Case report

Development of duodenal cancer in a patient with familial adenomatous polyposis

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Abstract: A Japanese woman with familial adenomatous polyposis in whom a duodenal ampullary adenoma underwent malignant change during a 10-year follow-up period is reported. After restorative proctocolectomy in 1989, and extensive small bowel resection for desmoid disease in 1991, regular surveillance duodenoscopies, including three to nine biopsies (mean, 4.8) were performed annually or biannually. Until 1995, the endoscopic findings of duodenal polyposis (including an ampullary polyp) did not progress and the histopathology did not worsen. In 1996, there was an increase in the number and size of the duodenal polyps, and the ampulla of Vater looked enlarged. Open surgery was discussed but not proceeded with because of the risk for short bowel syndrome. In January 1998, she was admitted with a diagnosis of acute pancreatitis. Duodenoscopy and radiological examination revealed that an advanced ampullary cancer had developed, and histopathology revealed a well-differentiated adenocarcinoma. Multiple hepatic metastases and ascites led to her death, in June, 1998. This in-vivo demonstration of the adenoma-carcinoma sequence highlights current limitations in the surveillance and treatment of duodenal lesions.

Key words: familial adenomatous polyposis, duodenal cancer, ampullary adenoma

Introduction

After prophylactic colectomy, periampullary cancer is one of the main causes of death for patients with familial adenomatous polyposis (FAP). Current treatment options are not optimal. Because prophylactic pancreaticoduodenectomy has been considered to be too aggressive, surveillance duodenoscopy is performed in order to detect duodenal cancer at an early stage.1

Approximately 10% of ampullary adenomas progress over a period of 5 years.2 The adenoma-carcinoma sequence is thought to exist in the FAP duodenum.3 However, clinical observation of the progression of duodenal adenoma to cancer has been rarely reported. We present a patient with FAP in whom a duodenal ampullary adenoma underwent malignant change during a 10-year follow-up period. This in-vivo demonstration of the adenoma-carcinoma sequence also shows the uncertainty of timing and appropriate intervention in patients with duodenal lesions in the presence of FAP.

Case report

A 21-year-old Japanese woman who presented with melena was diagnosed with rectal cancer in the presence of familial adenomatous polyposis (FAP).4 She underwent restorative proctocolectomy in 1989. Her father and younger sister were also affected with FAP. The APC gene was examined by polymerase chain reaction (PCR)-single-strand conformation polymorphism (SSCP) analysis and direct sequencing by a method reported previously.5 Codon 848 in exon 15 was found to be mutated(AAA → TAA).4 Duodenoscopy, on August 4, 1989, revealed that the patient was affected with severe duodenal polyposis (Spigelman stage IV6; Fig. 1). The histopathologic diagnosis of the ampullary lesion was adenoma with moderate atypia. Regular surveillance duodenoscopies, including three to nine biopsies (mean, 4.8) were performed, using a forward-viewer or side-viewer, annually or biannually, by experienced endoscopists (mainly by H.S.). Until 1995, the endoscopic findings of duodenal polyposis and ampullary...
polyp did not progress, and the histopathology of tissue taken from the ampulla of Vater did not worsen. In 1996, there was an increase in the number and size of the duodenal polyps. The ampulla looked enlarged, and histopathology of tissue taken from the ampullary adenoma had worsened slightly (Fig. 2; Table 1). These findings made us consider whether duodenal surgery was timely. However, because a large amount of small intestine had been removed in 1991 in order to resect an abdominal desmoid tumor, duodenal surgery was not performed for fear of the short bowel syndrome. At that time, devices for photodynamic therapy or argon plasma coagulation therapy were not available at our institute. Sulindac was then given to her, to suppress the rapid progression of the duodenal polyposis.

In January 1998, she was admitted to our hospital with a diagnosis of acute pancreatitis. Duodenoscopy