Extrophy of Bladder

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ABSTRACT

Bladder extrophy is an uncommon congenital anomaly of lower abdominal wall which is detected and managed at birth. As the bladder is open there is usually no upper tract involvement in these cases. Upper tract dilatation may be seen after successful bladder neck repair due to associated anatomical vesicoureteric reflux. In this case report we present an unusual association of bladder extrophy with bilateral upper tract involvement prior to bladder neck repair.

Key words: Ectopia vesicae; extrophy of bladder; extrophy-epispadius complex

Introduction

Bladder extrophy is a rare congenital anomaly in which bladder is inside and sticks out of the abdominal wall. The urine produced by the kidneys drain into this open area and is not stored normally in the urinary bladder. The incidence of bladder extrophy is about 1 in 30,000 to 50,000 births and is more common in males. Male to female ratio ranges from 1.5:1 to 5:1.[1] The cause of extrophy of bladder is not known. It varies in its severity. The pubic bones do not join thus leaving a wide opening and this causes hips to be externally rotated. In girls, the clitoris and labia minora are separated and spread apart, the vagina and urethra are shorter. The uterus, tubes and ovaries are generally normal.

Case Report

A 12-year-old female child presented to us with an obvious extrophy of bladder with the complaints of bilateral flank pain more on left side. She had undergone an attempt of primary bladder closure without bladder neck repair at new-born period which dehisced on the 3rd day of surgery. On examination, it was found that her stature was short as compared to her age and there was an extrophy bladder plate without any gross squamous metaplastic changes [Figure 1]. Both the ureteric orifices were normal and were showing normal efflux of urine. There was, however, operative scar of previous surgery visible just adjacent to the bladder. Her left kidney was palpable on bimanual examination.

Physical examination revealed general condition to be fair, pallor was present, hydration was normal, afebrile, pulse was 80/min, regular; blood pressure was 110/70 mmHg, respiratory system, cardiovascular system, and central nervous system were normal. Urinary bladder was visible in the hypogastrium covering partly the pubic region. Renal lump was palpable on both sides.

Hematology revealed hemoglobin 9.7 g/100 ml, Total leucocyte count 4.9 × 10^9/L; differential leukocyte count: Neutrophil 54%, lymphocytes 34%, eosinophils 4%, basophils 0%; polycythemia vera 26.5% total red cell count 3.39 × 10^{12}/L, mean cell volume 78 fl; mean cell hemoglobin (MCH) 28.6 pg; MCH concentration 36.6 g/dL, blood urea 22.6 mg/dL, serum creatinine 0.8 mg/dL, serum calcium 1.18 mmol/L, and serum phosphate 3.89 mg/dL.

Access this article online	
Quick Response Code:	Website: www.wajradiology.org
	DOI: 10.4103/1115-3474.172091

How to cite this article: Diwakar RK, Khurana O. Extrophy of bladder. West Afr J Radiol 2016;23:150-3.

Urine examination was normal.

Kidney, ureter, and bladder radiograph showed diastasis of pubic bones, enlarged kidneys with lower pole left kidney reaching up to the left iliac crest and soft tissue mass like extrophied urinary bladder in the lower pelvis [Figure 2].

Routine ultrasonography (USG) revealed enlargement of both the kidneys with marked hydroureteronephrosis on the right side. Left kidney showed poor definition of wall of calices and renal pelvis, which were filled with echogenic foci suggesting possible pyonephrosis [Figure 3].

Intravenous urography showed marked hydronephrosis of right kidney with hydroureter where the dilated distal end of ureter was seen in the pelvis. Left kidney showed delayed excretion of contrast with poorly defined calices and nonopacification of left ureter in films taken after 6 h and 24 h [Figure 4].

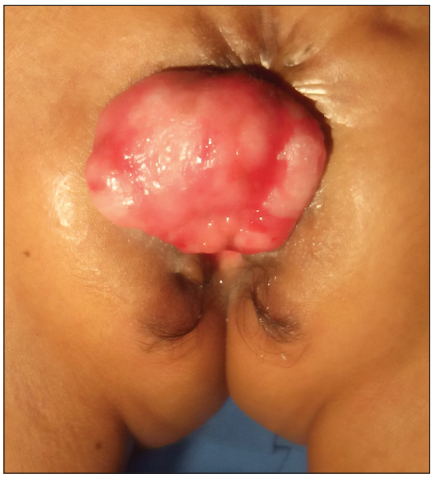

Figure 1: Preoperative photograph

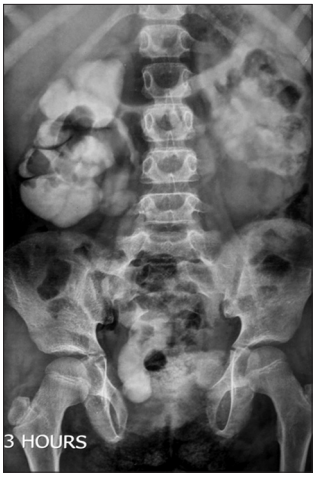

Figure 3: Delayed (6 h) intravenous urography skigram with marked hydronephrosis with dilated tortuous right ureter and poor opacification of calices in the left kidney

Computed tomography abdomen with contrast study revealed Grade IV bilateral hydronephrosis with thinning of renal cortex [Figure 5]. Extrophied bladder is seen in lower anterior abdominal wall with two ureteric openings on the sides [Figure 6].

99mTc-dimercaptosuccinic acid renal scan revealed enlarged right kidney with good tracer uptake with patchy distribution. Left kidney with irregular outline had reduced tracer uptake with uneven distribution. Scintigraphic evidence of scarring was not observed in either of the kidneys. The differential renal function was 73% in right kidney and 27% in left kidney [Figure 7].

During operation, it was found that there was a small defect in the lower abdominal wall with dense scarring through which the bladder was protruding. There was an obvious kinking of both ureters at this ring of fibrosis, which was possibly causing the proximal dilatation. As soon as the ring was released both the ureters could easily be cannulated with number 6 infant

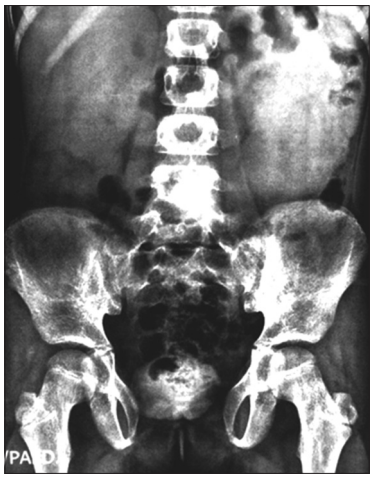

Figure 2: Kidney, ureter, and bladder skigram revealed enlarged left kidney and pubic diastasis

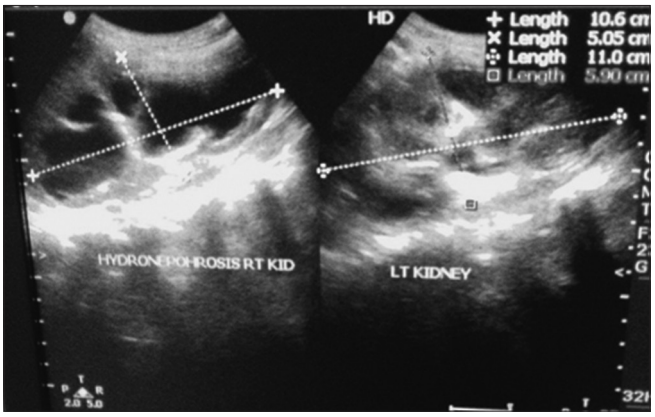

Figure 4: Renal ultrasonography showing enlargement of both kidneys with marked hydronephrosis and calices of left kidney filled with fine echoes

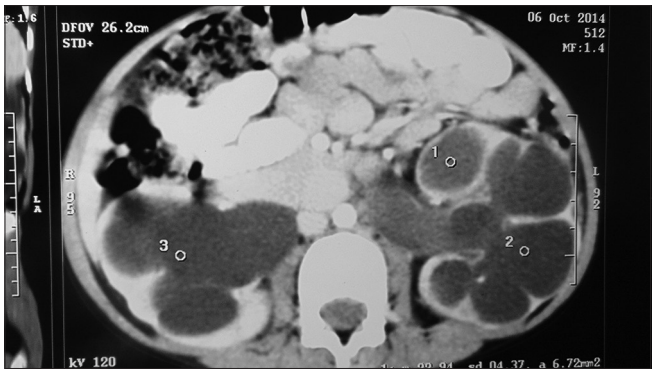

Figure 5: Grade IV bilateral hydronephrosis with thinning of renal cortex

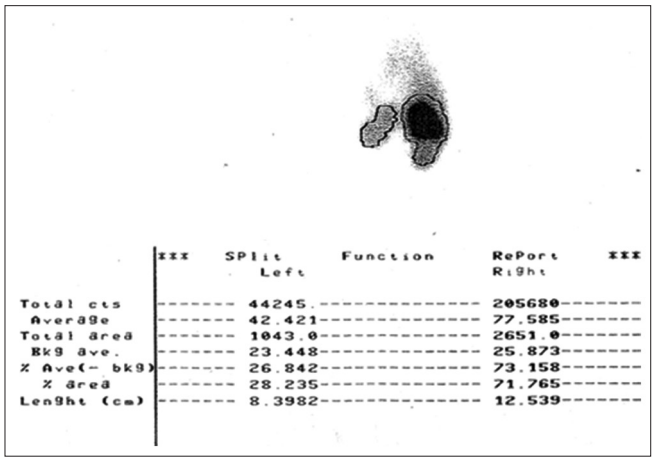

Figure 7: Differential renal function in 99mTc-dimercaptosuccinic acid scan

feeding tube [Figure 8]. The external opening of both the ureters in the bladder plate was wide. It was, thus, decided to do primary bladder closure with bladder neck repair on Bhatnagar's principle. In this procedure, the bladder plate is dissected circumferentially to free it completely of its attachment. The posterior urethra is tabularized up to just below the ureteric orifices. Moreover, then the bladder plate along with the neourethra is tabularized. It differs from other technique in that no part of the bladder tissue is used for buttressing the repair, but all are utilized for enhancing the bladder volume. The ureteric stents were removed 3rd week followed by the suprapubic catheter after 4 weeks. Postoperative period was uneventful. Renal USG, 4 weeks after the operation, revealed reduction in renal size.

Discussion

Bladder extrophy epispadius complex is an uncommon congenital defect of the lower abdominal wall. According to the accepted hypothesis, there is an abnormal persistence of the caudal portion of the insertion of the body stalk of the embryo.[3] As a consequence of this, the normal advancement and interposition of the mesenchymal tissue in the midline becomes impossible. The cloaca cannot be translocated backwards into the body cavity and the cranial end of the

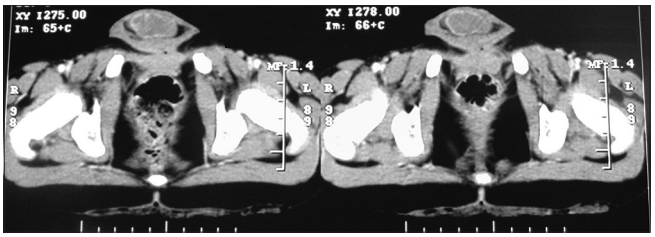

Figure 6: Computed tomography scan pelvis with extrophy of bladder

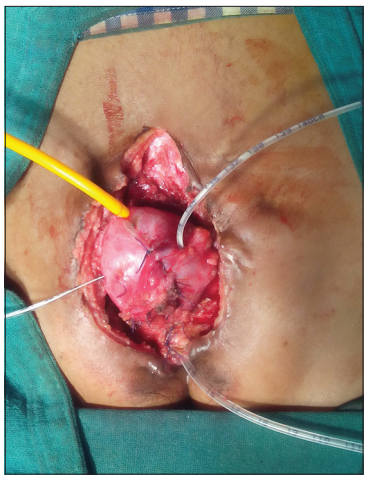

Figure 8: Operative photograph showing catheters in both ureters

cloacal membrane remains in contact with the inferior aspect of the low-set body stalk. This is in contrast to the previously abnormal rostral extension of the cloacal membrane. This ultimately leads to the eversion of the bladder mucosa which appears in the form of the extrophy.[2]

In females, the urethra is short often buried in the extrophied bladder. The clitoris tends to be bifid. The labia are also widely separated. The vagina is short and often stenotic. Uterine prolapse or unicornuate uterus may be present. Anus is anteriorly placed and may be patulous; this is more commonly seen in girls.[2]

Radiographic findings include a soft tissue mass extending from a large infra-umbilical anterior wall defect, which may be close to the umbilical arterial exits. Extrophy of bladder had widely been reported to be associated with the failure of the pubic bone to meet in the midline. As this condition is also sometimes associated with upper tract abnormalities a screening upper tract evaluation is advised in them. Bilateral hydroureteronephrosis is, generally, not associated preoperatively or after primary bladder closure due to the associated low pressure system owing to the open bladder. Once a bladder neck repair is done the open system gets converted to a closed system with an outflow resistance. As there is a lateral attachment of the ureters in the bladder plate the submucosal flap valve mechanism which normally happens and prevents reflux is absent. This case was unique due to the presence of gross upper tract dilatation in the

open system, which was due to the kinking of the ureters in the ring of fibrosis.

The approach in extrophy of bladder is a staged repair which involves the closure of the bladder plate with or without a bladder neck repair at 1st year of life. This is followed by an epispadius repair in males and monsplasty in females. As there is poor bladder growth in these patients the final procedure involves an augmentation of the bladder plate. This case was successfully managed by release of the ureteric kinks and primary bladder closure with bladder neck repair. In follow–up, she developed pyoureteronephrosis on the left side due to impacted pus flakes in the left ureteric orifice which was managed successfully by cystoscopy and flushing.

In untreated patients, due to continuous dribbling of urine, there can be mucosal erosions, infection, and squamous metaplasia resulting in acquired VUJ obstruction. The detrusor muscle may become fibrotic and scarred. Instances of adenocarcinoma of bladder have been reported in untreated adult patients.[2]

Another option is urinary diversion if continence is poor following bladder reconstruction. This can be done by utero-sigmoid anastomosis or formation of diversion of ileal conduit, colonic conduit, or continent urinary diversion.[3] Complications include stricture at the site of anastomosis, increased chances of adenomas and adenocarcinomas at the site of ureterocolic anastomosis and hyperchloremic acidosis.[3]

Surgical management of bladder extrophy demands patience and perseverance. The stages/procedures used in the surgical reconstruction of bladder extrophy includes: Primary closure of bladder extrophy with anterior abdominal wall reconstruction, bladder neck repair after 1 year of primary closure, ureteric reimplantation, and augmentation colocystoplasty in various combinations.

Renal USG is done to rule out renal agenesis, hydronephrosis and ectopic kidney. After bladder reconstruction is done, USG is done to look for upper tract deterioration which may result from increased bladder pressure or repeated infection.[4]

Evaluation methods include clinical evaluation and urodynamic assessment. Urodynamic assessment has emerged as an essential tool in the follow-up of these patients. Anticholinergic medication with imipramine or oxybutinin is a useful adjunct in the overall management.[5]

The role of pelvic osteotomy to achieve closure of the abdominal wall and in the achievement of continence is controversial.[5]

Follow-up studies include renal biochemistry, the upper tracts are monitored with radionuclide renography and ultrasound scans, and the bladder is evaluated with micturating cystourethrography and sonography for size, shape, residual urine, and VUJ reflux. Urodynamic studies are also done.[5]

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References

1. Gearhart JP, Jeffs RD. Extrophy-Epispadius complex and bladder anomalies.In-Campbell MF, Relik AB, Vaughan B. Campbell's Urology, 7th edition, Pholadelphia; Ps: WB Saunders 1998. p 1939-1990.
2. Mildenberger H, Kluth D, Dziuba M. Embryology of bladder exstrophy. J Pediatr Surg 1988;23:166-70.
3. Silverman FN, Kuhn JP. Abnormalities of pelvo-caliceal system, ureter, bladder and urethra. Epispadius-extrophy complex. In: Caffey's Paediatric X-ray Diagnosis: An Integrated Approach. Ch. 34, Sec. 3. Philadelphia: Mosby; 1993. p. 1298-301.
4. de Bruyn R, Gordon I, McHugh K. Imaging of the kidneys and urinary tract in children. In: Grainger and Allison's Diagnostic Radiology - A Textbook of Medical Imaging. 4th ed. Vol 2. Churchill Livingstone, London: Edinburgh 2001. p 1733-4.
5. Bhatnagar V, Lal R, Agarwala S, Mitra DK. Bladder-neck repair in urinary bladder exstrophy. Pediatr Surg Int 1999;15:290-3.