Adenocarcinoma in an Ileal Pouch Occurring 14 Years After Restorative Proctocolectomy

Report of a Case

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Restorative proctocolectomy (RP) is the operative procedure of choice for patients with severe ulcerative colitis refractory to medical therapy and for colitics with associated cancer or precancerous dysplasia.1–3 There have been many refinements in the operative technique and postoperative management during the past 15 years, with 92 percent of patients having a good long-term outcome.4 Adenocarcinoma arising at the site of ileoanal anastomosis or within an ileoanal reservoir is exceedingly uncommon and more often associated with familial adenomatous polyposis (FAP)5,6 than with ulcerative colitis.7–10 We report a rare case of adenocarcinoma arising in an ileal J-pouch reservoir 14 years after RP for ulcerative colitis.

REPORT OF A CASE

The patient was a 63-year-old male with a history of ulcerative colitis, diagnosed at age 19, who presented with unexplained pelvic pain. At the age of 49, because of chronic disease activity and inability to taper high-dose corticosteroids, the patient underwent restorative proctocolectomy by the senior author in 1986. This operation consisted of abdominal colectomy, proctectomy with endorectal mucosal resection, formation of a J-pouch ileoanal reservoir, and circumferential handsewn anastomosis of the distal pouch to the dentate line within a denuded muscular cuff. The pathology revealed pancolitis, focal dysplasia, and an 8-mm area of superficially invasive cancer (T1, N0) arising in the ascending colon. A proximal diverting loop ileostomy was constructed, which was closed 12 weeks later. This patient was evaluated every three months for the first year and then every six months for the second year, yet failed to maintain his scheduled yearly visits thereafter. The patient underwent yearly pouchoscopy with no documented episodes of pouchitis, and no biopsies were taken during the first two years of follow-up. The patient experienced a stable functional result for approximately 13 years after the procedure and then presented to an outside hospital with constant, low-grade pelvic pain. He denied any fever, visible bleeding per anum, or change in bowel or bladder function. The pain was characterized as dull and localized to the low presacral area. CT scan of the abdomen and pelvis was unremarkable. Magnetic resonance imaging of the pelvis showed a nonspecific area of enhancement along the interface of the posterior pouch and the sacrum. An examination under anesthesia at the outside hospital, including random pouch biopsies, revealed no unusual palpable, endoscopic, or histologic findings. Shortly thereafter, the pain worsened, and was associated with pouch filling and emptying. After returning to our institution and extensive discussion regarding the unremitting symptoms, the patient consented to an abdominoperineal resection of the ileoanal pouch and construction of a permanent ileostomy.

At surgery, abdominal exploration was unremarkable. The upper half of the ileoanal pouch was mobile
and relatively devoid of adhesions or fibrosis. The lower portion of the pouch, however, was densely adherent to the distal sacrum and coccyx. There were no other gross findings suspicious for malignancy. The ileal pouch, sphincter, and anal canal were removed and a Brooke ileostomy fashioned.

Pathologic inspection of the pouch revealed a 5-cm mucosal irregularity corresponding to an area of mural thickening (Fig. 1). Histopathology revealed Grade 2 adenocarcinoma arising from ileal mucosa, well proximal to the original anal anastomosis, with transmural invasion posteriorly (Fig. 2). The tumor measured 2.7 cm × 1.5 cm × 1.2 cm, with a deep posterior margin positive for malignant involvement. Lymph nodes were free of metastasis. Postoperatively, the patient underwent combined modality chemotherapy and radiation to the pelvis and greatly improved, but was still not completely asymptomatic. A distal sacrectomy had been recommended, but the patient declined.

**DISCUSSION**

With the rapid increase in the frequency of RP in the 1980s, there was speculation of the risk of malignant degeneration either in the reservoir itself or at the ileoanal interface.10 Yet, a review of more than 1,000 patients with restorative proctocolectomy revealed no cases of malignancy near the ileoanal pouch.11 Isolated reports of malignancy after RP appeared in the 1990s.8,9 Several of these reports were in patients with FAP.5,6 Adenomatous polyps in both continent ileostomy reservoirs and ileoanal reservoirs have also been found after proctocolectomy for FAP.12,13

Malignant transformation has been seen at the site of a Brooke ileostomy near the mucocutaneous junction.14 Pouch malignancy after RP for ulcerative colitis7,15 has been attributed to “backwash ileitis” as well as residual rectal mucosa.6 Indeed, islands of residual or regenerated “colonic” mucosa between the serosal surface of the reservoir and the opposed rectal muscular cuff10 are a source of potential malignant degeneration. The ileal mucosa in the ileoanal reservoir undergoing colonic metaplasia and even dysplasia is another possible source.17–19 Histologic studies of ileoanal reservoir biopsies have found colonic metaplasia in pouch mucosa characterized by goblet cell multiplication, persistent severe villous atrophy, and formation of crypts.16,19 In patients with chronic “pouchitis,” dysplasia has been found in up to 71 percent.17

A final mechanism for malignant transformation is specific to the use of a stapled anastomosis.20 Before 1991, our standard operative approach included a transanal endorectal mucosal resection that commenced at the dentate line and extended approximately 2 to 4 cm, ending just above the upper border of the anal sphincter apparatus. The denuded rectal muscularis propria was then transected. The muscular “cuff” remaining consisted of the internal sphincter and 1 to 2 cm of rectal muscularis propria. The ileoanal reservoir, typically a J-pouch configuration, was then pulled through the muscular cuff and handsewn to the anoderm and internal sphincter at the level of the dentate line. Since 1991, our distal extent of the proctectomy extends full-thickness into the anal canal, resulting in a transection of mucosa and internal sphincter at the dentate line or slightly above, in the area of the anal transitional zone (ATZ). No mucosal

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**Figure 1.** Gross pathology revealed a 5-cm raised mucosal lesion with an area of mural thickening (arrow).

**Figure 2.** Microscopic evaluation of the ileoanal pouch with hematoxylin and eosin stain revealed an adenocarcinoma, Grade 2, with transmural invasion extending into adipose tissue.