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Case Report

Congenital bilateral scrotal agenesis with bilateral cryptorchidism: a case report

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ABSTRACT

Congenital bilateral scrotal agenesis is very rare anomaly and characterized by the absence of scrotal rugae. It may be associated with other extra-genital anomalies as a part of a syndrome. Etiology is unknown but may be due to primary failure of labioscrotal folds development or secondary to localized 5-alpha-reductase type 2 deficiency. Various surgical techniques used for neoscrotum construction with different types of complications. In index case, we performed bilateral orchidopexy without creating neoscrotum.

Keywords: Cryptorchidism, Dihydrotestosterone, Scrotal agenesis, Testosterone

INTRODUCTION

Congenital bilateral scrotal agenesis is very rare anomaly of scrotal development. It is characterized by the absence of scrotal rugae in the perineum between the phallus and anus.[1,2] Till date approximately 6 cases were reported in literature.[3] Sometime, this anomaly can be associated with extra-genital anomalies as a part of a syndrome.[4] Bilateral scrotal agenesis may be due to primary failure of development of labioscrotal folds or secondary due to localized deficiency of 5-alpha-reductase type 2.[5] We are going to present a case of bilateral scrotal agenesis with bilateral non palpable undescended testes in 11 years old boy.

CASE REPORT

An 11-year-old male child presented to our outpatient department with history of empty bilateral hemi scrotum since birth. He was the first order child, born to non-consanguineous parents by normal vaginal delivery. There was no history of sibling death. On physical examination, normal size buried phallus of length 5 cm with normal urethral meatus. The patient had prominent mons-pubis and flat perineum without scrotal sac and rugosity (Figure 1 A). The midline raphe extended from the base of the phallus to the anterior anal verge. Bilateral testes were not palpable in the inguinal canal. Clinically patient had facial dysmorphism like bilateral maxillary bone hypoplasia and thin upper lip vermilion border. He had no others skeletal anomalies. Other systemic examinations including ophthalmic evaluation and echocardiography are within normal limits.

Abdominal ultrasonography (USG) did not exhibit any associated urinary system anomaly and mullerian structures remnant. Inguino-srotal USG suggestive of right testis of size 1.3×0.8×1.6 cm in right inguinal region and left testis of size approx. 1.8×0.9×0.6 cm in left inguinal region. The karyotype was 46, XY. Patient hormonal profile like serum testosterone, serum dihydrotestosterone, luteinizing hormone levels and testosterone/dihydrotestosterone ratio were normal. After proper clinical evaluation and pre-anesthetic clearance, we were planned for examination under anesthesia and diagnostic laparoscopy in view of non-palpable

undescended testes. Bilateral testes were not palpable under anesthesia. Laparoscopy findings suggestive of closed bilateral deep inguinal ring with vas and vessels entering through deep inguinal ring. There was no mullerian structures remnant visualized during laparoscopy. On inguinal exploration, both testes were found in bilateral inguinal canal respectively (Figure 1 B). After proper mobilization of cord structure both testes placed in subcutaneous tissue on either side of median raphe at proposed scrotal site (Figure 1 C). Post operative period was uneventful. We were kept patient on regular follow-up. At 3 months follow-up both testes palpated clinically and planned for Doppler sonography to look for testicular size and vascularity.

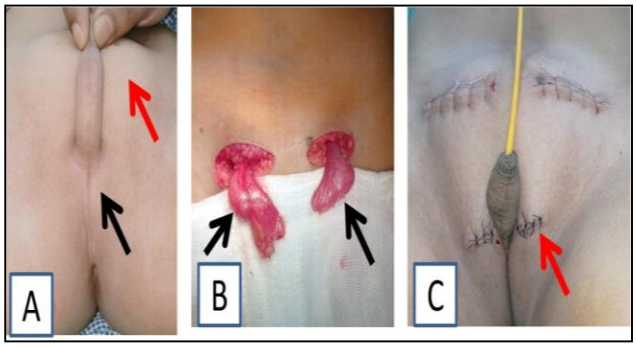

Figure 1 (A-C): Pre-op of absence of scrotal rugae, normal size phallus (black arrow) and prominent mons pubis (red arrow) and intra-op of bilateral testes (black arrow) and post-op picture of orchidopexy site (red arrow).

DISCUSSION

Scrotal agenesis is an extremely rare scrotal developmental anomaly.[2] The cause of scrotal agenesis is still unknown. Abnormal development of labio-scrotal folds is proposed to be the cause of scrotal agenesis.[2] Scrotal development starts by formation of labioscrotal folds on either side of urogenital folds at 7th week of gestation. Enzyme 5-alpha reductase converts testosterone to dihydrotestosterone (DHT). Dihydrotestosterone is more potent than testosterone which induces virilization of external genitalia.[1] At 16th week of gestation, labioscrotal folds fuse in midline to form scrotum.[6] However, very high level of dihydrotestosterone prevents labia formation without stimulation of scrotal development, as evidenced by presence of midline raphe.[4-6] Scrotal agenesis may present as an isolated anomaly or may be associated with other congenital defects such as face anomalies, growth retardation, cardiac anomalies, anterior ectopic anus, undescended testes and digit anomalies.[5,7,8] There are various surgical techniques for neoscrotum construction with different types of complications. Myocutaneous flaps may be used for neoscrotum creation but cosmetic point of view may be sub-optimal because of the obvious muscle deformity and unnatural look of the scrotum.[9] Preputial skin may be used to create neoscrotum but there is a risk of necrosis or infection of the preputial flap could compromise the safety of the repositioned testes.[8,10] Our patient had other anomalies like facial dysmorphism and bilateral cryptorchidism. In index case, we performed bilateral orchidopexy without creating neoscrotum. This procedure is easy and procedure related complications are almost nil.

CONCLUSION

Concluded that, scrotal agenesis is not an isolated anomaly. It may be associated with other anomalies. So, proper preoperative evaluation is essential for better surgical outcome.

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