Oncology

Adenocarcinoma in a Koff Urinary Ileal Diversion

Bradley Sherman a,*, Frederick Taylor b

a Department of Surgery, Doctor’s Hospital, Columbus, OH, USA
b Grant Medical Center, Central Ohio Urology Group, USA

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ABSTRACT

The use of an ileal conduit as a means of treatment for bladder cancer or dysfunction is widely used and understood. However, long term surveillance of that conduit has not been strongly established and set forth as a means of screening. We present a 76yo female with a history of neurogenic bladder secondary to paraplegia who underwent the formation of a “Koff” pouch as a conduit. Nineteen years later she presents with hematuria and was found to have adenocarcinoma originating in her conduit.

Introduction

This is a case of a 76 year old female with a 19 year history of an ileal pouch placed secondary to neurogenic bladder with resultant severe lower tract dysfunction as a result of becoming a paraplegic. She presented with hematuria and was found to have primary adenocarcinoma at the site of her ileal pouch with metastasis to her liver. It is well established that the construction of urinary diversion due to malignancy is used quite frequently. However, due to the majority of these diagnoses being performed in the late stage of life little to no surveillance is performed for these diversions. As a result there is no developed standard of care regarding surveillance for ileal pouches. This is a case report of an individual and a review of the literature of similar cases so that an estimate for possible surveillance can be established.

Case

76-year-old female, who has a longstanding continent catheterizable urinary diversion via a Koff pouch that was performed for lower urinary tract dysfunction subsequent to paraplegia 19 years prior to her presentation to the hospital. She had been in her normal state of health until she developed gross hematuria noted on catheterization. Physical examination showed a well nourished elderly woman who was high functioning despite her age and paraplegia. Past history was not contributory. She denied a smoking history. Denies ever having a colonoscopy or regular cancer screening. Workup of her gross hematuria included a CT scan of the abdomen and pelvis which demonstrated retroperitoneal lymphadenopathy, calcification of the urinary conduit, and multiple hepatic lesions, one of which was biopsied and found to be adenocarcinoma favoring intestinal primary (Fig. 1). Urine cytology did demonstrate atypical cells. A cystoscopy was undertaken and upon entering her ileal conduit she had multiple fibrinous, exophytic, calcified, areas that were suggestive of malignancy (Fig. 2). These were biopsied and demonstrated invasive, poorly differentiated adenocarcinoma.

Discussion

Patients undergo ileal pouch formation for a number of reasons, chronic inflammatory disease, neurogenic sources, detrusor over...
activity, infestation by parasites, bladder extrophy, and most commonly cancer. It is estimated that greater than 10,000 individuals a year undergo cystectomy with a conventional ileal conduit, 2000 of these undergo continent urinary diversion. The average age of those diagnosed with bladder cancer is 73 and with average life expectancy in the USA of 78.8 years. As a result these cases are likely underreported as there is not adequate time for this specific presentation to manifest. Greater awareness of both the patient and practitioner is important to limit the potential morbidity and mortality this may cause.

The typical time of transformation of intestinal cells to adenomas to early cancer is likely less than 5–10 years as evidenced by current established literature of colonic primaries. It is reasonable to propose that chronic exposure to urine can result in a dysplasia of the surrounding intestinal mucosa. The exact mechanism is not completely understood. Theories consist of chronic inflammation and irritation that contribute to the carcinogenesis within the intestinal segment. This is further supported by observable histological signs after long exposure to urine demonstrating inflammatory cells.

On a review of the literature there are several other cases of ileal diversion that subsequently develop primary intestinal malignancy. In 1978 by Shousha et al who described a patient with bladder extrophy that underwent urinary diversion. Twenty years later he developed metastatic disease to the liver. Two similar patients that underwent cystectomy secondary to malignancy developed carcinoma of their Indiana pouch, that required surgical removal 10 years following initial surgery. A review of Indiana pouch creation and detection of tumor was 2–15 years, with an average of 8–9 years development of appreciable malignant cells. Given the different techniques for the construction of an ileal conduit it is challenging to determine whether the formation of malignant cells originate in the colonic or small bowel mucosa. Based on the work of Austen et al a review of ileocystoplasties demonstrated the time to formation of malignancy was variable, 2–40 years, with a median of 22 years. However, examining colocystoplasties although not as common demonstrated a median of 17 years. A review of the literature does not demonstrate that strict ileal diversion, such as a Kock pouch would result in similar malignancy rates. In a study preformed in 2002, 645 patients that only used ileum for urinary diversion demonstrated tumor development in 0.3% of 350 ileal conduit, 0.8% of 260 ileal ureteral replacements, and 5.5% of 55 ileocystoplasties. In ileal ureteral replacement these cases were all preformed due to either tuberculosis or schistosomiasis and as a result the exposure of these pathogens may have increased the likelihood of malignancy. It is challenging to come to a solid conclusion in regards to colonic or isolated ileum tumor incidence. Furthermore, many of the cases are not able to differentiate the location of the tumor in a conduit. Making the determination of colonic vs ileal tumor genesis difficult. Regardless, a large database would aid in tracking tumor formation rates.

**Conclusion**

There are no current strict guidelines for routine screening for ileal conduits. An assumption could be made that some measure of flexibility for screening could be made for those that underwent ileal diversion as a result of malignancy vs poor function. Although as referenced prior a history tuberculosis or schistosomiasis increased the likelihood of malignancy. Also in cases of bladder extrophy the median time of malignant development was similar to those who had an initial malignancy. It may also be important to determine the anatomical components of the diversion as this may affect the malignant transformation potential of the conduit. Currently proposed guidelines presented in the literature recommend yearly follow up beginning with postoperative year 3–5. This should include endoscopic evaluation of the conduit or pouch with biopsies of suspicious sites. Urine cytology yearly could also be used for those preferring not to undergo cystoscopy although the sensitivity of this has not been studied.

**Conflict of interest**

There is no conflict of interest.

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