

Laparoscopic Total Proctocolectomy For A Case Of Familial Adenomatous Polyposis With Carcinoma Of The Rectum

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Citation

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Abstract

Familial adenomatous polyposis (FAP) is an autosomal dominant disorder characterized by multiple adenomatous polyps throughout colon and rectum with 100% risk of malignancy if not operated early. We came across a 32-year-old female patient who, after a battery of investigations was diagnosed as having multiple adenomatous polyps with low carcinoma of the rectum. Based on family history she was diagnosed to be a case of FAP for which LAPAROSCOPIC TOTAL PROCTOCOLECTOMY was done. Below is the discussion regarding presentation and surgical options.

ABBREVIATIONS: FAP - FAMILIAL ADENOMATOUS POLYPOSIS, APC - ADENOMATOUS POLYPOSIS COLI, CA - CARCINOMA

INTRODUCTION

FAMILIAL ADENOMATOUS POLYPOSIS (FAP) is an autosomal dominant disorder caused by germline mutation of APC gene on 5q21 with 100% penetration. FAP is characterized by multiple adenomatous polyps throughout colon and rectum and also in the stomach, duodenum and ileum. All affected individuals develop colorectal cancer, if prophylactic colectomy is not performed, usually by 35 years of age¹. The average age of patients with FAP at onset of adenoma detection is 15 years with almost 90% manifesting polyps by 30 years of age¹. Extraintestinal manifestations of FAP include desmoid tumors, osteomas and sebaceous cysts (GARDNER'S SYNDROME) and association with brain tumors (TURCOT'S SYNDROME).

CASE PROFILE

A 32-year-old female presented with complaints of episodic bleeding per rectum for 2 years, bright red and mixed with stools. The patient was operated for an abdominal wall swelling 6 years back. The patient's mother died at 38 years, diagnosed with carcinoma of the colon with a history of multiple abdominal surgeries. The patient's sister screened positive for multiple colonic polyps.

The patient's Barium enema revealed polyposis coli. Colonoscopy revealed multiple colonic polyps of varying size with a friable mass lesion in rectum. CT scan showed a 6cm anorectal growth with single perirectal lymph nodes. Upper GI Endoscopy was NAD. Retinoscopy showed bilateral peripheral retinal hyperpigmentation. Anorectal punch biopsy revealed moderately differentiated adenocarcinoma and colonoscopic biopsy revealed adenomatous dysplastic polyps.

With the diagnosis of FAP with low rectal carcinoma confirmed, the patient underwent total laparoscopic proctocolectomy. Intraoperatively, first, pelvic dissection was carried out, followed by mobilization of the descending colon, splenic flexure, transverse colon, hepatic flexure and terminal ileum, in that order. All vessels were ligated at their roots. The intact specimen was delivered by the peranal route after minimal perineal dissection. A permanent ileostomy was brought out through the RIF port site. Six ports were used in all.

Intraoperative blood loss was 400 ml. The patient had a remarkable postoperative recovery with mobilization and oral intake started from POD 2, minimal use of paraenteral analgesics and complete oral nutrition from POD5. The patient was discharged on POD12. Biopsy revealed more than 1000 polyps with dysplastic changes throughout the colon and a Dukes stage B1 moderately differentiated

adenocarcinoma of the rectum. Following surgery, the patient received a full course of chemotherapy.

DISCUSSION

FAP is among the most common inherited disease predisposing to cancer, occurring in approximately 1 in 10000 live births, and accounts for less than 1% of all colorectal carcinomas². The familial nature of FAP and its localization in the large bowel was first noted by Cripps in 1882³. The APC gene was identified in 1991.

Patients with severe phenotype (>1000 polyps) tend to present earlier, are more often symptomatic and more likely to develop colorectal cancer. A resected specimen with >1000 polyps has a double chance of containing a cancer compared with one with <1000 polyps⁴. There is association between severe polyposis phenotype and mutations at APC gene codons 1309 & 1328 and for these patients prognosis for retaining the rectum is poor⁵.

Besides the risk of CRC, other tumors associated with FAP include pancreatic, gastric, small intestine, thyroid and brain neoplasms, the most common being periampullary malignancy.

Treatment of FAP is chiefly surgical. Prophylactic surgery is commonly performed at 15-20 years when the risk of cancer is less than 3-4%⁶. Ideal operative management of colorectal polyposis should satisfy 3 criteria

- Removal of all colorectal mucosa at risk.
- Maintenance of low frequency, continence and transanal evacuation.
- Minimal operative complications.

Figure 1

TABLE : SURGICAL OPTIONS FOR MANAGEMENT OF COLO-RECTAL POLYPOSIS

PROCEDURE	ADVANTAGES	DISADVANTAGES	INDICATIONS
TOTAL PROCTOCOLECTOMY	Eradicates all mucosa at risk	<ul style="list-style-type: none"> • Pelvic & perineal dissection • Nerve injury • Permanent ileostomy • Difficult wound healing 	<ul style="list-style-type: none"> • Cancer of lower rectum at diagnosis.
COLECTOMY WITH ILEORECTAL ANASTOMOSIS	Normal continence Simple operation No pelvic dissection No ileostomy One stage procedure	<ul style="list-style-type: none"> • Risk of rectal cancer • Requires life-long surveillance. 	<ul style="list-style-type: none"> • Age <30 years • Absence of symptoms • Spared rectum (<100 polyps) • Metastatic cancer of colon/rectum
RESTORATIVE PROCTOCOLECTOMY WITH MUCOSECTOMY	Eradicates all mucosa at risk. Acceptable function & continence	<ul style="list-style-type: none"> • Usually two stage procedure. • Pelvic dissection • Sepsis/nerve injury • Mucosectomy 	<ul style="list-style-type: none"> • Age >30 years • Presence of symptoms • Rectal adenomas >100 • Poor compliance to follow-up • Family history of rectal cancer
RESTORATIVE PROCTOCOLECTOMY WITHOUT MUCOSECTOMY	Acceptable function Near normal continence	<ul style="list-style-type: none"> • Pelvic dissection • Retained mucosa at risk. • Requires surveillance 	<ul style="list-style-type: none"> • Age <30 years • Absence of symptoms. • Spared rectum.

Chronological age is an important risk factor. After IRA, the risk of rectal cancer developing by 50 years is 5-10% & by 60 years, the risk is 14-29%.

Major benefits of the laparoscopic approach to total proctocolectomy may prove to be long term rather than short term. Laparoscopy is associated with markedly less adhesion formation than laparotomy. Other benefits include decreased disability, less immune system trauma, diminished pain, expedited recovery and lower morbidity as compared to laparotomy. Young, thin, cosmetically conscious and physically active patients who wish to try to minimize the disability period are ideal candidates for the procedure.

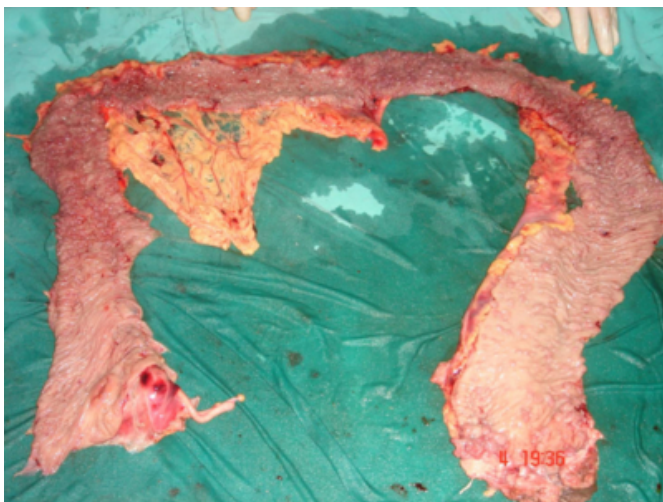
Figure 2

FIGURE 1: PROCTOCOLECTOMY SPECIMEN BEING DELIVERED THROUGH THE PERIANAL ROUTE



Figure 3

FIGURE 2: SPECIMEN SHOWING MULTIPLE ADENOMATOUS POLYPS THROUGHOUT THE COLON AND A CARCINOMA IN THE ANO-RECTUM REGION



CONCLUSION

Total proctocolectomy with Brooke's ileostomy completely eliminates the risk of CRC and is indicated for established low-lying rectal cancer

IPAA is indicated for patients with increased risk of rectal cancer (related to the number of rectal adenomas, presence of colonic cancer at colectomy and type of APC gene mutation), patients with symptoms and in patients where long-term follow-up cannot be assured.

IRA with close endoscopic surveillance and ablation of rectal polyps is an option for patients with low risk of rectal cancer and willing to comply with aggressive surveillance of the rectal stump.

Laparoscopic-assisted total proctocolectomy is a technically feasible procedure with considerable reduction in post-operative morbidity and early return to work, and it is oncologically safe.

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