



Ectopic scrotum: Single stage rotational flap reconstruction with orchidopexy

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ABSTRACT

Introduction: Congenital anomalies of the scrotum are rare; furthermore, ectopic scrotum and accessory scrotum are rarer still. We describe our series of four cases with their surgical management and long-term outcomes. We performed single stage rotational flap reconstruction with orchidopexy.

Materials and methods: Data was collected retrospectively for three cases of ectopic scrotum and one of accessory scrotum over the last twenty years in a tertiary paediatric surgery unit.

Results: Two infants with supra-inguinal ectopic scrotum underwent rotation pedicle flap procedure to relocate the scrotum and testis. The follow-up at 5 years shows excellent cosmetic outcome. The third infant had anorectal malformation and ectopic scrotum with absent testis. The fourth child had a large perineal hamartoma and an overlying accessory scrotum. In the later two, the empty scrotums were excised.

Conclusion: Single stage rotational flap reconstruction with orchidopexy, is feasible and should be considered. It is a safe single stage technique to achieve excellent cosmesis and hopefully function of the ectopic gonad. Collaboration between paediatric surgeons and plastic surgeons in these rare cases is advisable. The published reports show cases operated in stages and at times sacrificing the ectopic scrotal skin. We elected to operate early without sacrificing scrotal tissue and preserving the testis.

1. Introduction

Congenital scrotal disorders are rare. Approximately 28 cases of ectopic scrotum have been reported [1–6]. Scrotal anomalies include scrotal hypoplasia, penoscrotal transposition, bifid scrotum, ectopic scrotum and accessory scrotum. The last two entities are extremely rare, whereas the first three are relatively common.

We report a series of 4 cases seen in the last twenty years at a tertiary Paediatric Surgery centre. We describe single stage rotational flap reconstruction with orchidopexy. The rarity of the primary condition, i.e. ectopia, along with associated anomalies make this series unique and worth reporting. A review of literature to study the etiology, still quite enigmatic, was undertaken.

2. Materials & methods

The study population comprised of three patients of ectopic scrotum and one of accessory scrotum between January 1998 - December 2009. Detailed history and physical findings in each of the cases are outlined below:

2.1. Case 1

A newborn male had severe reduction deformity of left lower limb. There was partially developed left hemipelvis with absent hip joint and a rudimentary femur. He had a normal right scrotal sac with a median raphe and a very rudimentary left scrotum adjacent to this. In addition, he had an ectopic but well developed scrotum in the left supra-inguinal region containing the left testis. The renal tract was normal (Fig. 1).

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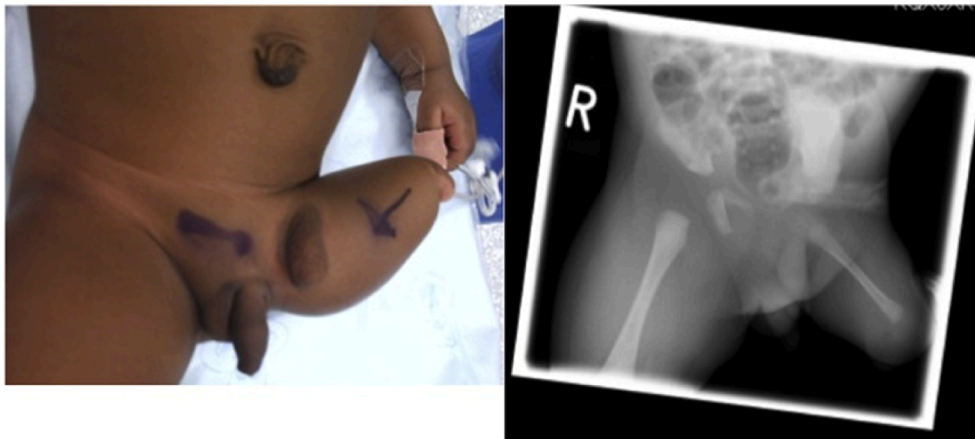


Fig. 1. Case 1 with inguinal left ectopic scrotum and X-Ray.

2.2. Case 2

A neonate presented with an ectopic supra-inguinal left hemi scrotum without any associated anomaly (Fig. 2).

2.3. Case 3

A male neonate presented with anorectal malformation, left supra-inguinal ectopic scrotum, ipsilateral renal agenesis and absent left testis (Fig. 3). There were no other abnormalities. The right scrotum had a hypertrophied testis.

2.4. Case 4

A 3-year-old male presented with a perineal hamartoma and an overlying accessory ectopic scrotum. He otherwise had a normal scrotum and normally located testes.

2.5. Surgical technique of relocation

The well developed ectopic scrotum was relocated where scrotal tissue in the normal location on the ipsilateral side was hypoplastic. The cases were managed jointly with the paediatric and plastic surgical teams. Case 1 and 2 were operated at 7 months and 9 months of age respectively. The ectopic scrotal skin was relocated adjacent to the median raphe as a perforator based island flap based on the per-

forators of the external pudendal artery. Simultaneously, the ectopic left testis and cord were mobilised out of the ectopic scrotum. This allowed sufficient length to place the testis in the scrotum in its new position (Fig. 4). Both patients had an uneventful post-operative course with follow up demonstrating a good cosmetic result and normal testicular growth at 5 year follow up (Figs. 5 and 2).

Case 3, underwent surgery for the anorectal anomaly and the small, empty, ectopic scrotum was excised. The other testis and hemiscrotum was noted to be larger than normal. In Case 4, the accessory ectopic scrotum was excised and histology confirmed it to be normal scrotal skin with dartos. The hamartoma being static in size and asymptomatic has been treated conservatively.

3. Results

Surgical and cosmetic outcome at 5 years in the operated cases has been excellent. The functional outcome is difficult to assess at this stage.

4. Discussion

We have described our single stage technique of scrotal transposition without sacrificing scrotal tissue, while preserving and relocating the testis. The published reports show cases operated in stages [5] and at times sacrificing the ectopic scrotal skin [3]. We have elected to operate early, as opposed to several reports of operation after one year



Fig. 2. Case 2 with ectopic left scrotum and at 1 year follow up.



Fig. 3. Case 3 with ectopic left scrotum and absent left testis.

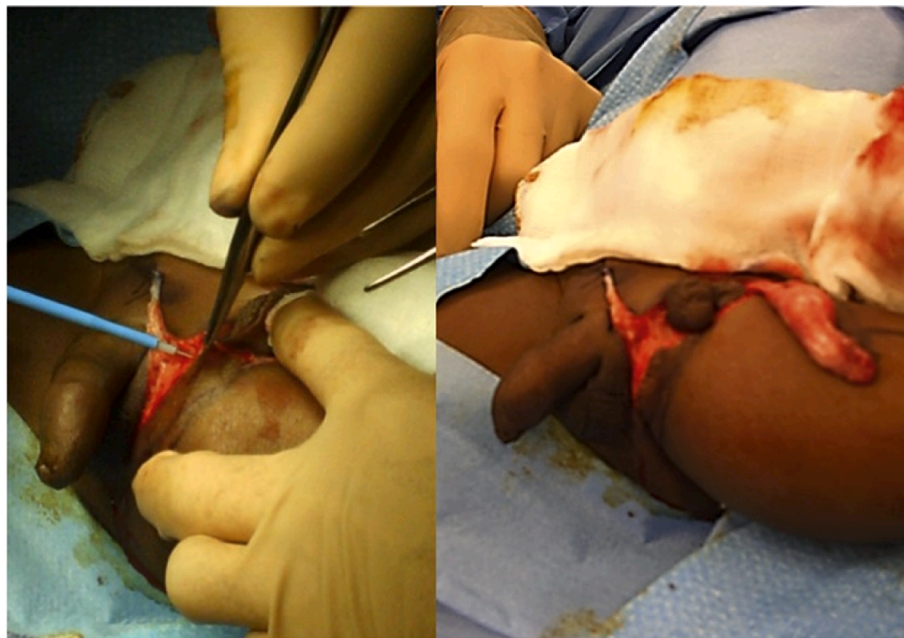


Fig. 4. Case 1 intra operatively.

of life. We do not believe that cases of ectopic scrotum need delayed surgery. Our justification being the testis is technically fully descended, albeit in an ectopic position. The long term follow up shows excellent cosmetic outcome.

Through these four cases, we have reported varying presentations of ectopic scrotum. As such ectopic/accessory scrotum is a rare condition, the commoner scrotal anomalies being hypoplasia, bifid scrotum and penoscrotal transposition often associated with hypospadias. Other anomalies with ectopic scrotum are chordee, undescended testes, renal anomalies [7], patent urachus [8], VATER association [3,9], anorectal malformation [10], perineal hamartomas and lipomas [11]. Interestingly, patients have also been noted to develop inguinal herniae within the ectopic scrotum itself [7]. Knowing that labia majora has the same embryogenesis as scrotum, we searched the literature and found only one reported case of an ectopic labia associated with VATER anomaly [12].

Depending on the location, ectopia can be classified as (a) perineal, (b) infrainguinal, (c) suprainguinal and (d) femoral. In our series, three cases belonged to the category of suprainguinal ectopic testis and one had perineal ectopic testis.

The etiology for an accessory and ectopic scrotum is not exactly known. Scrotal formation begins at approximately the fourth week of gestation. The scrotum forms from the labioscrotal swellings that are adjacent to the urogenital fold, which forms the penis. The labioscrotal swellings migrate postero caudally fusing in the midline to form the scrotum at approximately 10–12 weeks of gestation.

Several hypotheses for the development of an ectopic scrotum have been proposed. Embryological, mechanical, genetic, chromosomal and teratogenic factors have been implicated. Embryological theory suggests abnormal division of labioscrotal fold allowing the scrotum to get dislodged. The second possibility is abnormal caudal migration of labioscrotal swelling. In cases of perineal hamartoma, the



Fig. 5. Case 1 post operatively and at 1 year follow up.

etiology is thought to be due to presence of intervening mesenchymal tissue or a teratoid growth impeding normal labio scrotal swelling [13].

However, in our first case, none of the aetiologies appear to explain the genesis of ectopia. This makes us postulate a mechanical factor in scrotal migration. It could be that the limb buds normally keep the labioscrotal fold located medially, adjacent to the midline. However, in the absence of a limb bud there was no force to keep the labio scrotal fold in the centre and therefore mechanical shearing force of the opposite side allowed lateral migration of labio scrotal fold, which forced the scrotum to be positioned laterally. All this may have resulted from abnormal division not allowing the formation of the limb bud. Stephen [14] proposed that pressure from the contralateral heel causing compression during intrauterine life, could be a cause for abnormal scrotum formation. This also explains perineal abnormalities such as ano-rectal malformation. In addition to pregnancy related complications such as oligohydromnios and breech presentation, the abnormally flexed limb could also be responsible for the reduction deformity of the contralateral limb.

In cases where there are associated anomalies like in our Case 3, the etiology could well be a VATER association, genetic or teratogenic factors.

It may be pertinent to discuss if the gubernaculum [15] has a role in inguinoscrotal testicular descent. Several authors have claimed that the cremasteric sac forms by eversion of the gubernaculum cone whereby regression of the extra abdominal part of the gubernaculum creates a space into which the gubernaculum cone everts to form the processus vaginalis within the scrotum. Some authors have described the cause of testicular ectopia as a mystery and over years many hypotheses are suggested [16]. It is proposed that gubernacular eversion is more apparent than real and that there is some degree of gubernacular migration.

Hoar [17] proposed that the gubernaculum could be a prerequisite for the ultimate location of both the testis and the scrotum, its role is complicated by differential growth of labioscrotal folds in which the gubernaculum is stabilized. If the interaction is disturbed, the result could be suprainguinal ectopia, penoscrotal transposition or perineal scrotum. A femoral scrotum on the other hand is the result of aberrant gubernacular stabilization. Hoar also acknowledged that the etiology of these abnormalities is likely to be multifactorial.

In 1988 Ikadai et al. established a new rat line (TS inbred rats) showing uni- or bilateral ectopic scrota in about 70% of the males. Genetic analysis of the TS inbred rats suggested that the formation of ectopic scrotum was controlled by multiple genes [18].

There have been only 2 previously reported clinical cases, of ectopic/accessory scrotum with associated lower limb skeletal deformities, with some similarity to our first case [19,20]. Zoran [19] presented a case of left tibial aplasia, associated with hypoplastic distal femoral epiphysis, shortened/thickened fibula, and an accessory scrotum. On further elaboration they describe a bifid scrotum with cryptorchid testis. They postulate that a combination of such skeletal and genital abnormalities could be accounted for by an insult occurring during the late blastogenesis stage of embryological development, i.e. close to 28 days of gestation, causing an effect on the development of both systems. Kendirci [20], describe a case of accessory scrotum, proximal femoral focal deficiency and diastasis of the symphysis pubis. They suggest that such defects, involving two organ systems, could be accounted for by an embryologic mesenchymal disorder, as this forms the initial precursor for connective tissue, bone, cartilage, the circulatory and lymphatic system, as well as the external genitalia.

Accessory scrota, such as that described in our last case, are often associated with perineal lipomas. Sule et al. [11] reported that up to 83% of accessory scrotums are associated with perineal lipomas, and hypothesised that the accessory labioscrotal fold, and hence accessory scrotum, develops because the lipoma, otherwise described as intervening mesenchymal tissue, disrupts the continuity of the caudally developing labioscrotal swelling. Park et al. postulated that the position of the lipoma itself may be directly related to the type of scrotal abnormality that the child develops [21]. It has been noted that most accessory scrotums associated with a perineal lipoma are not associated with other abnormalities [11,22]. Interestingly, one case of accessory scrotum has been diagnosed at the 32 week antenatal ultrasound scan [23].

In the absence of a perineal lipoma, accessory perineal scrotums are usually associated with other anomalies [9,22]. Accessory scrota have been reported with anorectal malformations such as imperforate anus [24,25], glandular hypospadias [26], VATER association [9], penoscrotal transposition and pseudodiphallia [27], retrocerebellar arachnoid cysts [28] and partial 'prune-belly' syndrome [29]. One report of an associated Becker's Nevus, previously shown to contain androgen receptors in levels similar to genital skin, suggests that both abnormalities may have developed due to androgen sensitivity and stimulation [30]. We noted only 2 other reported cases of accessory scrotum being associated with a perineal hamartoma as was present in our case.

5. Conclusion

It is concluded that a single etiological factor may not be responsible for the genesis of ectopia. Defects in embryogenesis (abnormal migration, interruption of developing labioscrotal folds), mechanical factors, genetic, teratogenic, gubernacular, syndromic association, are some postulated hypotheses.

Single stage rotational flap reconstruction with orchidopexy, is feasible and should be considered where the orthotopic scrotum on that side is hypoplastic. It is a safe single stage technique to achieve excellent cosmesis and hopefully function of the ectopic gonad. Collaboration between paediatric surgeons and plastic surgeons in these rare cases is advisable.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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