

## SURGICAL MANAGEMENT OF ECTOPIA VESICAE- A TERTIARY CARE CENTRE- OUR EXPERIENCE WITH THREE CASES

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### PRESENTATION OF CASE

The first patient is a 30-year-old female who presented with an irregularly fungating growth (Figure 1) just above the anteriorly placed vagina in the area of the exposed urinary bladder. She gives history of surgery for exstrophy bladder at 5 years of age. Since the last six months, she noticed that the swelling in the lower abdomen was prone to ulceration, infection and bleeding and was referred to our center from local hospital.

On examination, labia majora was found to be separated at the mons area. There were no significantly enlarged inguinal lymph nodes. Gross divarication of recti was found. Systemic examination was normal. Secondary sexual characters and breasts were well developed.

X-ray pelvis showed symphysis diastases. Routine laboratory investigations including ultrasound of abdomen and chest x-ray were within normal limits. Contrast-enhanced CT scan of the abdomen and pelvis with CT urogram showed the localised growth from the bladder wall with no ureteric involvement. Iliac/retroperitoneal region showed no significant lymphadenopathy. Both ureters showed moderate dilatation up to the bladder due to narrow orifices that were difficult to locate on the bladder wall. Vaginoscopy revealed a roomy vagina and normal cervix. Biopsy was taken from representative sites from the mass.



**Figure 1. Preoperative Image of Patient-1**

The second patient is a 28-year-old female who presented with ectopia vesicae with no prior history of any surgery (Figure 2). Local examination showed dysplastic changes on the skin and a few areas suspicious of malignancy.

Biopsy from the bladder mucosa showed severe dysplastic changes, but no evidence of malignancy.



**Figure 2. Intraoperative Image of Patient-2**

The third patient is a 42-year-old male who presented with bleeding and infection from an ulcerating mass in the lower abdomen. He had undergone bladder closure as an adult and the CECT of abdomen revealed a fungating mass within the repaired bladder. The tumour had infiltrated into the lower ureters bilaterally. Biopsies were taken from the mass and showed malignant transformation.

**CLINICAL DIAGNOSIS-** Adult ectopia vesicae.

**DIFFERENTIAL DIAGNOSIS-** Malignancy arising from patent urachus.

### SURGICAL MANAGEMENT

Ectopia vesicae (bladder exstrophy) is a rare congenital anomaly characterised by failure of closure of abdominal wall and urinary bladder. The baby is born with a mucosal mass in the lower abdomen that protrudes between the umbilicus and symphysis pubis. Ectopic vesicae are more common in males.

As surgical correction is often made during childhood, it is rare to see adults with this condition. The management of these patients is demanding and depends on the clinical situation and the surgeon's experience. Extensive reconstructive procedures are required in all cases, but each case is highly individualistic.

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In this case series, two patients had developed malignant transformation of their exstrophic bladders and the third had severe dysplastic changes in the exposed mucosa. Surgical management in each case was different as the situation demanded. There is no singular principle for operating on patients with ectopia vesicae.

**Patient-1-** Patient was taken up for radical cystectomy and pelvic lymphadenectomy (Figures 3, 4). Urinary diversion was in the form of continent Mainz pouch using 20 cm of terminal ileum, caecum and ascending colon with ileocaecal valve as the antireflux mechanism. Continence was achieved by using appendix as appendicostomy (Figure 5). The defect in the abdominal wall was closed with musculocutaneous pedicle flap from anterolateral thigh (Figure 6). Postoperative period was uneventful. Patient was put on regular pouch washes with sodium bicarbonate solution. The pouch capacity on follow up was found to be on an average about 530 mL. Patient is on CIC every 3-4 hours through the appendicostomy and is under regular follow up.

Histopathology showed well-differentiated squamous cell carcinoma of the excised bladder segment with free surrounding margins including ureter and no positive lymph nodes.



**Figure 3. Peroperative Image of Patient-1**



**Figure 4. Identification and Stenting of Ureters (Post Cystectomy Image of Patient-1)**



**Figure 5. Mainz Pouch with Appendicostomy as Catheterisable Stoma of Patient-1**



**Figure 6. Postoperative Appearance (Midline Defect was Covered with Musculofascial Flap from Anterolateral Thigh) Patient-1**

**Patient-2-** She underwent cystectomy and pelvic lymphadenectomy. The defect on the anterior abdominal wall was repaired with musculocutaneous flap from left anterolateral thigh. We chose to perform urinary diversion in the form of ileal conduit as the patient was too uneducated and unwilling to perform intermittent catheterisation (Figure 7). Postoperative period was uneventful.

Histopathology report of the cystectomy specimen and pelvic nodes was similar to preoperative report. There was no evidence of malignancy, only severe dysplastic changes.



**Figure 7. Postoperative Appearance of Patient-2**

**Patient-3-** Patient underwent radical cystectomy with penectomy and pelvic lymphadenectomy. Since the ureteric length was short after radical excision, bilateral ureterostomies were performed before the abdominal wall defect was closed with anterolateral thigh flap.

Histopathology showed high-grade squamous cell carcinoma with infiltration into both ureters. But, the pelvic nodes were free of neoplasm.

**Postoperative Follow up-** All the three patients are doing well and on regular follow up with CT urogram, conduitogram, pouchogram, urine cultures, quality of life questionnaires and routine investigations. Our follow up is short, but the results so far are highly encouraging. The patient with Mainz pouch is able to hold urine for around four hours without urine leak. Till far, there have been no infective or other complications during follow up. Quality of life has improved dramatically and one patient has already applied for work with agencies.

**DISCUSSION MANAGEMENT-** Ectopia vesicae is a congenital anomaly in the spectrum of epispadias-exstrophy complex with a prevalence of 3.3 per 1,00,000 births. It is two to three times more common in males than females (2.3-6:1).<sup>1,2</sup> Risk of recurrence in any family is 1:100. The child is born with anomalies of the genital and urinary systems, anterior abdominal wall, bony pelvis and pelvic floor.<sup>3</sup> The aetiology of bladder exstrophy is unknown and there is no definite inheritance pattern. Risk factors include Caucasian race, maternal tobacco usage, young age of mother, multiparity and assisted-reproductive techniques.<sup>4</sup>

#### Clinical Features

- 1) Mucosal mass in the lower abdomen representing the exstrophied bladder in the defect between two divergent rectus muscles with two ureteric orifices opening laterally leading to continuous urine dribbling. Inferiorly, the bladder plate is seen in continuity with the urethra.
- 2) Umbilicus is low set and seen at the upper border of the exstrophied bladder.

The pubic bones (pubic diastasis) are widely separated and connected by a strong ligament. In plain radiograph of the pelvis, this appearance has been likened to a manta ray (manta ray sign) (Figure 8) (Bull, M. 2014).



**Figure 8. X-Ray of KUB of Patient-1**

- 3) Inguinal rings are wide with risk of inguinal hernias.
- 4) The pelvis is externally rotated.
- 5) Anus maybe displaced anteriorly with a normal sphincter.
- 6) Upper urinary tracts are always normal.

#### In Females-

- a) Labia are separated and the vaginal orifice is visualised anteriorly. Clitoris is bifid and visualised beneath the separated labia.
- b) Vaginoscopy shows normal-appearing vagina, uterus and cervix. Patients are usually fertile, but prone to develop prolapse due to the weak pelvic floor muscles.

#### In Males-

- a) The testes are normal, usually descended in an anteriorly-displaced scrotum.
- b) The penis is small, broad and epispadiac with dorsal chordee and ventral hooded prepuce. It is located at the lower border of the bladder plate and turned upwards. The glans is open and urethral plate can be visualised the entire length of the penis. The urethral meatus maybe located beneath the bladder neck or maybe penile or glandular. The penis and scrotum are widely separated. In some cases, there is complete nonunion of the two halves of the penis and scrotum.<sup>3</sup>
- c) Bilateral inguinal hernia maybe present.
- d) The prostate and seminal vesicles are rudimentary.
- e) The corpora cavernosa are dissociated from their bony attachment and displaced back in the perineum.

#### Management Principles in Ectopia Vesicae in the

**Newborn-** In the first year of life, the urinary bladder is primarily closed following osteotomy of both iliac bones just lateral to the sacroiliac joints. Primary closure is indicated only in those patients with bladders that are contractile and elastic and adequate capacity.<sup>5</sup>

Once primary closure is achieved, the patient will be incontinent due to absent bladder neck sphincter. In males, epispadias is corrected around 6 months to 2-3 years of age and the bladder neck is reconstructed at around 4-5 years of age.

Complications of primary bladder exstrophy closure include the following.<sup>6</sup>

- 1) Failed surgical correction and wound dehiscence.
- 2) Bladder prolapse.
- 3) Bladder neck stenosis/urethral stricture/bladder outlet obstruction.
- 4) Bilateral vesicoureteric reflux and reflux nephropathy.
- 5) Recurrent bladder infection.
- 6) Bladder calculi/renal calculi.
- 7) Urethrocuteaneous fistula.
- 8) Pubic separation.
- 9) Osteotomy nonunion and persistent joint pain.
- 10) Limb anomalies.



**EMBRYOLOGY-** Ectopia vesicae develops due to defects in development of the pelvis minor and cloacal membrane. During 4-7 weeks of intrauterine development, the infraumbilical mesenchyme migrates between the ectodermal and endodermal layers of the cloacal membrane and divides it into primitive urogenital sinus anteriorly and the anorectal canal posteriorly.<sup>7</sup> This occurs at the same time as maturation of the anterior abdominal wall.<sup>8</sup> When mesoderm fails to migrate around the cloacal membrane, the anterior abdominal wall and anterior wall of urinary bladder fail to develop.<sup>9,10</sup>

Rarely, the cloacal membrane persists longer than usual and it prevents completion of developmental steps. Then, it becomes unstable and ruptures. If this rupture takes place at 6-8 weeks, classic bladder exstrophy develops. The lower urinary tract becomes exposed to the outside through the defect on the anterior abdominal wall. The pelvic ring normally closes by 8 weeks, but this is not achieved in bladder exstrophy. The visceral structures attached to the pubis do not join in the midline and remain open. This bony defect is the most constant feature of bladder exstrophy. Bladder exstrophy consists of three conditions.

- Epispadias.
- Classic bladder exstrophy.
- Cloacal exstrophy where the bladder as well as the intestines are exposed externally along with other malformations.

### **PATHOLOGICAL DISCUSSION**

Exstrophy increases the risk of developing a bladder tumour by 700 times as compared to the general population.<sup>11</sup> There is no decrease in the risk for malignancy even if primary closure of the bladder is achieved during childhood. The exact cause of carcinogenesis in exstrophy is unknown. Various hypotheses such as recurrent infections, environmental exposure or genitourinary secretions have been proposed. Rieder et al have suggested that chronic irritation of the adjacent skin by urogenital secretions leading to metaplasia is the cause of SCC and adenocarcinoma in exstrophic bladder.<sup>12</sup> Smeulders et al have identified different types of metaplastic changes in the colonic epithelium of the untreated exstrophic bladder suggesting a potential intrinsic predisposition to malignant transformation.<sup>11</sup> Adenocarcinoma may also result from malignant degeneration of embryonic rests of gastrointestinal tissue.

Most of the malignant tumours (60%) associated with an exstrophy of the bladder occur during the fourth and fifth decades of life.<sup>13</sup> Of the remaining, about 20% each occur after 60 years and before 40 years. The most common tumour is adenocarcinoma (95%)<sup>14,15</sup> and 3% to 5% are squamous cell carcinomas.<sup>12,16</sup> Squamous metaplasia has been found in 80% of cases.<sup>16</sup>

Since uncorrected exstrophic bladders in adults are more prone to malignancy and dysplastic changes common, biopsy from these bladders on presentation is suggested. If there is any suspicion of malignancy, a primary from other

sites, especially the gastrointestinal tract in case of adenocarcinoma should be ruled out.

Radical cystectomy followed by urinary diversion remains the standard treatment for invasive bladder cancer. Several techniques for urinary diversion have been described in literature. Simon performed the first surgical urinary diversion into the bowel as a treatment for ectopia vesicae in 1851. In 1878, Smith performed ureterosigmoidostomy by anastomosing ureters to sigmoid colon. Nearly, one century later, Bricker introduced ileal conduit diversion in the early 1950s, while Camay created the first ileal neobladder in 1959. Orthotopic diversion became popular in the 1980s.

In ectopic vesicae, the fibrotic nature of the bladder wall and absence of sphincter at bladder neck makes reconstruction of a distensible and continent bladder impossible. Therefore, after cystectomy, a Mainz pouch with its continent reservoir has become the popular diversion in this condition. The reservoir is created by antimesenteric opening and spherical reconfiguration of the ileocecal segment and ascending colon. Ureters are implanted using submucosal tunnels into the colonic segment. Appendix is tunneled into the skin and acts as the catheterisable continence mechanism.<sup>17</sup>

Systemic chemotherapy/radiotherapy used to treat transitional cell carcinoma bladder are ineffective in non-urothelial carcinomas like SCC and adenocarcinoma. Hence, ablative surgery remains the only option in such situations.

### **FINAL (PATHOLOGIC) DIAGNOSIS**

Squamous cell carcinoma and severe dysplasia complicating adult ectopia vesicae.

### **CONCLUSION**

We conclude that cystectomy with urinary diversion in the form of intestinal pouches with self catheterisable stoma is the optimal approach of treatment for patients with ectopic vesicae as it vastly improves the quality of life in these patients. Ileal conduit should only be considered in old and unskilled patients, in patients with no healthy bowel segments and in patients who will have difficulty or are unwilling for CIC. Cutaneous ureterostomy and ureterosigmoidostomy should be considered only in desperate situations.

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