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LETTER TO THE EDITOR

Management of an Obese Patient with Familial Adenomatous Polyposis: Surgical Implication of Biliopancreatic Diversion and Total Colectomy

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Introduction

Obesity, defined as body mass index (BMI) greater than 30 kg/m^2, is a major worldwide multisystem condition considered to be the fifth leading risk factor for mortality. Obesity is associated with several comorbidities: type two diabetes, dyslipidemia, hypertension, heart disease, stroke, asthma, and cancer. Bariatric surgery (BS) is indicated when lifestyle modifications are ineffective. BS is effective both for weight loss and for improving and remitting obesity-related comorbidities [1]. Biliopancreatic diversion (BPD) is considered very effective for marked weight loss and comorbidity reduction [1]. Familial adenomatous polyposis (FAP) is a highly penetrant rare hereditary autosomal dominant syndrome caused by germline mutations in the tumor suppressor gene APC. FAP is characterized by the development of hundreds to thousands of adenomas of the colorectal epithelium from the 2nd decade of life [2]. FAP is characterized by lifetime risk of 95–100% to early develop colorectal cancer (CRC) if not adequately treated [2]. The severity of FAP could be also associated with the type of APC mutation [3]. FAP is a systemic syndrome that could be also characterized by the development of extracolonic tumors (duodenal, thyroid, brain, liver, and pancreas). Ten to 25% of FAP patients develop desmoid tumors (DT) in the abdomen and the abdominal wall, especially after surgical procedures. DT are fibrous benign tumors characterized by aggressive infiltrating growth harming the surrounding vital structures [4]. Surgery is the mainstay of FAP treatment. Extended colonic resections (e.g., total colectomy or proctocolectomy) are performed with prophylactic aim (CRC prevention) generally from the second decade of life [5]. The association between obesity and cancer has been studied with identification of different cellular and molecular pathways; however, no relation between obesity and FAP has yet been studied [6, 7].

Case Study

A 25-year-old Caucasian woman with morbid obesity (BMI 41, body weight 107.6 kg) was submitted in 2002, after failed diet therapy, to BPD and cholecystectomy in another institute. Preoperative radiological upper and gastrointestinal series were negative for neoplastic lesions of the digestive tract. BPD was successful with following weight loss (minimum weight 64 kg, %TWL 40.52, BMI 24.2). In 2006, the patient complained fatigue with evidence of microcytic anemia (Hb, 6.5 g/dl). The fecal occult blood test was positive. Gastroscopy was negative; however, colonoscopy reported > 40 polyps (diameter, 2–3 mm, low-grade dysplasia tubular adenoma). The patient was admitted to our Hereditary Digestive Tract Tumors Unit with the clinical suspect of FAP. Familial history

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was negative for polyposis and CRC. DNA sequencing (Sanger) identified a pathogenic variant of APC, c.509_512delATAG (p.D170VfsX4), which is reported to be associated with an attenuated form of FAP [3]. Clinical examination and abdominal MRI were negative for extracolic manifestations. Preoperative colonoscopy confirmed the presence of numerous diffused small polyps (> 100; diameter < 5 mm) with lower count in the left colon. The evidence of only 10 small polyps (< 3 mm), at the preoperative proctoscopy, indicated the planning of rectum sparing total colectomy. The patient was submitted to total colectomy with ileocecal valve preservation and cecum stump-rectal latero-terminal anastomosis. In order to reduce the risk of postoperative malabsorptive disorder and to have the chance to explore the duodenum, it was also decided to reverse the previous BPD by taking apart the ileo-ileal anastomosis and performing a jejuno-ileal latero-lateral anastomosis. The ileocecal valve was preserved in order to possibly restrain postoperative bowel evacuation frequency already high for the previous intervention. The colon specimen was 90 cm long with more than 100 polyps with dimension between 1 and 3 mm. All polyps analyzed were low-grade dysplasia tubular adenomas. During surgery, a 2-cm stiff nodule was removed "in toto" with a small portion of the gastric greater curvature. Immunohistochemistry of the nodule was actine (1A4) negative, CD117 negative, b-catenine positive, COX2 positive, PDGFR-alpha negative. The nodule was reported as a perigastric small desmoid lesion. Postoperative course was uneventful with discharge on the 7th postoperative day. Postoperative follow-up was structured as follows: proctoscopy every 6 months, annual clinical evaluation including blood tests, gastroscopy, and abdominal MRI every 1–2 years. During follow-up, we assisted a fast weight gain of 9 kg in 12 months (weight at colectomy was 64 kg). The maximum postoperative weight was 86 kg (total weight gain, 21 kg) gained at 44 months from surgery. The patient reported to regain a bulimic behavior worsened by the occupation as a canteen staff member with easy access to food. Psychotherapy was ineffective with the final weight of 90 kg. In the same period, we assisted a gradual increase in rectal polyp count at endoscopy with a mean of 7 polyps (< 3 mm of diameter; low-grade dysplasia tubular adenomas). Moreover, at 37 months, we documented high-grade dysplasia tubular adenomas at the cecal stump-rectum anastomosis. Proctectomy was proposed to the patient; however, the psychological condition, the high risk of desmoid formation (from a third laparotomy), and above all the will of the patient to avoid surgery with a possible consequent definitive ileostomy indicated to continue strict endoscopic surveillance. At 164 months from the colectomy, there was evidence of rectal stump adenoma and fundic gland polyps without occurrence of malignancy despite inadequate upper gastrointestinal surveillance.

Conclusions

This case report shows a rare case of FAP patient previously submitted to BS for morbid obesity. Several points should be highlighted. First, before BPD, the patient was submitted to upper and lower gastrointestinal series, both negative for lesions. We suggest the need, especially in patients submitted to BPD (with impossible duodenal endoscopic view), to always consider family history especially for polyps, CRC, or FAP extracolic manifestations in order to do a preoperative colonoscopy.

Second, in the case of FAP patients, the high risk of postoperative desmoid development (10–25%) should be always remembered when considering a surgical treatment for morbid obesity. Third, in the case of BS in FAP patients, it should be discussed if it is better to prefer restrictive techniques (e.g., sleeve gastrectomy) instead of malabsorptive techniques for obesity-related comorbidity improvement and remission, especially considering postoperative bowel evacuation frequency (which could be worsened from the combination of BS and FAP surgery) with a consequent strong impact on quality of life.

However, it is possible that BPD had a protective role in the present case delaying the diagnosis of large-bowel adenoma that occurred 10 years later (at the age of 30) compared with FAP patients. The patient has a type of APC mutation which is theoretically linked to an attenuated form of FAP, and, despite the relevant number of polyps (> 100), they were all small (1–3 mm) with no evidence of CRC. The son, who inherited the mother's APC mutation, instead received a standard prophylactic total colectomy at the age of 17 with > 100 colonic adenomas.

During surgery, the BPD was reversed in order to balance the postoperative malabsorptive symptoms and bowel movements. This did not benefit the patient who referred 5 daily movements with gradual weight gain. Moreover, the polyp burden and the report of high-grade dysplasia tubular adenomas have alerted our team especially considering that the patient has an attenuated form of FAP, therefore raising the question of whether BS has a protective role in FAP.

To our knowledge, this is the first reported case of FAP patient submitted to BS. Further studies are needed especially considering the high epidemiological impact of obesity.

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Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Ethical Approval For this type of study, formal consent is not required.

Informed Consent The patient has given her written consent.

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