Case Analysis

Surgical Management of Adenomatous and Juvenile Polyposis Coli – A Single Surgeon's Experience

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Adenomatous;
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Subtotal colectomy;
Restoratvie proctocolectomy

Purpose. Early operation is recommended for patients with polyposis coli because cancer will develop in 100% of patients if untreated. This is a retrospective analysis of a single surgeon's experience of surgical management of polyposis coli in a period of 25 years.
Materials and Methods. From 1984 to 2010, 46 cases of polyposis coli were operated by a single colorectal surgeon (TCH). There were 44 patients with adenomatous polyposis coli and two patients with juvenile polyposis coli. Twenty-three patients were male, and 23 patients were female. Age ranged from 11 to 58 years old, with an average of 34.2 years old. Three patients had colectomies for polyposis coli previously.
Results. Restorative proctocolectomy (IPAA) was performed in 22 patients, followed by total proctocolectomy with ileostomy in nine patients and subtotal colectomy with ileorectal anastomosis in five patients. Thirty-two patients were found to have invasive adenocarcinoma in the specimen. Four patients were re-explored postoperatively for postoperative complications. Thirteen patients died of metastatic colorectal cancer and four patients died of cause other than cancer. Within the group of patients who had IPAA, a patient did not have the ileostomy closed, a patient died of metastatic cancer, and a patient was a non cancer death years later. There were 19 patients with good function of pouch. One patient is currently receiving treatment of desmoid tumor.
Conclusion. Colectomy should be recommended to all patients with polyposis coli to avoid death due to colorectal carcinoma. Restorative proctocolectomy is not a perfect operation, but can offer a reasonable good quality of life to the patient with polyposis coli if the operation is successful.
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Polyposis coli is defined as having at least 100 polyps present in the colon. Familial adenomatous polyposis coli is characterized by the development of hundreds to thousands of adenomatous polyps in the colon and rectum of affected individuals. Classic FAP is inherited in an autosomal dominant manner and results from a germline mutation in the adenomatous polyposis (APC) gene, on chromosome 5q21. Although it is a hereditary disease in most of cases, there are 20-30% of patients without family history, and is

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caused by gene mutation. Associated manifestations include gastrointestinal tract polyps, congenital hypertrophy of the retinal pigment epithelium, desmoid tumors, and other extracolonic malignancies. Gardner syndrome is more of a historical subdivision of FAP, characterized by ostemas, dental anomalies, epidermal cysts, and soft tissue tumors. Other specified variations include Turcot syndrome with associated central nervous system malignancies and hereditary desmoid disease.

FAP has an incidence of about one in 7,000 to 24,000 live birth.[1,2] It manifests equally in both sexes, and accounts for less than 1% of colorectal cancer cases. In 2009, the European medicines Agency (EMEA estimated that FAP affected approximately 3-10/100,000 people in European Union.[3]

Unless the colon is removed early in life, most of patients will develop cancer before 40 years old.[4,5] Early operation is recommended for patients with polyposis coli because cancer will develop in 100% of patients if untreated.

Juvenile polyposis coli is a rare disease and frequently seen in the families of adenomatous coli. Most of experts believe that the juvenile polyposis may eventually become adenomatous polyposis, so the disease should be considered as high in malignant potential. Early operation is suggested for the patients with juvenile polyposis coli.[6]

Because of low incidence of the disease in our country, experiences of most physicians of management especially surgical management of the patients are limited. This is a retrospective analysis of a single surgeon's experience of indications, procedures and results of surgical management of polyposis coli in a period of 25 years.

Materials and Methods

The study used the Mackay Memorial Hospital (MMH) administrative database of colorectal surgery and we identified all patients who had surgical intervention for FAP. From February 1984 to December 2010, 46 cases of polyposis coli were operated by a single colorectal surgeon (TCH). Excluded were the patients with hyperplastic polyposis or Peutz-Jegher's polyposis, patients who refused surgery, patients who were operated by other surgeons. Demographic data including age and gender, surgical indications, type of first operation at MMH, morbidities, and mortalities were analyzed.

Results

There were 44 patients with adenomatous polyposis coli and two patients with juvenile polyposis coli. Twenty-three patients were male, and 23 patients were female. Age ranged from 11 to 58 years old, with an average of 34.2 ± 11.7 year old. Three patients had colectomies for polyposis coli in other hospital previously.

The most common chief complaints was bloody stool in 22 patients (47.8%), diarrhea in 12 patients (26.08%), and abdominal pain in 9 patients (19.6%). The most common surgical indication was malignancy in 32 patients, followed by polyposis coli in 14 patients. The stage of colorectal cancer is listed in Table 1.

Restorative proctocolectomy (IPAA) with ileostomy was the operative procedure for 22 patients, subtotal colectomy and ileorectal anastomosis was performed in five patients, and total proctocolectomy with ileostomy was the procedure in nine patients. The rest of procedures included abdominoperineal resection in three patients, subtotal colectomy with ileostomy in two patients, segmental colectomy in three patients, Hartmann's resection in one and colostomy in one patient. (Table 2)

The complications after operation included intestinal obstruction in eight patients, upper gastrointestinal bleeding in three patients, and ileoanal anastomosis dehiscence in two patients.

Thirty-two patients were found to have invasive adenocarcinoma in the specimen. Three patients had two colon resection by the same surgeon because of uncertainly of diagnosis prior to the first operation. A 51 years old female who initially had APR followed by total colectomy with an ileostomy, a 53 years old male who initially had APR followed by total colectomy with an ileostomy, and a 33 years old male who initially had subtotal colectomy followed by restorative proctocolectomy. Four patients were re-

Table 1. Stage of colorectal cancer (32 patients)

	Male	Female
Colon cancer		
Stage I	2	1
Stage II	0	1
Stage III	2	1
Stage IV	0	3
Rectal cancer		
Stage I	1	1
Stage II	4	3
Stage III	4	6
Stage IV	0	1
Colon and rectal cancer		
Stage II	0	1
Stage IV	1	0

Table 2. Operative procedure in patients with polyposis coli

	Male	Female	Total
Restorative proctocolectomy	11	11	22
Total proctocolectomy with ileostomy	5	4	9
Abdominoperineal resection	2	1	3
Subtotal colectomy with ileoproctostomy	3	2	5
Subtotal colectomy with ileostomy	1	1	2
Segmental colectomy	0	1	3
Hartmann's resection	1	2	3
Colostomy	0	1	1

explored postoperatively for postoperative complications: two patients with intestinal obstruction following IPAA, a patient with bleeding following IPAA and a patient with intestinal obstruction following closure of ileostomy. A patient had a segmental colectomy died of postoperative bleeding. Thirteen patients died of metastatic colorectal cancer and four patients died of cause other than cancer. Within the group of patients who had IPAA, a patient did not have the ileostomy closed, a patient died of metastatic cancer, and a patient was a non cancer death years later. There were 19 patients with good function of pouch. One patient is currently receiving treatment of desmoid tumor.

The mean duration of fisrt admission is 19.3 ± 10.9 days. The mean duration of remaining life in patients who died of CRC was 976 ± 1427 days. The mean age of expired patients was 43.0 ± 11.1 years old, with male at 42.6 ± 16.0 years old and female at 43.2 ± 8.8 years old.

Discussion

Some individual case reports of patients with multiple polyps were described in the eighteenth and nineteenth centuries, but Cripps is generally credited with being the first to note the condition in two members of the same family in 1882.[7] He recognized a familial tendency of polyposis coli. Handford described the association of cancer in 1890.[8] Lockhart-Mummery in 1925[9] and Dukes in 1930[10] brought out the malignant potential of the disease. Others had also reported that the disease was premaligant. Besides intestinal manifestation, it was well known that adenomatous polyposis coli has associated extraintestinal manifestation, such as osteoma and desmoid tumor in Gardner's syndrome, malignant brain tumor in Turcot's syndrome, hypoproteinemia and alopecia in Cronkhite-Canada syndrome, and association of congenital hypertrophy of the retinal pigment epithelium etc.[11] The average age of appearance of symptoms was 20 years old, cancer usually develops at approximately 35 years old, and average age of death of the patients with cancer was 41 years old.[12] Following discovery of the disease, it was soon recognized that the disease should be treated surgically before it became malignant.[13]

In the Polyposis Registry of Japan, The mean age of diagnosis of FAP in patient with CRC was 28 year old, compared to 33 year old for those with early cancer (in situ or submucosal) and 40 years old for advanced cancer. The cumulative risk of CRC exceeded 50% by the age of 42 year old in women, and 44 years old in men.[14] In our study, the mean age of diagnosis of FAP in patients without colorectal cancer was 23.8 ± 10.2 year (27.5 ± 10.7 years old in male patients, 20 ± 8.9 years old in female patients), compared to 38.2 ± 9.6 years old (38.7 ± 10.9 years old in male patients, 37.9 ± 9.6 years old in female patients) for those patients with colorectal cancer.

Screening of patients and family members, with regular treatment of affected individuals, has led to a 55% reduction in the incidence of colorectal cancer at diagnosis of FAP, and an increase in cumulative survival for all FAP patients.[15,16] The American Gastroenterological Association recommends an annual sig-

moidscopy, beginning at the age of 10-12 year, for patients with a genetic diagnosis of FAC, or at-risk family members who have not undergone genetic testing.[17] Mutations at condons 1309 and 1328 in exon 15G were associated with a uniformly severe polyposis phenotype.[18] The mutational analysis may be a more accurate way of selecting patients for IRA or IPAA. James S. Wu et al. suggested that patients with mutations at these locations be strongly considered for total proctocolectomy and IPAA at the time of initial surgery.[18]

Principle of surgical management of adenomatous polyposis coli is early operation for all patients with adenomatous polyposis coli, operation especially should be carried out as soon as possible in patients who are diagnosed later in life.[12,13] Forty years ago, the choice of the surgery was either subtotal colectomy with ileorectal anastomosis to preserve anal function or total proctocolectomy with permanent ileostomy. Subtotal colectomy with ileorectal anastomosis has advantages of relatively low morbidity and maintaining good quality of life without a stoma. However, in spite of aggressive follow up with frequent removal or electrocoagulation of rectal polyps, cancer still develops in the rectal stump.[19] The disadvantages of the subtotal colectomy with ileorectal anastomosis included false security of subsequent development of cancer in the rectal segment, a threat of subsequent development of colorectal cancer and troublesome of follow up.[13] Although total colectomy with permanent ileostomy does have advantages of low morbidity and without further threat of development of colorectal cancer. However, a permanent stoma is associated with poor quality of life.[12,19] Restorative proctocolectomy of total proctectomy, mucosal proctectomy with ileal pouch and pouch anal anastomosis (IPAA) has been popularized by several surgeons since 1970'.[20-22] However the operation is frequently technically impossible, not suitable for people of old age, weak anal sphincter, locally advanced or widely spread rectal cancer and those who can not withstand long time of anesthesia. Besides, the IPAA is associated with many complications which included pelvic sepsis, subphrenic abscess, wound infection, fistula from pouch, rectovaginal fistula, hemorrhage from the reservoir, anastomotic stricture, pouchitis, pouch perforation, small bowel obstruction, electrolyte imbalance and malabsorption.[23,24] To make the things worse, about 20% of patients would end of losing the pouch after five years due to pouch-related complications.[25] So although IPAA has the advantages of fair to good quality of life and without threat of developing rectal cancer, it is associated with high morbidity and usually needs at least two operations.

Juvenile polyposis coli is a rare disease and frequently seen in the families of adenomatous coli. Besides juvenile polyps, the patients may also have adenomatous polyps in the colon. Most of experts believe that the juvenile polyposis may eventually become adenomatous polyposis, so the disease should be considered as high in malignant potential. Early operation is suggested for the patients with juvenile polyposis coli with efforts to preserve anal function without a permanent stoma, which means that subtotal colectomy with ileorectal anastomosis or IPAA should be considered as the procedure of choice.

Although restorative proctocolectomy (IPAA) with ileostomy was the operative procedure of choice for polyposis coli, but was only performed for 22 patients for different reasons. Sixteen patients still end up with a permanent stoma. Besides, IPAA is not without complications, four patients even had re-exploration for complications.

Thirty-two patients were found to have invasive adenocarcinoma in the specimen, which means that in spite of improvement in the diagnosis of the disease, many patients still present late in the course of disease. Thirteen patients died of metastatic colorectal cancer also meaned that early diagnosis should be pursued especially for the families who could inherited the disease. Although there are still several controversies in IPAA surgery currently,[27-29] the senior author of the article prefer transanal mucosectomy with hand-sewn anastomosis and not the double-stapled technique, the use of diverting ileostomy in all cases, open instead of laparoscopy colectomy and avoid IPAA in patients older than 55 years old. There were 19 patients with good function of pouch which encourage us to continue this time-consuming, complicating procedure especially in patients who might have long life expectancy. One patient is currently re-

ceiving treatment of desmoid tumor which alerts us that all the patients with polyposis coli should be closely followed for possible extraintestinal manifestations and malignancies.[19]

Conclusion

Colectomy should be recommended to all patients with polyposis coli to avoid death due to colorectal carcinoma. Restorative proctocolectomy is not a perfect operation, but can offer a reasonable good quality of life to the patient with polyposis coli if the operation is successful.

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病例分析

外科手術治療大腸腺瘤及青年息肉症的經驗

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目的 大腸腺瘤及青年性息肉症通常採用外科療法以預防大腸息肉的惡性變化。對於大多數的臨床醫師來說，由於息肉症在台灣地區的發生率不高，因此息肉症之手術治療的經驗的確比較缺乏。這篇回顧性研究分析，主要是呈現單一外科醫師對大腸腺瘤及青年性息肉症的手術經驗。

方法 從 1984 年 2 月到 2010 年 12 月，共收集 46 位罹患腺瘤及青年性息肉症患者資料，這些患者都接受同一位外科醫師的手術治療。這些研究資料包括患者的流行病學資料、手術治療的適應症、手術術式、手術併發症及死亡率。

結果 針對大腸息肉症患者的收集，共計有男性 23 位及女性 23 位。平均年齡為 34.2 ± 11.7 歲。最常見的開刀適應症為惡性腫瘤共有 32 位 (69.6%)，其次為只有大腸息肉症共有 13 位 (30.8%)。最常見的開刀術式為復原式結腸直腸切除術共計有 22 位 (47.8%)，其次為全結腸直腸切除術併迴腸造瘻共計有 9 位 (19.6%)，及次全結腸切除併腸吻合共計有 5 位 (10.9%)。治療追蹤期間共有 18 位患者死亡，其平均年齡為 43.0 ± 11.1 歲。術後併發症最常見的是腸道阻塞共計有 8 位 (17.4%)，其次為上消化道出血共計有 3 位 (6.5%)。

結論 預防性結腸切除術是目前大腸腺瘤及青年性息肉症患者預防大腸直腸癌發生的最佳治療方式，但決定採用何種術式，應視患者情況做最適當的選擇。

關鍵詞 多發性息肉症、次全結腸切除術、復原式結腸直腸切除術。