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Signet ring cell cancer of colon 48 years after ureterosigmoidostomy, treated with low anterior resection and continent urinary diversion

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Background	A male patient presented 48 years following ureterosigmoidostomy with a signet ring cancer at the urinary colonic anastomosis.
Summary	Our patient presented at age 53 with a change in his bowel habits. He had a ureterosigmoidostomy at age five for the diagnosis of benign bladder extrophy. He initially ignored the increased frequency of stools until he had recurrent urinary tract infections. Subsequent studies led to the findings of hydroureteronephrosis and a pelvic mass. Colonoscopic examination demonstrated a tumor at the ureterosigmoid anastomotic site. The patient underwent surgery to resect the mass with a low anterior resection and continent urinary diversion. Final pathology revealed the tumor to be a poorly differentiated signet ring carcinoma. While the increased incidence of adenocarcinoma at the urinary colonic anastomosis is well documented, signet ring cancer is reported less often. Generally, it is seen at an earlier age and has a worse prognosis. Peritoneal metastases are more common than hepatic spread. The need for lifelong surveillance is underscored by this case report.
Conclusion	Ureterosigmoidoscopy has a high incidence of adenocarcinoma. We present a case of signet ring carcinoma presenting 48 years following the patient's urinary tract surgery. It highlights the need for continued surveillance regardless of the lengthy time interval.
Keywords	Ureterosigmoidostomy, signet ring carcinoma

Case Description

Tumor development within the colon following ureterosigmoidostomy is a rare but well-documented event.[1, 2, 3] Multiple etiologic factors contribute to the increased incidence of colon cancer following urinary diversion, including increased exposure of the colonic epithelium to urine flow and activation of carcinogenic compounds. These tumors have a variable latency period but most commonly present within two decades, usually as an adenocarcinoma. We reported a rare presentation of signet ring cell cancer at the urointestinal anastomosis 48 years after urinary diversion for bladder exstrophy.

The patient is a 53-year-old male with history of benign bladder exstrophy diagnosed and treated with ureterosigmoidostomy at the age of five. The patient reported a history of frequent bowel movements (approximately 15 per day) but never sought the attention of a physician. The patient subsequently developed a persistent urinary tract infection which prompted a referral to our facility. Ultrasound and CT scan revealed left hydroureteronephrosis as well as a suspicious mass within the pelvis (figure 1).

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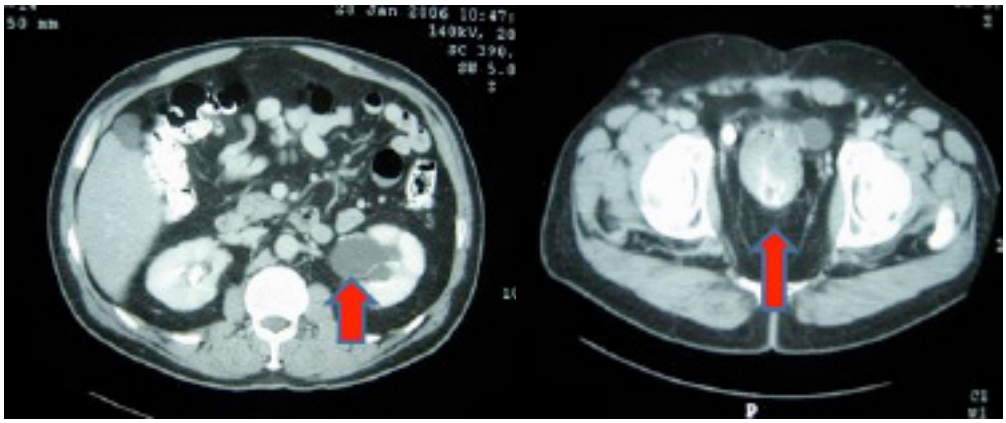

Figure 1. Left hydroureteronephrosis and suspicious mass within the pelvis

Given the patient's prior surgical history, he was referred to a colorectal surgeon for colonoscopy and further workup.

Colonoscopy revealed a suspicious mass at one of the ureteral orifices in the distal rectum (figure 2).

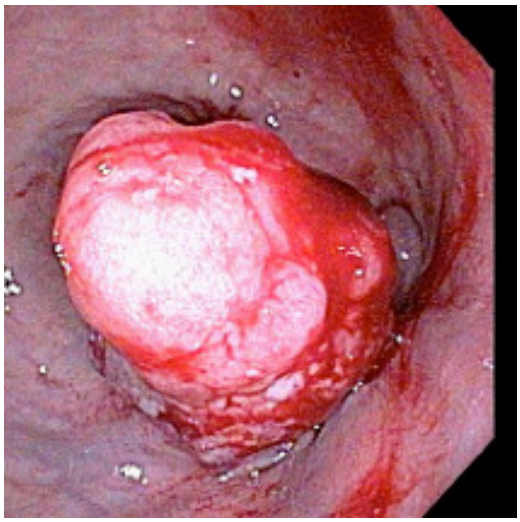

Figure 2. Suspicious mass at one of the ureteral orifices in the distal rectum

The contralateral ureteral orifice was grossly free of any mass. Plans were then made to resect the mass. The risks of cancer, particularly with urinary-colonic diversion, were thoroughly explained to the patient, who was adamant on having a continent urinary diversion. The patient agreed to careful follow-up, with an understanding about recurrence risks. CT scan of the abdomen and pelvis revealed no evidence of metastatic disease.

The patient was taken to the operating room and a low anterior resection was performed along with a continent catheterizable ileocolonic stoma (Indiana pouch). The distal ureteral margins were free of tumor. Final pathology of the left ureter and associated colon revealed high-grade poorly differentiated signet ring cell cancer (figure 3).

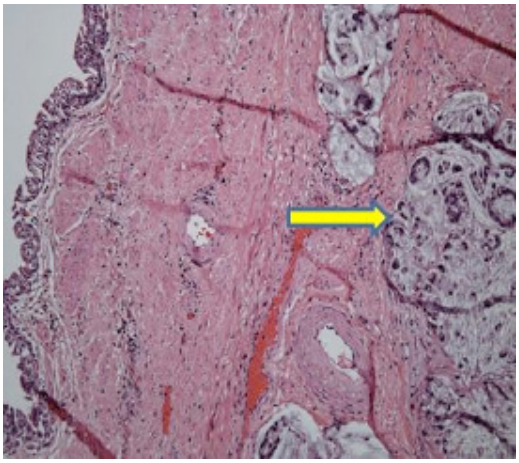

Figure 3. High-grade poorly differentiated signet ring cell cancer

A colonoscopy one year after surgery revealed no evidence of tumor and the patient's Indiana pouch continues to function well.

Discussion

Ureterosigmoidostomy is an acceptable but outdated means of continent urinary diversion, first described in 1852 .4 Ureterosigmoidostomy is a technically straightforward operation with relatively good results for benign conditions requiring cystectomy (such as bladder exstrophy), but this procedure has fallen out of favor over the past half-century for numerous reasons. The advantages of ureterosigmoidostomy are that it avoids the need for an abdominal stoma or any catheterization, it has an excellent continence mechanism, and it avoids the need for complicated reconstructive surgery. Specific problems include recurrent urinary tract infections and ascending pyelonephritis (secondary to colonic bacteria refluxing up the ureter), nephrolithiasis, ureteral stricture, anal incontinence and diarrhea with resultant metabolic abnormalities, and most importantly, an increased incidence of colonic carcinoma at the ureteral anastomosis.[1,5]

The incidence of colon carcinoma at the site of the ureteral implant ranges from 100 to 7,000 times that of the general population.[6,7] The etiology of carcinogenesis is not fully delineated, but it is probably related to the persistent flow of urine across the colonic epithelium. The mixture of urine and feces at the suture line of the uroepithelium and colonic mucosa results in bacterial activation of carcinogenic N-nitroso compounds. Persistent postsurgical inflammatory responses may also promote carcinogenesis at the anastomotic line.[8,9] The latency period between surgery and cancer onset ranges from 6 to 50 years, with a mean time of 21 years and a median age at diagnosis of 33.6

Signet ring cell cancer is characterized by mucin-secreting adenocarcinoma cells that contain intracytoplasmic mucin and peripherally placed nuclei. There is some diagnostic overlap between signet ring cell cancer and mucinous colorectal cancer. In western countries, the incidence is quite low, ranging from 0.8 to 2.6%, but in eastern nations such as Jordan and Lebanon, there is a much higher incidence of both mucinous and signet ring cell carcinomas (as high as 18.5%). The most common location of primary signet ring cell carcinoma is the stomach (>96%), followed less frequently by the breast, gallbladder, and urinary bladder. Rarely, colorectal signet ring cell cancers are found (0.1–2.6% of all colorectal cases). The majority of colorectal signet ring cancers are located in the right colon and rectum.[10-12]

There are no clear studies identifying an association between colon polyps and adenomas and signet ring cell colon cancer, but there have been some findings in the literature suggesting an association between inflammatory bowel disease and signet ring cell cancer.[11,12] Unlike adenocarcinoma of the colon, positive family history is not an established risk factor for signet ring cell colon cancer. The mean age of patients with signet ring cell cancer is lower (23–57) than that of patients with non-signet ring colon cancer. Presenting symptoms include abdominal pain, change of bowel habits, and rectal bleeding.[10, 11]

In general, signet ring cell carcinomas have a worse prognosis compared to other primary colorectal cancers. They frequently are diagnosed at an advanced tumor stage and have higher rates of peritoneal seeding, resulting in lower rates of curative resection. A characteristic feature of signet ring colon cancer is the high incidence of peritoneal metastastes and relatively low incidence of hepatic metastases compared to nonsignet ring colon cancer. Distant metastasis appears to result from tumor cells spreading in a continuous manner via a more aggressive type of infiltrating growth.[10-12] A recent study noted that only 71% of patients diagnosed with signet ring colon cancer were treatable with surgery and only half of those patients achieved curative resection—much lower than in non-signet ring cell colon cancer.[10] Neoadjuvant and adjuvant regimens are utilized frequently, but prognosis and survival remain poor. Reported overall five-year survival rates range from 9% to 50%, with median survival times ranging from 15 months to 24 months.[11]

Our case underscores the importance of close follow-up after ureterosigmoidostomy. Late detection of colon cancer after urinary diversion leads to a high mortality rate, so lifelong surveillance is recommended for all patients who undergo ureterosigmoidostomy. Annual surveillance with colonoscopy and fecal occult blood testing should be started soon after surgery, and conversion to an alternative means of urinary diversion should be made if recurrent polyps, dysplasia, or cancer is found.1 Our patient is unique because of his longer than average latency period (48 years) between his surgery and his cancer diagnosis at 53; in addition, the patient was found to have a relatively rare signet cell colonic carcinoma, treated successfully with low anterior resection and Indiana pouch continent urinary diversion.

Conclusion

Ureterosigmoidoscopy has a high incidence of adenocarcinoma. We present a case of signet ring carcinoma presenting 48 years following the patients urinary tract surgery. It highlights the need for continued surveillance regardless of the lengthy time interval.

Lessons Learned

Urinary diversion into the colon increases the risk of cancer. Rare cancers like signet ring cancer are possible and have a worse prognosis. Surveillance for the patient's lifetime is critical; our patient presented 48 years following urinary diversion.

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