



Oncology

Adenocarcinoma of the Ileal Conduit in a Patient Born With Classic Bladder Exstrophy

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ABSTRACT

Bladder exstrophy is a rare birth defect that typically requires patients to undergo multiple surgical procedures throughout the course of their childhood. Many ultimately undergo operations that use segments of bowel for the reconstruction and/or augmentation of the urinary tract, which imparts an increased risk of malignancy in these patients. We present the case of a 59-year-old man with a history of bladder exstrophy managed with ureterosigmoidostomies revised to an ileal conduit who developed a large adenocarcinoma in the ileal conduit that extended into small bowel, sigmoid colon, and ureter.

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Introduction

Bladder exstrophy is a rare birth defect that typically requires multiple operations using bowel segments for urinary tract reconstruction and/or augmentation. When bowel is incorporated into the urinary tract, the risk of neoplasia increases¹; thus adult patients with a history of bladder exstrophy require close surveillance for potential malignancies.

In the following presentation of a 59-year-old man born with bladder exstrophy, a large adenocarcinoma involving small bowel, sigmoid colon, and ureter developed in an ileal conduit revised from ureterosigmoidostomies.

Case presentation

A 59-year-old man with classic bladder exstrophy presented with abdominal pain. Prior to 10 years of age, he underwent bilateral ureterosigmoidostomies, which were revised to an ileal conduit at age 18 when an 8-cm benign adenomatous sigmoid colon polyp necessitated sigmoid colectomy. He required multiple

ileal conduit revisions. Most recently, an additional segment of ileum was used to “patch” the distal aspect of the existing ileal conduit at age 44. His last surveillance colonoscopy, performed the year before presentation, demonstrated no suspicious lesions.

The patient had a history of recurrent left lower quadrant pain and abdominal fullness managed with nil per os and antibiotics. When such an episode did not resolve with this regimen, a non-contrast computed tomographic (CT) scan of the abdomen and pelvis revealed left hydronephrosis to the level of the conduit with renal parenchymal thinning and a soft tissue density in the conduit. Dynamic nuclear renal imaging with Tc99m-mercaptoacetyltriglycine (MAG3) and lasix confirmed left ureteral high-grade obstruction and 93% and 7% split function of the right and left kidneys, respectively. On immediate transfer to the authors’ institution for further management, CT scan confirmed severe left hydronephrosis and an infiltrating 4-cm soft tissue mass in the region of the ureteroenteric anastomosis of the ileal conduit, most prominent on the left side, involving adjacent small bowel loops and partially encasing the left iliac artery, that was not appreciated on CT scan 1 year before.

Serum creatinine was elevated to 2.2 mg/dL (baseline 0.9 mg/dL). A retrograde ureteral stent could not be placed, so a right-sided percutaneous nephrostomy tube was inserted, eliciting a decrease in serum creatinine to 1.4–1.6 mg/dL. At endoscopic evaluation, a large mass visualized in the ileal conduit was biopsied. Pathology demonstrated mild fibrosis and chronic inflammation but no evidence of tumor. Colonoscopy revealed no suspicious lesions.

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On exploratory laparotomy by urology and general surgery, an infiltrative 15-cm tumor in the proximal ileal conduit was seen directly extending into the left ureter, sigmoid colon, descending colon, several loops of small bowel, and small bowel mesentery. It was situated over the aortic bifurcation, overlying both iliac arteries. A retroperitoneal lymph node dissection, extensive enterolysis, and subtotal omentectomy were performed. The mass was removed en bloc; the specimen included the left kidney, ureter, ileal conduit, sigmoid colon, and small bowel. The entire ileal conduit was resected. The right ureter was free of tumor, and a new ileal conduit was created with remaining terminal ileum. Coloproctostomy and enteroenterostomy were performed after large and small bowel resection, respectively.

Final pathology revealed a 6.2-cm moderately differentiated adenocarcinoma arising in the ileal conduit with mucinous features and angiolymphatic and perineural invasion. The tumor infiltrated through the ileal conduit muscularis propria into underlying fat and adjacent organs, including small bowel, sigmoid colon, and left ureter. Left iliac periarterial tissue contained adenocarcinoma with a positive surgical margin. One pericolic lymph node of 69 total lymph nodes contained metastatic adenocarcinoma.

At 2 months, the patient was doing well with a stable creatinine and no evidence of disease on CT scan.

Discussion

Carcinogenesis in urinary diversions incorporating bowel is well recognized in the medical published data. After ureterosigmoidostomy, the incidence of carcinoma is 2%–15%. This translates into a 500-fold increase in colon cancer incidence when compared with the general population. The average time between ureterosigmoidostomy and cancer diagnosis is 20–26 years. However, patients with bladder exstrophy can develop adenocarcinoma within just 10 years of urinary diversion.² Prolonged exposure of colonic mucosa to urine is believed to contribute to cancer risk after ureterosigmoidostomy. Although the exact mechanism has not been elucidated, many theories involve the interaction among urine, stool, intestinal mucosa, and the healing anastomosis.³ In screening colonoscopies after ureterosigmoidostomy, the presence of precursor lesions (dysplasia and adenomas) suggests an adenoma–adenocarcinoma sequence characteristic of colorectal cancer.⁴ Urinary conduits display histologic signs of chronic inflammation after chronic exposure to urine. Substances released by inflammatory cells may promote carcinogenesis, contributing to the development of dysplasia and eventual malignancy.¹

This patient had ureterosigmoidostomies for approximately 15 years until a benign polyp prompted sigmoid colectomy and creation of ileal conduit. Theoretically, sigmoid colectomy decreased the

malignancy risk conferred at the time of ureterosigmoidostomy. However, sigmoid colon and/or the ureterosigmoidostomy anastomoses may not have been completely resected. A case report of a patient with ureterosigmoidostomies revised to an ileal conduit after 8 months who developed poorly differentiated sigmoid colon adenocarcinoma at 36 years has been reported.⁵

In this case, the mass arose from the ileal conduit. In 2013, only 8810 new small bowel cancers have been estimated in the general population, in contrast to 102,480 for colon cancer.⁶ It appears, however, that chronic exposure of intestinal mucosa to urine in urinary diversions and bladder augmentation increases this risk.¹

Although strict guidelines for surveillance are not established, urologists advocate initiating yearly screening 10–20 years after incorporating bowel into the urinary tract. Some demonstrated that screening is not required for at least 15 years after augmentation or replacement cystoplasty.⁷ Owing to high malignancy risk after ureterosigmoidostomy, initiating yearly sigmoidoscopy after 10 years is recommended.⁸ This patient had a CT scan and colonoscopy the year before presentation, which showed negative findings. At diagnosis, colonoscopy again showed negative findings, but endoscopy of the ileal conduit revealed tumor. His last endoscopic evaluation was approximately 4 years before presentation; therefore, yearly endoscopy of the ileal conduit may have prompted earlier diagnosis.

Conclusion

This case highlights the importance of frequent screening in patients with major congenital urologic defects and urinary tract reconstruction and the importance of examining the entire reconstructed urinary tract during screening.

References

1. Austen M, Kälble T. Secondary malignancies in different forms of urinary diversion using isolated gut. *J Urol*. 2004;172:831–838.
2. Spence HM, Hoffman WW, Fosmire GP. Tumour of the colon as a late complication of ureterosigmoidostomy for exstrophy of the bladder. *Br J Urol*. 1979;51:466–470.
3. Azimuddin K, Khubchandani IT, Stasik JJ, et al. Neoplasia after ureterosigmoidostomy. *Dis Colon Rectum*. 1999;42:1632–1638.
4. Stewart M, Macrae FA, Williams CB. Neoplasia and ureterosigmoidostomy: a colonoscopy survey. *Br J Surg*. 1982;69:414–416.
5. Khan MN, Naqvi AH, Lee RE. Carcinoma of sigmoid colon following urinary diversion: a case report and review of literature. *World J Surg Oncol*. 2004;2:20.
6. Siegel R, Naishadham D, Jemal A. Cancer statistics, 2013. *CA Cancer J Clin*. 2013;63:11–30.
7. Hamid R, Greenwell TJ, Nethercliffe JM, et al. Routine surveillance cystoscopy for patients with augmentation and substitution cystoplasty for benign urological conditions: is it necessary? *BJU Int*. 2009;104:392–395.
8. Woodhouse CRJ. Guidelines for monitoring of patients with ureterosigmoidostomy. *Gut*. 2002;51:v15–v16.