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Case Report

Duplicate Bladder Exstrophy with Superior Vesical Fistula in a Male Infant, A Rare Variant

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Abstract

We report a rare variant of exstrophy-epispadias complex, a duplicate bladder communicating with exstrophy bladder via a fistula. On presentation appeared to be a case of classical bladder exstrophy with diastasis of the pubis and rectus muscles, a low lying umbilicus and a form of epispadias. On careful examination phallus appeared normal with normal external urethral meatus.The micturating cysto-urethrogram was done which elicited a rare anomaly; duplicate bladder exstrophy.

Introduction

Classic bladder exstrophy is an extremely rare congenital anomaly with an incidence of 1:50,000 live births [1]. Variants of the exstrophy-epispadias complex are even rarer, making up to 8% of these midline defects [2]. Duplicate exstrophy is a rare variant in which patient presents with a patch of exstrophied bladder, a normal appearing abdominal bladder, diastasis of the symphysis pubis and rectus muscles and a form of epispadias. We report a rare variant of duplicate exstrophy with a fistula between the exstrophy bladder and normal abdominal bladder in a male infant.Tomita et al have suggested this as a hybrid form of duplicate bladder exstrophy and superior Vesical fistula [3].

Case Report

A one month old male neonate presented with leaking urine from an abnormal mucosal patch over his lower abdomen. The baby was passing urine normally per urethra. Clinically the baby appeared healthy with a mucosal patch on lower abdomen, below a low lying umbilicus, diastasis of the pubis and rectus muscles and urine leak from mucosal patch. Infant had a well formed penis with normally placed external urethral meatus with shiny dorsal penile skin and dorsal chordee. Prenatal ultrasound was reported normal. Post natal ultrasonography, demonstrated normal kidneys, ureters and bladder, beneath exstrophied bladder patch. Child was catheterized and surprisingly, catheter extruded from midline through mucosal patch (Figure 1). The catheter was withdrawn and micturating cysto-urethrogram was done which showed a normally situated bladder with bilateral grade 1 vesico-ureteric reflux and urine leak in exstrophied bladder via Vesical fistula; (Figure 2) and normal urethra(Figure 3). The plain abdominal radiograph film showed a widened symphysis pubis. Intra-venous urography also showed normal kidneys, ureters and bladder and urine leak in exstrophied bladder.

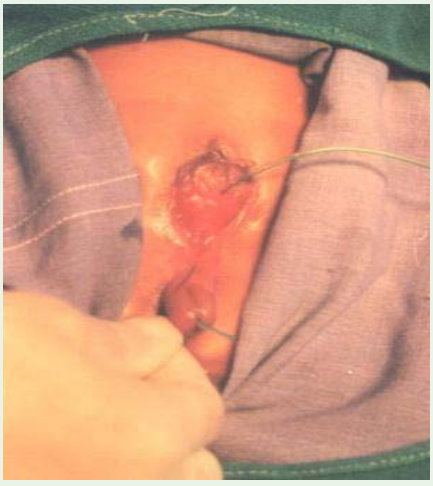

Figure 1: Catheter seen extruding from midline through mucosal patch.

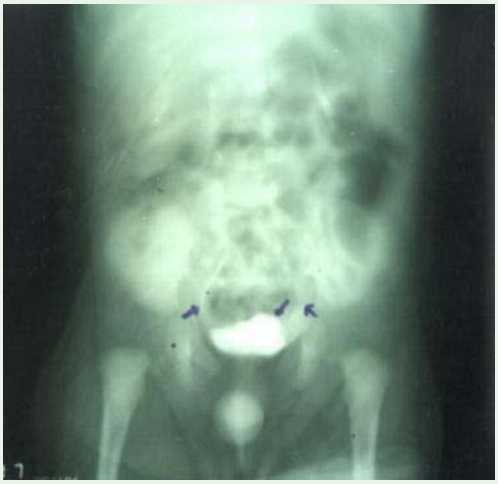

Figure 2: Micturating cysto-urethrogram showing a normally situated bladder with bilateral grade 1vesico-ureteric reflux and urine leak in exstrophied bladder via vesical fistula.

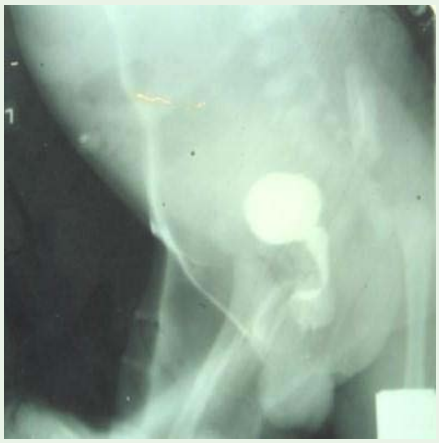

Figure 3: Micturating cysto-urethrogram showing a normally situated bladder and normal Urethra.

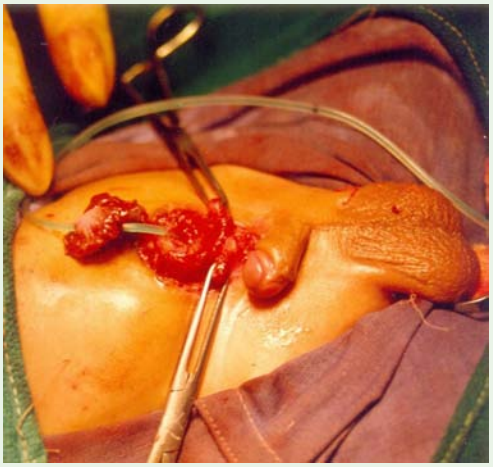

Figure 4: Excision of exstrophy bladder mucosa with fistula tract.

The clinical examination and micturating cysto-urethrogram confirmed the diagnosis of a rare exstrophy variant, duplicate bladder exstrophy with superior Vesical fistula. Child was taken for cysto-urethroscopy, which demonstrated a normal urethra and bladder with normally placed ureteric orifices. The bladder dome demonstrated a fistula, with urine leak from exstrophied bladder. During same anesthesia exploration was done, a small single midline orifice from which urine leaked was identified and cannulated with an infant feeding tube. The exstrophy bladder mucosa with fistula tract was excised (Figure 4), and cystostomy tube was placed through the fistula site. As the separation of recti was not very wide, the abdominal closure was possible without tension and an osteotomy was not required. The mucosal extension along the dorsum of penis which appeared to be epispadias urethra of exstrophied bladder was left in situ for chordee correction in future. The histopathology of the exstrophied patch showed urothelial lined fibro muscular tissue. Postoperatively, child passes urine normally with a good stream. The ultrasonography at 6 months of age showed normal bladder with normal upper urinary tracts.

Discussion

Marshall and Muecke have discussed the embryological basis of exstrophy –epispadias complex. They have suggested that an abnormal cloacal membrane prevents the lateral mesoderm from migrating and closing towards the midline and disrupts the development to the lower abdominal wall [4]. Exstrophy variants can be categorized as duplicate exstrophy, superior Vesical fistula; pseudo exstrophy and covered exstrophy [1]. There are two different forms of duplication, antero-posterior duplication and side by side duplication. The first form is considered a duplicate bladder exstrophy with a patch of exstrophy bladder mucosa on the anterior abdominal wall with a second bladder lying in the pelvis with both ureters opening in the normal bladder [4,5]. Superior Vesical fistula, sometimes mistaken for superior Vesical fissure, consists in a small communication between the normal bladder and exstrophied bladder and is the most common variant form [6].

According to Tomita, et al. this exstrophy variant could be a hybrid of duplicate bladder exstrophy and superior Vesical fistula. Our case is similar to one reported by Tomita, et al. [3], where it was operated as classical bladder exstrophy and neonate underwent bilateral posterior iliac osteotomies; a major intervention for a neonate.Bouali et al have reported duplicate exstrophy in a female newborn, this neonate was managed by simple excision of the exstrophy bladder, without requiring osteotomy [7], similar to our case.

Fabrizio et al have presented a unique method of reconstructing the urethra by tubularizing the exstrophic plate while sparing internal covered bladder. This case demonstrates the wide variety of re-constructive procedures that can be utilized for the repair of exstrophy-epispadias complex [8].

Although exstrophy variants are quite confusing, a high index of suspicion, a normal appearing phallus, and inability to demonstrate ureteric orifices on exstrophied bladder should confirm the diagnosis. Hence recognition of exstrophy variant is important because the treatment and prognosis are very different.

Conclusion

Although exstrophy variants are extremely rare, their pre-

operative diagnosis is important because treatment and prognosis are very different from classical exstrophy. The variant we describe was managed surgically with good functional and cosmetic results.

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