

Sigmoid Colon Adenocarcinoma at the site of prior, now defunctionalized, ureterosigmoidostomy for bladder exstrophy.

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Abstract

Patients with a history of ureterosigmoidostomy are at an increased risk of rectosigmoid adenocarcinoma. The cancer can occur many decades after the diversion, even if there was conversion to ileal urinary conduit. These patients may still be considered as high-risk individuals and screened for colorectal cancer accordingly.

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Abstract : Patients with a history of ureterosigmoidostomy are at an increased risk of rectosigmoid adenocarcinoma. The cancer can occur many decades after the diversion, even if there was conversion to ileal urinary conduit. These patients may still be considered as high-risk individuals and screened for colorectal cancer accordingly.

Key Clinical Message : History of ureterosigmoidostomy increases the risk of rectosigmoid adenocarcinoma decades after diversion even after conversion to ileal urinary conduit and these patients may be screened for colorectal cancer as high-risk individuals.

Keywords : Ureterosigmoidostomy, rectosigmoid adenocarcinoma, case report

Background

There is a 100-fold increase in risk for colorectal adenocarcinoma in patients who underwent urinary diversion by ureterocolic anastomosis, usually by ureterosigmoidostomy for bladder exstrophy [1]. After recognizing

this association, surgeons converted many patients who had previously undergone ureterosigmoidostomy as neonates to ileal loop urinary diversion when they reached adulthood. It has been recommended that patients who have an intact ureterosigmoidostomy undergo yearly screening flexible sigmoidoscopy, as well as patients who had been converted to another form of urinary diversion, but had the ureterosigmoidostomy anastomosis left intact [2, 3]. Herein we report a case of sigmoid adenocarcinoma developing in a patient with a remote history of neonatal ureterosigmoidostomy and subsequent revision to ileal loop urinary diversion, who was not undergoing enhanced screening. We present the following case in accordance with the CARE reporting checklist.

Case Presentation

A 72-year-old woman was referred for further evaluation of left lower quadrant abdominal pain, blood per rectum and dark stools. Colonoscopy had been attempted but could not be completed due to sigmoid tortuosity. There was suggestion of a sigmoid mass, but this could not be confirmed endoscopically and biopsies of the area in question revealed necrotic tissue and atypical cells indeterminate for malignancy. The patient's most recent colonoscopy was 7 years prior.

She had a complicated medical history mostly related to her multiple congenital anomalies. Her bladder exstrophy was addressed by ureterosigmoidostomy diversion in the neonatal period that was subsequently revised to ileal conduit at age 21. She had undergone numerous prior abdominal and pelvic operations. Her other comorbidities included right sided aortic arch, coronary artery disease, atrial fibrillation, congestive heart failure, chronic kidney disease and chronic vaginal prolapse. The patient had no personal or family history of colon cancer or inflammatory bowel disease.

The initial step in our evaluation was another attempt to establish a diagnosis with colonoscopy, which confirmed the presence of a partially obstructing mass in the distal sigmoid [Fig 1]. Biopsies revealed adenocarcinoma. Staging revealed no evidence of distant metastatic disease. At operation, the tumor was found to be invading the retroperitoneum, and remnant distal ureter stumps were found in the region of the tumor, still anastomosed to the sigmoid [Fig 2]. Anterior resection of the rectosigmoid with en bloc resection of the retroperitoneum and the remaining ureters attached to the colon, and construction of colorectal anastomosis was performed. Histologic evaluation revealed pT3-4N0 adenocarcinoma with negative margins and 0/30 nodes involved with tumor. The patient had an uncomplicated recovery.

Discussion

The association between urinary diversion with ureterosigmoidostomy and subsequent development of rectosigmoid adenocarcinoma has been described in the medical literature [4, 5]. Prevalence of ureterocolostomy in the population is thought to be approximately 2 in 100,000 people, with post diversion neoplasia occurring in 24% of these patients at a mean of 20 years follow up [3].

Since the first case report by Hammer in 1929, the mechanism of this increased risk of malignancy has remained elusive [6]. There are varying theories regarding the etiology, including exposure of colon mucosa to urine inciting hyperplasia, and alteration in bacterial flora resulting in elevated nitrite compounds or elevated free oxygen species [7, 8]. The tumor in our patient was near the site of original anastomosis with the ureter stump still in place, suggesting that the area was indeed exposed to urine during the 21 years that the ureterosigmoidostomy was functional.

There is usually a latent period between creation of ureterosigmoidostomy and cancer occurrence ranging from 6 to 55 years, with a median of 21-33 years reported in small series [9]. Our patient was 51 years removed from takedown of ureterosigmoidostomy and conversion to ileal conduit and 72 years from the original ureterosigmoidostomy as a neonate, which to our knowledge is the longest latent period reported to date. Due to her history of ureterosigmoidostomy, our patient should have been considered high-risk for rectosigmoid cancer. As recommended by other authors, a reasonable option for screening would be frequent flexible sigmoidoscopy and screening the remainder of the colon based on standard guidelines [2]. Although the number of patients with a history of prior ureterosigmoidostomy is dwindling, clinicians should be aware

of the increased risk of rectosigmoid cancer in these patients and screen them appropriately.

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Figure 1: Ulcerated mass in the sigmoid colon.

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Figure 2: Remnant ureteral stumps from prior ureterosigmoidostomy (encircled with blue vessel loops) attached to sigmoid colon.

Conflict of Interests

All authors have completed the ICMJE uniform disclosure form. The authors have no conflicts of interest to declare

Ethical Statement

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013).



1 Sigmoid Colon : Mass



2 Sigmoid Colon : Mass

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