnormal gubernaculum, aberrant amnion bands, meconium periorchitis, and abnormal fusion of labioscrotal folds [8]. In our literature search, the most recent cases were reported in 2022 by Nguinzanemou et al. in Central Africa, constituting the 18th reported case, followed by a study reported by Eshibe et al., which was the 19th [2,9]. We believe that our previous study on scrotoschisis from Indonesia represents the 20th case reported globally, as a similar case had not been previously documented [3].

EMBRYOLOGY OF SCROTAL DEVELOPMENT

The testis originates embryologically from primordial germ cells, which are generated in the epiblast and migrate to the endoderm cells in the genital ridges during the early 5th week of development. This process is initiated by the activation of the XY sex chromosome, particularly the SRY gene encoding the testis-determining factor. Subsequently, sex cords develop into testes, which are then covered with a layer of tunica albuginea. This differentiation process also involves the formation of specialized cells such as Sertoli and Leydig cells [10].

By the 9th week of gestation, Sertoli cells begin to produce Anti-Mullerian Hormone (AMH), which inhibits the development of the Mullerian duct, the precursor of the female genital tract. Concurrently, Leydig cells produce testosterone, which induces the differentiation of Wolffian ducts into the male accessory reproductive tract and masculinizes external genital organs in response to testosterone. These processes facilitate the further development of male genital organs, primarily the descent of the testes from the abdomen [10,11].

The migration of the testis from the abdomen occurs between the 8th and 15th gestational weeks and is divided into two phases: transabdominal and inguinal-scrotal migration. Initially, the gubernaculum enlarges at the caudal end under the influence of Insulin-like Hormone 3 (INSL3) produced by Leydig cells. The testis then migrates toward the gubernaculum and descends into the abdomen through the processus vaginalis [11,12].

Upon reaching the processus vaginalis, inguinoscrotal migration occurs. During this phase, the gubernaculum further develops and migrates toward the scrotum. Testosterone influences this process by elongating the processus vaginalis through the inguinal canal, pulling the testis downward into the scrotal cavity where the gubernaculum is located. This migration process may vary in timing, typically occurring between the 25th and 35th weeks of gestation [11,12].

Simultaneously, external genitalia development commences around the 6th week of gestation. In the presence of dihydrotestosterone, the phallus develops into the penis, and the labioscrotal (urogenital) folds fuse to create the closed structure of the scrotum. In females, the absence of dihydrotestosterone prevents the fusion of the labioscrotal folds, resulting in the formation of the labia majora. In males, the fusion of both urethral folds and labioscrotal folds in the midline forms a scrotal raphe, burying the penile shaft deep below the scrotal cavity and resulting in the formation of the scrotal sac, which is comprised of excess folds of skin [13,14].

ETIOPATHOLOGENESIS OF SCROTOSCHISIS

Scrotoschisis arises when there is a failure in the closure of the labioscrotal folds, resulting in a defect in any part of the scrotum, which poses a risk of evisceration of its contents. This mechanism stands as the most widely accepted explanation for scrotoschisis, as its etiology remains unknown. Various theories have been proposed, all converging on the common theme of a defect or failure in the fusion of the labioscrotal folds during scrotal formation [13,15].

One prevalent theory suggests the absence of the cremaster muscle, weakness in the development of scrotal mesenchyme, chemical-induced inflammation of the scrotum, meconium periorchitis, external compression, and iatrogenic injury as potential factors contributing to this anomaly. However, none of these theories have been definitively proven as the underlying mechanism for this rare condition [15].

Many reported cases have associated this anomaly with meconium periorchitis, wherein patients exhibited testicular tissue with debris and thick green fluid during surgical exploration, consistent with meconium. Inflammatory signs were often observed in the defect and the exteriorized testicular tissues. Meconium, typically associated with defects in the gastrointestinal tract, may leak into the peritoneal and scrotal cavity through a patent processus vaginalis. The presence of meconium in the scrotal sac triggers an inflammatory reaction, leading to a scrotal wall defect. Histopathological examination confirming periorchitis with meconium residuals has supported this theory [14,16].

However, some cases have shown no evidence of meconium residuals, prompting consideration of

other potential mechanisms such as mechanical compression and iatrogenic injury. Increased intraabdominal pressure may force the descent of the testis over the developing scrotal sac. Iatrogenic injury, particularly during vaginal delivery, could also occur due to surgical or mechanical compression during labor. Nonetheless, evidence of lacerating trauma associated with iatrogenic injury has not been reported [17,18].

Another proposed mechanism by Gongaware et al. suggests a failure in the differentiation of scrotal mesenchyme. This theory suggests that scrotal closure occurs but with only a thin epithelial layer and a lack of anchoring mesenchymal structures. Consequently, the structure becomes prone to rupture under compression or avascular necrosis, resulting in a defect through which the testes eviscerate [14,19].

TYPES OF SCROTOSCHISIS

Scrotoschisis can manifest at various sites depending on the type, which is determined by the number of testes involved in the extrusion and the location of extrusion, including scrotal and inguinal "bubonoschisis" [4]. The majority of scrotoschisis cases are unilateral and affect typically developing males. However, bilateral cases have also been documented, as well as unilateral cases involving both testes [7,8,20]. There are even instances where scrotoschisis occurs in the midline, involving both hemiscroti and resulting in the extrusion of both testes [21].

Scrotoschisis can be further categorized based on the site of extrusion into scrotal and inguinal "bubonoschisis" [4]. Bubonoschisis refers to the condition where the testicle protrudes through the inguinal canal. In most cases, the malformation of the scrotum is located on the proximal anteromedial scrotal wall, with the testes extruding through a defect in the anterior and cranial wall of the ipsilateral scrotum adjacent to the median raphe and the penoscrotal junction. Additionally, there have been cases where the malformation is situated at the scrotal base [3,22]. For example, Haidar and Gharmool reported a case of inguinal "bubonoschisis" in a male infant in Libya, where left testicular extrusion was observed through a small defect in the inguinal canal at the level of the superficial inguinal ring [1].

DIAGNOSTIC WORKUP

Scrotoschisis should be considered in the differential diagnosis when a newborn presents with abnormal testicular shape, as it is most commonly observed in full-term newborns of normal weight and age [2,22]. Gross identification of scrotoschisis can be achieved by its macroscopic appearance. Upon localized examination, a lesion may appear as an amalgamated mass extruding through a defect in the scrotal wall or inguinal canal at the level of the superficial inguinal ring. A fibrotic layer covering, similar to gastroschisis, may also be present [3]. The cord is typically of normal length with an intact processus vaginalis, and almost no hernial sac or penile anomalies have been reported in any case to date. The size of the extrusion site is usually smaller than the diameter of the extruded testicles, indicating that the disorder likely occurred early in pregnancy. Occasionally, the scrotum may appear empty but well-formed and similar in size to the contralateral side [4]. See Figure 1 for an illustration of a scrotoschisis case [18].

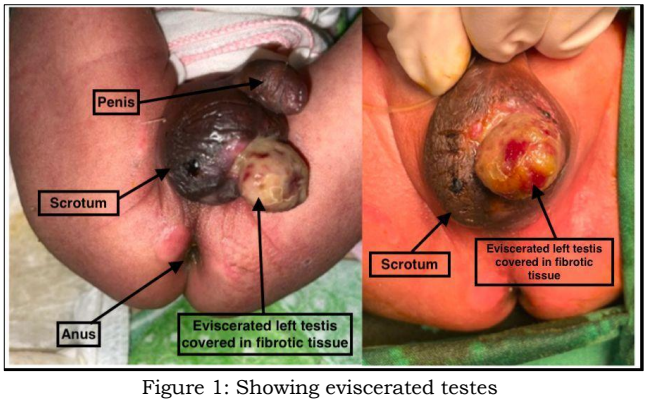

Figure 1: Showing eviscerated testes

During general examination, slight impairment of vital signs may be noted, with temperature being the most commonly affected, occasionally rising to a febrile state [6]. In such cases, laboratory studies, including a complete blood count, may indicate presumptive sepsis, characterized by pancytopenia [2].

Further evaluation with multimodal diagnostic tools is warranted if other accompanying congenital anomalies are suspected. Anomalies associated with scrotoschisis include jejunal atresia, intestinal atresia, Beckwith-Wiedemann syndrome, ruptured omphalocele, meconium periorchitis, and testicular torsion [4,23]. Plain abdominal radiography and whole abdomen ultrasound are essential diagnostic tools and can also aid in detecting bowel perforation and peritonitis [4].

MANAGEMENT

Most previous studies have recommended surgery as the definitive treatment for scrotoschisis, typically involving dartos pouch orchidopexy and closure of the defect using absorbable sutures [8]. The objective is to return the testis to the scrotal sac. Treatment typically involves testicular cleaning, scrotal exploration, orchidopexy, and defect closure [24, 25]. Upon diagnosis, administering broad-spectrum antibiotics is recommended. In cases awaiting surgery, wet aseptic gauze or warm compresses

should be applied to the testis to prevent dessication and dehydration of the organ [6,21]. The surgical approach varies depending on the severity of the presentation, ranging from simple dressing with secondary healing to defect repair in layers under general anesthesia (GA) or local anesthesia (LA) [4]. While treatment of scrotoschisis is not usually considered an emergency unless complicated by torsion, early repair may help prevent advanced complications [2,26]. See Table 1 for a comparison of treatment methods and outcomes from various antecedent studies, including complications accompanying the anomaly presentation or occurring during surgery.

Table 1: Treatment of scrotoschisis in prior studies

Study	Scrotoschisis type	Treatment	Complication	Outcome
Eshiba et al. (2022)[9]	Unilateral	Broad spectrum antibiotics, orchidopexy, and defect closure under GA	-	Successful
Sibomana et al. (2019)[8]	Unilateral with both testes extruded	Wet gauze cover, scrotal exploration, and orchidopexy under GA	-	Successful
Keita et al. (2021)[6]	Unilateral	Broad spectrum antibiotics, orchidopexy, and defect closure under GA	Fever when admitted	Successful
Nguinzanemou et al. (2022)[2]	Unilateral	Broad spectrum antibiotics, orchidopexy, defect closure under GA	Sepsis presumption when admitted	Died 3 hours after surgery, due to GA side effects
Premkumar et al. (2009)[27]	Unilateral	Moist saline gauze, orchidopexy, and defect closure under LA	-	Successful
Syed Waqas et al. (2021)[21]	Bilateral	Scrotal exploration, orchidopexy, and defect closure under GA	-	Successful
Togami et al. (2002)[28]	Unilateral	Scrotal repositioning and defect closure under LA	-	Successful
Sidibe et al. (2017)[5]	Unilateral	Broad spectrum antibiotics, orchidopexy, and defect closure under GA	-	Successful
Abubakar et al. (2015)[24]	Bilateral	Parenteral antibiotics, antiseptic gauze, orchidopexy, and defect closure	Fever when admitted	Successful
DeRoo et al. (2016)[20]	Unilateral	Orchidopexy and defect closure	Meconium periorchitis, admitted to the operating room two times	Successful
Eleweke et al. (2018)[29]	Unilateral	Saline-soaked gauze and orchidopexy under GA	-	Successful
Jesus et al. (2012)[22]	Unilateral	Moist warm sponges, broad-spectrum antibiotics, and orchidopexy under GA	-	Successful
Mahfouz (2018)[30]	Unilateral	Broad spectrum antibiotics, orchidopexy, and defect closure under GA	Ventricular septal defect	Successful
Onuora et al. (2013)[31]	Unilateral with both testes extruded	Broad-spectrum antibiotics with tetanus toxoid, orchidopexy, and defect closure	-	Successful
Perveen et al. (2021)[32]	Bilateral	Prophylactic antibiotic, testis replacement, and defect closure under GA	-	Successful

CONCLUSION

Scrotoschisis is a rare congenital anomaly affecting the scrotum and testis. The limited reports and evidence available pose challenges in identifying its cause and mechanism. However, most cases can be treated surgically, with generally good prognoses proposed. Nevertheless, further studies focusing on cohorts of testicular development will be necessary to assess the long-term prognosis of testicular function. Additionally, conducting animal model research to investigate the various theories regarding the mechanism of scrotoschisis may prove valuable in both understanding and preventing further cases of this anomaly.

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