



Covered bladder exstrophy with heterotopic hind gut duplication cyst: A case report with review of literature

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ABSTRACT

There are many different variants of classical bladder exstrophy (BE) and covered BE is one of these rare variants which has an association with heterotopic hind gut duplication cyst. We report a case of a 20 months old girl with covered BE, who had a 5 cm cystic mass attached anteriorly to the bladder, which turned out to be a hind gut duplication cyst on histopathology examination. So a possibility of a heterotopic hind gut duplication cyst should be kept in mind if a cystic mass is found anterior to a covered BE.

Key Words: Bladder exstrophy, hind gut duplication cyst, covered bladder exstrophy.

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Introduction

Bladder exstrophy (BE) is a congenital genitourinary malformation belonging to the spectrum of the exstrophy-epispadias complex [1]. It is characterized by an open bladder plate over the anterior abdominal wall, epispadias or bifid clitoris, divergent recti, a wide pelvic diastasis and rotation deformity of the pelvis [1]. Covered BE is a rare variant of Classical BE and consist of all the features of a classical BE but has a closed bladder with thin intact

overlying skin [1-3]. The incidence of BE is about 1 in 30,000-50,000 live births, while the variants are estimated to be 10 times less frequent than the classical exstrophy [1-3]. So far there has been no proper classification for these variants. Covered BE has a known association with heterotopic hind gut duplication cyst mainly colonic [1, 2-6]. To our knowledge no more than 15 cases have been reported so far of this association. We are reporting another case of heterotopic hind gut duplications cyst in a covered bladder exstrophy with review of literature.

Case report

A 20 months old girl was referred to our center as a case of epispadias from a secondary care

hospital. She was born at term with normal antenatal scans. The anomaly was missed at the neonatal examination. The family asked for medical advice as the child had abnormal voiding pattern. On physical examination in the clinic, she had low-set umbilicus with lower abdominal swelling, rectus abdominis diastasis, wide symphysis pubis, bifid clitoris, and normal vaginal and anal openings. Pelvic X ray showed pubic diastasis and bilateral developmental dysplasia of hip joints (Fig. 1). Ultrasound renal tract was unremarkable. Micturating cystourethrogram revealed a normal bladder with no vesicoureteral reflux. A diagnosis of covered BE was made and surgery was planned to repair it.



Fig. 1. Pelvic X-ray revealed features of bilateral DDH and pubic diastasis.

During surgery she underwent cystoscopy first, which showed the urethra was 1.5 cm from the bladder neck to the external urethral meatus. There was normal bladder wall and normal ureteric openings with no abnormal findings. It was followed by bilateral pelvic osteotomy by orthopedics team. To repair the exstrophy the bladder was exposed through a

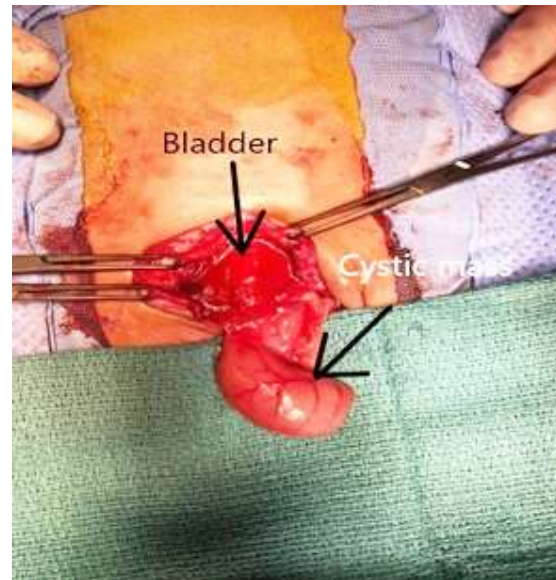


Fig. 2. The cystic mass anterior to bladder.

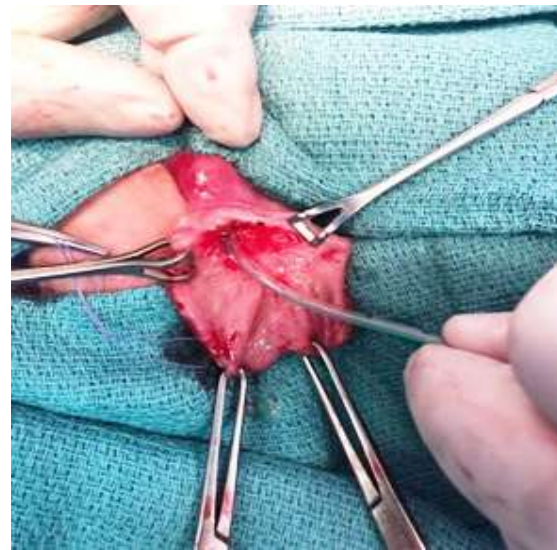


Fig. 3. The cyst after incision.

midline incision. During dissection of the bladder, which appeared otherwise normal, a 5cm cystic structure was found anterior to it. It was separate from the bladder wall. After initial aspiration it was opened and we found it to be filled with mucous, with no communication with, either the bladder or any abdominal viscera including intestine (Fig. 2 and 3). There were no further cysts in the small bowel or large bowel. The cyst was completely

excised and sent for histology. The bladder was freed all around and dropped down into the pelvis. Bladder neck was not repaired. The pubic symphysis was approximated, recti brought to midline, the abdominal wall was closed in layers with no tension and the umbilicus was refashioned. Postoperative recovery was uneventful.

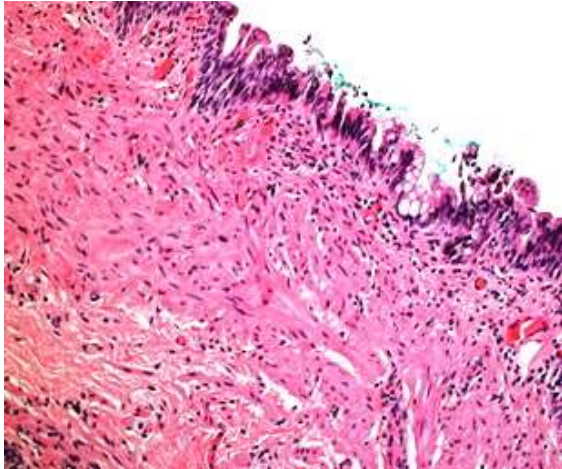


Fig. 4. Histology showing duplication cyst of colon with well-formed all layers. M: Mucosa; SM: Sub mucosa; MM: Muscularis mucosa; S: Serosa.

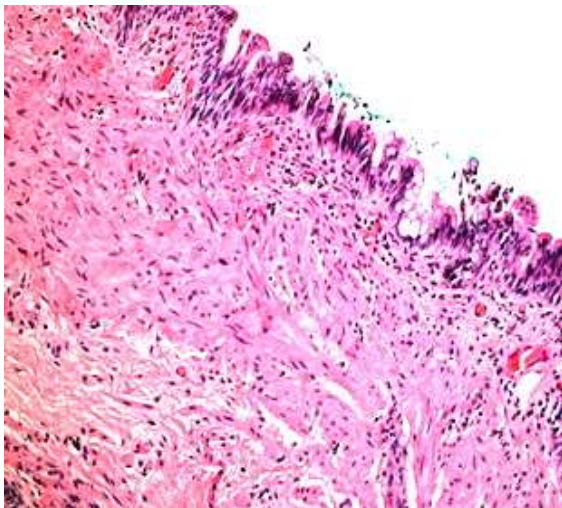


Fig. 5. Transitional epithelium likely representing the mid zone of anal canal.

Histology of the cyst showed normal colonic mucosa and submucosa, inner circular and

outer longitudinal muscularis propria and serosa. There was also a transition from normal colonic epithelium to anal transitional epithelium (Fig. 4 and 5). The only abnormality noted was absence of ganglion cells (Fig. 4). These features were suggestive of heterotopic hind gut duplication cyst.

On follow up, 4 months after surgery at 2 years of age, she was still not potty trained but did have dry intervals. We intend to assess her bladder function after potty training age and plan further management accordingly.

Discussion

The embryological origin of BE is due to abnormal bladder development and malformation of the ventral wall of abdomen. Mildenberger et al. [7] suggested that the BE and their variants result primarily from the abnormal persistence of the caudal position of the insertion of the body stalk on the embryo which prevents normal advancement and interposition of normal mesenchymal tissue.

Intestinal duplications are either cystic or tubular could be found anywhere from esophagus to anus [8]. These are lined by intestinal mucosa with smooth muscles in their wall and are normally attached to adjacent bowel with which they were shared a common blood supply. In covered exstrophy, the duplication cyst is neither adjacent to the intra-abdominal bowel nor share blood supply with it and for that reason it is called heterotopic duplication cyst. The embryological explanation of this has been proposed by Arin et al. [6]. They believe that it represents a common link BE and cloacal exstrophy with the large un-ruptured cloacal membrane as the cause of BE features and a “snared-in” bowel leading to the sequestered bowel duplication cyst. Somehow this snared in loop lose connection with the rest of the bowel, which

restores its continuity. However, Cerniglia et al. [9] suggested delayed invasion of the mesoderm with secondary closure of the membrane after it is ruptured either completely or partially may leave a sequestered loop of bowel over the ventral surface of bladder with no communication with intra-abdominal viscera.

There are many variants of classical BE which includes superior vesical fissure, superior vesical fistula, inferior vesical fistula, duplicate exstrophy, split symphysis variant, pseudo exstrophy, covered exstrophy and pubic umbilicus [1-6]. Attempts have been made to classify these variants but due to rarity of the condition, a lot of crossover of various subtypes and naming the same condition with different names have resulted in a great deal of confusion [3,6,10-12]. A simpler classification has been proposed by Maruf et al. [3] who subdivided BE variants into 3 broad categories: 1) Covered/skin-covered BE, in this there is a thin sheet of skin below the umbilicus, with an intact bladder present just beneath this thin layer of skin. The recti are also laterally displaced. 2) Superior vesical fissure (SVF). In this there is a defect in the anterior abdominal wall, which communicates with partially opened up bladder. Depending on the size it can be called SVF or superior vesical fistula. 3) Duplicate BE in which the bladder is duplicated either in the anterior posterior direction or side to side. Additionally, all these variants have typical skeletal abnormalities of BE with varying degree of epispadias.

So far not more than 15 cases of covered BE with heterotrophic bowel sequestration have been reported. In covered BE without sequestration, male slightly predominate as compared to females but due to small number of covered BE with sequestration this cannot

be deduced [3]. Similarly covered BE without sequestration has known association with gastrointestinal anomalies such as omphalocele, anorectal malformations, intestinal duplication, and gastroschisis; neurological anomalies such as hemivertebrae, and genitourinary anomalies; such as undescended testes, solitary kidneys, ectopic kidneys and mega ureter. This has yet to be established in cases of covered BE with sequestration due to small number of cases reported so far [1-4,6,9]. As the embryological origin of covered BE with and without sequestration appears to be similar, we feel that these associated anomalies may be similarly present in covered BE and should be searched for.

The general principles of repair are similar to the repair of classical bladder exstrophy with some modifications. These include correction of pelvic anatomy by performing bilateral osteotomy as we did in our case. Extensive mobilization of the bladder from the surrounding structures, pushing bladder into pelvis and a tension free abdominal wall closure [1-3,6,9]. As the bladder is completely developed so it does not need to be opened and for similar reasons ureteric re implantation is not needed if there is no vesicoureteral reflux on MCUG. The bladder neck repair can be done around 4 years of age if indicated [1,3,6,9]. We did not do any bladder neck repair in our case. We will assess the patient around 4 years of age and if needed we will consider it. Other issues like bladder neck incompetence, VUR and small bladder capacity can be managed in a similar way as classical BE [1,3,9].

As these patients have epispadias with possible involvement of bladder neck, one has to follow them carefully to assess urinary incontinence. Though preliminary outcomes in single case

reports suggest good outcome in terms of continence but long term follow up by Maruf et al suggest that nearly all of the patients of BE variants including covered BE with and without bowel sequestration, despite bladder neck repair will need either augmentation or bladder neck closure or both to remain dry [1-4,6].

Conclusion

Covered bladder exstrophy has association with heterotrophic bowel sequestration. The principles of surgical management are similar to classic BE repair. Long term follow up is needed to assess continence.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: Patient confidentiality has been maintained and written consent for the publication of patient details and clinical pictures have been obtained from the patient's father and can be furnished when required.

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