



## Case Report

# Adult Presentation of Untreated Exstrophy–Epispadias Complex with Squamous Cell Carcinoma: A Rare Case Report

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### Abstract

Exstrophy–epispadias complex (EEC) is a rare congenital anomaly characterized by a midline defect of the lower abdominal wall involving the urinary bladder, external genitalia, and pelvic bones. Early diagnosis and surgical reconstruction in the neonatal period is the standard of care. Untreated cases persisting into adulthood are extremely uncommon and are associated with a significant risk of malignant transformation of the exposed bladder mucosa. We report a rare case of untreated exstrophy–epispadias complex in a 38-year-old male who presented with an ulcerated bladder plate that was subsequently diagnosed as invasive squamous cell carcinoma. The patient underwent organ-sparing radical cystectomy

with continent urinary diversion using an ileocecal segment and a Mitrofanoff catheterizable channel, along with epispadias repair and umbilicoplasty. Histopathology confirmed grade II invasive squamous cell carcinoma with tumor-free surgical margins. This case highlights the importance of early diagnosis and treatment of bladder exstrophy and demonstrates the potential for malignant transformation in long-standing untreated cases.

### Keywords

Exstrophy–epispadias complex; adult presentation; bladder exstrophy; squamous cell carcinoma; radical cystectomy; Mitrofanoff procedure

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### Introduction:

Exstrophy–epispadias complex (EEC) represents a spectrum of rare congenital anomalies involving the urinary bladder, urethra, genitalia, abdominal wall, and pelvic bones. The anomaly results from abnormal development and premature rupture of the cloacal membrane during early embryogenesis. The estimated

incidence of bladder exstrophy is approximately 1 in 10,000 live births, with a male predominance ranging from 1.5:1 to 6:1 [1].

With modern advances in pediatric urology and neonatal surgery, most patients undergo staged reconstruction shortly after birth. As a result, untreated cases reaching adulthood are extremely rare. Chronic exposure of the bladder mucosa to the external environment predisposes patients to persistent inflammation, metaplastic changes, and malignant transformation over time [2]. Although adenocarcinoma is the most frequently reported malignancy arising in exstrophic bladders, squamous cell carcinoma has also been reported in long-standing untreated cases [3].

We report a rare case of untreated exstrophy–epispadias complex presenting in adulthood with invasive squamous cell carcinoma of the bladder.

### Case Report

A 38-year-old man presented with a painful ulcerated infra-umbilical mass associated with continuous dribbling of urine and occasional bleeding from the

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lesion. According to the patient's history, he had been born with a fleshy infra-umbilical mass and abnormal genitalia with continuous leakage of urine from the exposed bladder plate.

His parents initially sought medical advice from physicians at a local upazila hospital. They were informed that surgical correction would be difficult and that the outcome might be uncertain. Consequently, no treatment was undertaken during childhood. During early childhood, the patient's mother covered the exposed bladder plate with cotton and cloth to absorb urine. Later in life, the patient used a polythene bag as a urine collector.

Despite the congenital anomaly, he remained otherwise healthy and worked as a manual laborer in his village. Three months prior to presentation, he noticed foul-smelling discharge from the bladder plate along with rapid enlargement of the mass, ulceration, and intermittent bleeding.

Clinical examination revealed a 5 × 5 cm irregular ulceroproliferative growth arising from the exposed bladder plate with areas of slough and necrosis. The bladder plate demonstrated visible ureteric orifices and an open bladder neck with exposed urethral plate. The scrotum was well developed and both testes were palpable. The penis was short and broad. Systemic examination revealed no other abnormalities.

Preoperative biopsy of the lesion revealed invasive squamous cell carcinoma (grade II). Ultrasonography of the abdomen did not reveal any metastasis or upper urinary tract abnormalities.

The patient underwent surgery through a lower midline incision encircling the bladder plate. The peritoneum was carefully separated from the bladder. Six-French feeding tubes were inserted into both ureteric orifices and the ureters were mobilized. En-bloc excision of the bladder with the tumor was performed while preserving the prostate, vas deferens, and seminal vesicles, constituting an organ-sparing radical cystectomy.

Epispadias repair was performed using the modified Cantwell–Ransley technique over a 14-Fr catheter. A 15-cm segment of terminal ileum with the caecum and proximal ascending colon was isolated with its vascular pedicle to construct a heterotopic continent urinary reservoir. Intestinal continuity was restored with ileo-ascending colon anastomosis. The appendix was utilized to create a Mitrofanoff catheterizable channel, which was brought out through the right iliac region.

The ureters were implanted into the ileocecal reservoir. Drains were placed in the pelvis and Morrison's pouch, and the abdomen was closed in layers with umbilicoplasty.

Oral feeding was started on postoperative day six. Drains were removed gradually and ureteric stents were maintained for one month. The patient was trained in clean intermittent catheterization (CIC). Final histopathology confirmed invasive squamous cell carcinoma grade II with tumor-free margins.

## Discussion

Bladder exstrophy results from abnormal development of the cloacal membrane and failure of mesodermal migration during early embryogenesis. The timing of cloacal membrane disruption determines the severity of the anomaly within the exstrophy–epispadias spectrum [1].

Untreated exstrophic bladder mucosa is exposed to chronic irritation, infection, and mechanical trauma. These conditions predispose the mucosa to metaplastic and dysplastic changes that may eventually lead to malignant transformation [4].

The incidence of malignancy in untreated bladder exstrophy has been reported to range between 3.3% and 7.5%. Among these malignancies, adenocarcinoma accounts for nearly 90% of cases, whereas squamous cell carcinoma represents approximately 5% [5].

Several authors have reported cases of malignant transformation in adult patients with untreated exstrophy. Radical cystectomy with urinary diversion is generally recommended due to the poor functional capacity of the exstrophic bladder and the high risk of malignancy [6].

Recent studies also emphasize that adult presentation of untreated bladder exstrophy remains rare and is mostly reported from resource-limited regions where access to pediatric surgical care is limited [7,8]. In our patient, lack of awareness and limited access to specialized surgical services contributed to delayed treatment until adulthood.

In the present case, we performed organ-sparing radical cystectomy with creation of a continent urinary reservoir using an ileocecal segment and a Mitrofanoff catheterizable channel. This approach provides urinary continence and improves quality of life compared with non-continent diversion techniques.

## Conclusion

Untreated exstrophy–epispadias complex presenting in adulthood is extremely rare. Long-standing exposure of the bladder mucosa significantly increases the risk of malignant transformation. Early diagnosis and neonatal surgical reconstruction remain the cornerstone of management. In adult patients presenting with malignancy, radical cystectomy with appropriate urinary diversion offers a definitive treatment and can provide satisfactory functional outcomes.



Figure 1. Clinical photograph of untreated bladder exstrophy in a 38-year-old male showing an ulcerated growth arising from the exposed bladder plate.

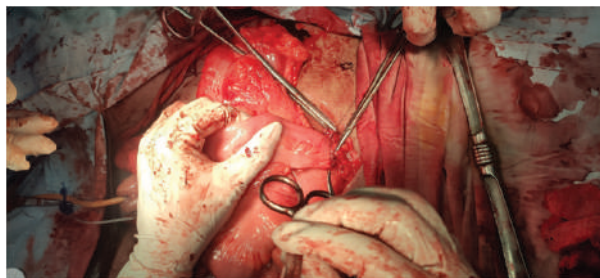


Figure 2. Intraoperative photograph demonstrating ureteric implantation during construction of the continent urinary reservoir.



Figure 3. Postoperative appearance after organ-sparing radical cystectomy with continent urinary diversion using Mitrofanoff appendicovesicostomy and modified Cantwell–Ransley epispadias repair

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