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Restorative proctocolectomy for familial adenomatous polyposis coexisting with colorectal cancer

Kolorektal kanser gelişmiş familial adenomatöz polipozislerde restoratif proktokolektomi

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The aim of this study was to assess whether restorative proctocolectomy was suitable as an initial procedure for selected familial adenomatous polyposis patients with coexisting colorectal cancer. Six malignancy patients who underwent restorative proctocolectomy for familial adenomatous polyposis were reviewed. At the time of restorative proctocolectomy, cancer was not suspected in four patients. The two remaining familial adenomatous polyposis patients had a known associated colorectal cancer. Operative procedures, pathological findings, complications and long-term follow-up were evaluated. All patients were Stage I-II cancers. There was no mortality or pouch failure. No evidence of tumor recurrence was found and all the patients are still alive and disease-free in follow-up (mean 28 months). As an initial procedure, restorative proctocolectomy for patients with familial adenomatous polyposis with coexisting Stage I-II colorectal cancer seems to be a dependable procedure.

Key words: Colorectal cancer, familial adenomatous polyposis, restorative proctocolectomy

Bu çalışmanın amacı, kolorektal kanser gelişmiş bazı hastalarda ilk seçenek olarak restoratif proktokolektominin yapılıp yapılamayacağını araştırmaktır. Restoratif proktokolektomi yapılan familial adenomatöz polipozisli altı kolorektal kanser hastası retrospektif olarak incelendi. Restoratif proktokolektomi yapılırken dört hastada kanser teşhis edilememişti. Kalan iki hastada ise ameliyat öncesi kanser tanısı bilinmekteydi. Ameliyat bulguları, patolojik değerlendirmeler, komplikasyonlar ve uzun dönem takipler incelendi. Tüm hastalar Evre I-II kanserlerden oluşuyordu. Hiçbir hastada mortalite ve poş kaybı olmadı. Hiçbir hastada tumor nüksü gözlenmedi ve tüm hastalar halne hastalıksız olarak hayattadırlar (ortalama 28 ay). Evre I-II kolorektal kanserlerle birlikte olan familial adenomatöz polipozis hastalarında ilk seçenek olarak restoratif proktokolektomi uygulanması mantıklı görünmektedir.

Anahtar kelimeler: Kolorektal kanser, familial adenomatöz polipozis, restoratif proktokolektomi

INTRODUCTION

Familial adenomatous polyposis (FAP) is an autosomal-dominant inherited premalignant disease in which the risk of adenocarcinoma of the colon and rectum increases with age. Although screening programs of family members and prophylactic surgery have provided a significant decrease in the incidence of colorectal cancer (1), some FAP patients still present with colorectal cancer at the time of FAP diagnosis. The stage and the location of the cancer are the main determinants of the type of surgical procedure; surgical alternatives are total proctocolectomy with end ileostomy, total colectomy with ileorectal anastomosis and restorative proctocolectomy (1, 2). Although restorative proctocolectomy has some advantages over the other two procedures and is usually preferred as the first surgical option in FAP patients without cancer (3), results are still conflicting for the cases coexisting with cancer. The aim of this study was to assess whether restorative proctocolectomy was suitable as an initial procedure for selected FAP patients with coexisting colorectal cancer.

MATERIALS AND METHODS

From January 1992 to January 2001, 36 patients underwent surgery for FAP. The diagnosis of FAP was confirmed by endoscopic examination, radiological contrast studies and biopsies. Preoperative diagnosis of malignancy was made in 17 (47%) patients: seven required proctocolectomy with end

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ileostomy for rectal cancers, eight required total colectomy with ileorectal anastomosis and two underwent restorative proctocolectomy. In 19 (53%) cases, there was no preoperatively noticed coexisting colorectal cancer and all were treated by restorative proctocolectomy. Four (21%) of these patients were diagnosed as malignancy after histopathological examinations of the excised specimens. These six malignancy patients in total in whom restorative proctocolectomy for FAP was performed were retrospectively reviewed. Operative procedures, pathological findings, complications and long-term follow-up were evaluated.

The operative procedure consisted of total abdominal colectomy and mobilization of the rectum. The distal rectal mucosa was excised to the dentate line via perianal approach and a manual ileal J pouch anastomosis was constructed in two patients. No mucosectomy was performed; double stapling technique was performed in four patients. The choice of the surgical technique was left to the surgeons. Four patients had temporary diverting loop ileostomies, which were closed two months later after pouchograms. Preoperatively known cancers were staged by abdominal ultrasound, computed tomography and surgical findings. Pathological staging was based on the TNM classification (4). No patients received postoperative chemoradiotherapy. Cancer follow-up included periodical physical and endoscopic examinations, chest X-ray, carcinoembryonic antigen level, blood biochemistry and abdominal ultrasound.

RESULTS

The six patients who underwent restorative proctocolectomy for FAP associated with colorectal cancer are outlined in (Table 1). Preoperatively unknown cancers were small and Stage I-II cancers. One of the known cancers was a non-palpable tumor and the other was not observed to have penetrated the serosa during the surgical exploration. No enlarged lymph nodes were determined during surgical explorations. Histopathological examinations of the resected specimens did not demonstrate extensive cancers (Stage I-II). Cancers of the rectum were at least 8 cm above the anal verge.

Postoperative complications were documented in two patients (33%). One patient without a diverting ileostomy complicated with anastomotic leakage and was treated by temporary ileostomy. The other patient had pulmonary embolism and was treated medically. All the temporary ileostomies were closed. There was no mortality or pouch failure. No evidence of tumor recurrence was found and all the patients are still alive and disease-free in the follow-up (mean 28 months).

DISCUSSION

Because FAP carries a serious risk of malignancy, preoperative diagnosis of cancer should be essential. But preoperative cancer diagnosis may be a problem, due to the progressive formation of hundreds, even thousands, of colorectal adenomas. We preoperatively diagnosed 79% of the malignancies of FAP cases and missed only 21% of the cases. Other studies reported higher rates of undetected malignancy cases for FAP (28%-50%) (5, 6). None of our incidentally found malignancies were advanced cancers. These findings were supported by other authors, and, as far as we know, there was no FAP case in whom an advanced colorectal cancer was missed (3, 5, 6). This is quite different from ulcerative colitis in which incidentally found coexisting advanced colorectal cancers are not uncommon (7). The difference may originate from the structure of the colorectal wall. FAP has a pliable, soft intestinal wall, and detection of an advanced cancer during endoscopic and surgical examination is much easier than in ulcerative colitis, which is characterized by thick, inflamed intestinal wall and mucosa.

Several operations have been used to treat patients with FAP. In total proctocolectomy with end ileostomy, the disease is cured because all of the mucosa of the large intestine is eliminated. This operation, however, requires a permanent ileos-

Table 1. Demographics and the results of the patients

No	Age/Sex	Location	Diagnosis	TNM	Follow-up	Result
1	44/F	Colon	Preoperative	T3,N0,M0	63 months	Disease free
2	56/M	Rectum	Postoperative	T1,N0,M0	57 months	Disease free
3	45/F	Rectum	Preoperative	T1,N0,M0	23 months	Disease free
4	52/M	Rectum	Postoperative	T3,N0,M0	7 months	Disease free
5	28/F	Colon	Postoperative	T1,N0.M0	16 months	Disease free
6	36/M	Colon	Postoperative	T1,N0,M0	10 months	Disease free

tomy and it is an important issue in life quality of patients who are mainly young and active. Until the advent of restorative proctocolectomy, the mainstay of surgical therapy for patients with FAP was total colectomy and ileorectal anastomosis that eliminates most of the large intestinal mucosa (1, 6). Madden and coworkers suggested that ileorectal anastomosis should be preferred as the first surgical option in most FAP patients and that restorative proctocolectomy should be restricted to some select patients (2). We do not support this approach because of the high incidence of reported rectal cancer risk ranging from 7.1% to 32.1% after ileorectal anastomosis and of the reported poor outcomes in the cases of rectal cancer developing after ileorectal anastomosis (1). The most recent studies advocate restorative proctocolectomy as the initial operation of choice for most FAP patients (3, 8, 9). It eliminates all large intestinal mucosa, avoids permanent stoma and, when compared with ileorectal anastomosis, has a minimal risk for rectal cancer and provides an acceptable life quality.

This study showed that restorative proctocolectomy was not only a preventive procedure at FAP, but also a therapeutic modality with the patients who had an existing early stage colorectal cancer. Despite the favorable results, we believe that Stage III-IV colorectal cancer patients and those with distal rectal cancers are not candidates for restorative proctocolectomy.

The best timing for restorative proctocolectomy for colorectal cancer complicated by FAP remains a matter of debate. Taylor and associates suggested that all patients with carcinoma who are candidates for restorative proctocolectomy should have it as their initial procedure (10). They reported the results of four patients with FAP and of 13 with ulcerative colitis complicated by cancer; 45% of the patients had Stage C disease. Metastatic disease developed in only one patient nine months later. Stelzner and Fonkalsrud reported 12 cancer patients with complicating ulcerative colitis and FAP-three Stage C and two Stage D (11). Two patients in their series with Stage C cancer died of metastatic disease after 18 and 43 months, respectively, and both patients with Stage D were followed up for only two months postoperatively. Ziv and coworkers reported eight FAP patients with coexisting colorectal cancer treated with restorative proctocolectomy (6). Only one patient had Stage III disease and this patient developed metastatic disease after surgery. Contrary to these studies, Wiltz and associates refused to perform immediate restorative proctocolectomy on patients with a proven or suspected cancer. For all such cases, they recommended total abdominal colectomy combined with Hartmann's resection with end ileostomy. After accurate staging, adjuvant therapy was offered to patients who would benefit, and after a period of observation (12-24 months), restorative proctocolectomy was performed (5). We accept neither routine restorative proctocolectomy nor routine total colectomy and Hartmann's procedure. Instead, we recommend restorative proctocolectomy selectively to Stage I-II cancers as an initial procedure. Other cases should be treated by other surgical options with the combination of adjuvant chemoradiotherapy.

CONCLUSIONS

Restorative proctocolectomy for patients with FAP coexisting with Stage I-II colorectal cancer seems to be a dependable procedure. These cases have favorable prognosis. Theoretically, restorative proctocolectomy should provide the treatment of the cancer, elimination of the large intestinal mucosa without permanent ileostomy, and a good disease-free long-term survival. This study shows that restorative proctocolectomy can achieve these results for FAP patients with Stage I-II colorectal cancers.

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