

Anal transitional zone adenocarcinoma following restorative proctocolectomy for ulcerative colitis : case report and review of literature

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Abstract

Conservative proctocolectomy with ileal pouch-anal anastomosis (IPAA) has become the intervention of choice for severe ulcerative colitis (UC) requiring surgery. Sporadically, the occurrence of carcinoma arising in or near the ileo-anal pouch after IPAA for UC has been reported. This issue is of utmost importance as an endoscopic follow-up is mandatory also after the operation. The present paper reports a case of cancer which developed four years after proctocolectomy with IPAA in a young man suffering from UC. Moreover, the authors provide update (PubMed research) by literature review on this issue. (*Acta gastroenterol. belg.*, 2009, 72, 441-443).

Key words : ulcerative colitis, dysplasia surveillance, cancer in IBD, IPAA.

Introduction

Ulcerative colitis (UC) is characterized by mucosal inflammation that affects the rectum (proctitis) and extends proximally to involve a variable tract of the colon (1).

Severe UC, failing to respond to medical therapy, persistent inflammation and cancer risk are the main surgical indications in patients with UC. Conservative proctocolectomy with ileal pouch-anal anastomosis (IPAA) has become the intervention of choice (1,2). IPAA can be hand-sewn at the dentate line, after mucosectomy of the rectal stump or stapled at the anorectal ring, thus preserving a very short cuff of transitional mucosa (columnar cuff). Stapled IPAA is claimed to be quicker, technically less demanding and to have superior functional results, possibly because the anal transitional zone (ATZ) is preserved and the anal canal is manipulated to a lesser extent (2).

From a theoretical viewpoint, the preserved ATZ carries an increased risk of cancer, while after mucosectomy this event should be impossible. However, even with the latter, malignancy is reported to occur, probably because mucosectomy is incomplete, even in very experienced hands (3).

Since 1978 (4) a great number of IPAA have been performed. However, to our knowledge, only very few cases of ATZ malignancy have been described after this intervention. Here, we report a case of cancer developed after proctocolectomy with IPAA in a man suffering from UC and, we provide updates on this issue by literature review.

Case report

In 2001, a 38 years old man, with a 12 year history of UC and without familial history of cancerous or precancerous colorectal lesions, underwent follow-up colonoscopy, which revealed a rectal polyp, 7 cm from the anal verge, measuring 2.5 cm with macroscopically adenomatous feature. Biopsies were performed on the polyp and histologic examination revealed a tubular adenoma with high grade dysplasia. No biopsy was performed in the vicinity of the polyp. Both macro- and microscopic diagnosis were confirmed at repeated colonoscopy and histological examination. He thus underwent restorative proctocolectomy with stapled IPAA. Pathologists confirmed the diagnosis of UC with high grade dysplasia.

After the intervention, the patient joined the standard protocol of follow-up at our institution, with endoscopy assessment and multiple biopsies of the ATZ performed yearly. No abnormal findings were observed for 3 years. In the fourth year of follow-up, biopsies revealed high grade dysplasia. Following the technique described by Fazio and Tjandra (5), mucosectomy and advancement of the pouch to the dentate line were performed. Since histologic examination of the resected mucosa revealed intramucosal adenocarcinoma, an abdominoperineal resection of the pouch and anal canal with construction of a permanent ileostomy was performed. Post-operative histopathological examination showed a well-differentiated adenocarcinoma from a remnant of rectal mucosa at the anastomosis, infiltrating the muscularis mucosae (Fig. 1). No lymph nodes metastasis was observed (pT1N0) and no signal of distant spread of the disease was documented.

After 27 months (last consultation in June 2007) the patient was disease-free and well, without evidence of recurrence.

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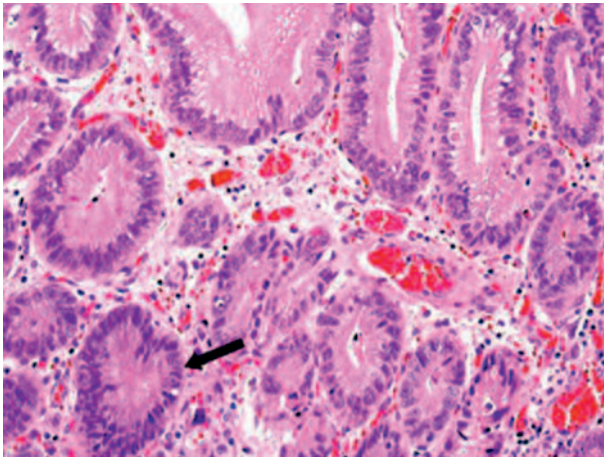


Fig. 1. — Microscopic examination (H&E $\times 20$) of the resected tissue showed a well-differentiated adenocarcinoma from a remnant of rectal mucosa at the anastomosis, infiltrating the *muscularis mucosae* (arrow).

Discussion

Despite the efforts in the fight against colorectal cancer, the burden due to this malignancy is expected to remain relevant in the next years. Therefore, the adoption of prevention policies is mandatory (6).

As a result of the disease, rather than an inherited phenomenon (7), patients with UC have a cumulative probability to develop colorectal cancer of 2% at 10 years, 8% at 20 years and 18% at 30 years from the diagnosis (8). After colectomy and ileorectal anastomosis, the risk of rectal cancer is not very different, 6% at 20 years and 15% at 30 years (9). On the contrary, total proctocolectomy completely eliminates such a risk, at the price of a permanent ileostomy. Conservative proctocolectomy and hand-sewn IPAA with mucosectomy would eliminate cancer risk without the need of a permanent ileostomy, while the ATZ which persists after stapled IPAA would carry a small but definite risk of malignancy. However, also after hand-sewn IPAA with mucosectomy cancer can occur. As a matter of fact, retained islands of rectal mucosa have been found in 20.7% of patients requiring removal of the pouch after initial mucosectomy (3). This is of particular concern, because retained rectal mucosa remains under the pouch and cannot be examined or biopsied. In practice, due to the potential cancer recurrence, a surveillance programme should include annual biopsies from the ATZ and the ileal pouch itself (10).

A PubMed research (www.pubmed.com) of all studies published from 1965 to 2008 was conducted. The final date of the search was August 30, 2008. The following medical subject headings were used : IPAA, ileal pouch-anal anastomosis, cancer, adenocarcinoma, ulcerative colitis, anal transitional zone. Only papers published in English language were considered. In addition to the case presented herein, another 21 cases of carcinoma arising in or near the ileo-anal pouch after IPAA for UC

(Table 1) (11-29) were found. In 15 cases, IPAA was hand-sewn with mucosectomy, in 5 it was stapled and in 1, the anastomotic technique was not stated. Two more cases were reported or cited without detailed data (30,31). The sole information about the patient cited by Ravitch MM. in his 1984 American Surgical Association presidential address is that the IPAA was performed in the '50s (30), before the description of this intervention by Parks and Nicholls. In the report by Rotholtz *et al.*, the stapled ileo-J-pouch-rectal anastomosis 10 cm cephalad to the dentate line cited (31) cannot be defined as a IPAA.

In some cases, the carcinoma could have arisen from pouch mucosa rather than from retained rectal mucosa, possibly in cases of chronic long-standing pouchitis. However, this cannot be proven with certainty when the time elapsed since pouch construction is short.

We did perform a stapled IPAA in a patient with rectal dysplasia. Remzi *et al.* stated that if dysplasia is demonstrated in the lower two-thirds of the rectum, then a hand-sewn anastomosis with mucosectomy should be done (32). We conclude that probably many surgeons would not have performed a stapled IPAA in such a situation.

One can suppose that if a hand-sewn IPAA with mucosectomy was performed, the patient described here might have been spared from subsequent rectal cancer. However, in the event of carcinoma arising from a retained mucosal island, malignancy would have been diagnosed at a more advanced stage, as follow-up biopsies would have been impossible. As underscored by Lee *et al.*, all cancers after mucosectomy were at least locally advanced (25) ; on the contrary, stage I cancers, including our case, have been detected only after stapled IPAA.

Table 1. — Cancer following ileal pouch-anal anastomosis : review of published cases

Publication year	Authors	Number of cases	Type of anastomosis
1990	Stern <i>et al.</i> (11)	1	Mucosectomy
1992	Puthu <i>et al.</i> (12)	1	Mucosectomy*
1995	Rodriguez-Sanjuan <i>et al.</i> (13)	1	Mucosectomy
1997	Sequens R. (14)	1	Stapled
1998	Vieth <i>et al.</i> (15)	1	Not stated
2000	Iwama <i>et al.</i> (16)	1	Mucosectomy
2001	Heuschen <i>et al.</i> (17)	1	Mucosectomy
2002	Baratsis <i>et al.</i> (18)	1	Stapled
2002	Hyman N. (19)	1	Stapled
2002	Laurenti <i>et al.</i> (20)	1	Mucosectomy
2003	Bell <i>et al.</i> (21)	1	Stapled
2003	Bentrem <i>et al.</i> (22)	1	Mucosectomy
2003	Hassan <i>et al.</i> (23)	1	Mucosectomy
2003	Negi <i>et al.</i> (24)	1	Mucosectomy
2005	Lee <i>et al.</i> (25)	3	Mucosectomy
2006	Knupper <i>et al.</i> (26)	1	Stapled
2006	Walker and Radley (27)	1	Mucosectomy
2007	Ota <i>et al.</i> (28)	1	Mucosectomy
2007	Das <i>et al.</i> (29)	1	Mucosectomy

* not stated in the case report, but it can be inferred as such from the discussion.

In conclusion, two important considerations can be made. First, stapled IPAA should not be performed in the case of rectal dysplasia. Second, stapled IPAA should indeed be performed in other cases in order to allow follow-up biopsies. As, even in the most experienced hands, rectal mucosa is retained in more than 20% of the cases of mucosectomy with hand-sewn anastomosis, we suggest that less experienced centres should perform stapled anastomosis and start a follow-up surveillance programme.

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