A duodenal villous adenoma associated with in situ carcinoma: A case report

SABAHATTİN ASLAN¹, BAHADIR ÇETİN¹, FATMA MARKOÇ², ABDULLAH ÇETİN¹

Departments of ¹General Surgery and ²Pathology, Ankara Oncology Hospital, Ankara-Turkey

Villous adenomas of the duodenum behave in a manner similar to villous adenomas of the colon and rectum, having both a high rate of recurrence after local treatment and a high incidence of malignant transformation. Villous adenomas of the duodenum are the most common benign periampullary neoplasms. They are considered as premalignant lesions with a malignant transformation rate as high as 60%. Periampullary carcinomas arising from the villous adenomas of the duodenum are also the most frequent extracolonic cause of death in Familial Adenomatous Polyposis patients. Although the Whipple procedure remains the procedure of choice in treating adenomas containing invasive cancer, the selection of an appropriate procedure to treat benign villous adenomas of the duodenum, especially those with carcinoma in situ is controversial. Here we report a 63-year old woman who presented with epigastric pain secondary to a mass of the ampulla of Vater. An endoscopic biopsy revealed adenocarcinoma while pathological workup after curative pylorus preserving pancreaticoduodenectomy revealed villous adenoma of the duodenum associated with carcinoma in situ. [Turk J Cancer 2001;31(4):162-167]

Key words: Adenoma, Villous, Vater's ampulla, Duodenum, Pancreaticoduodenectomy

Villous tumors are premalignant lesions, described as an entity in the rectum (1). Although uncommon, villous tumors of the duodenum (VTD) are encountered with increasing frequency, mainly as a result of the widespread application of gastroduodenoscopy for evaluation of patients with upper gastrointestinal complaints (1,2). VTD are the most common benign periampullary neoplasms and those arising from the ampulla of Vater has a high risk of malignant transformation (1,3,4).
Case Report

A 63-year old woman complaining of abdominal pain and fatigue for a few weeks was admitted to our clinic. She had no previous history of jaundice, weight loss or diabetes mellitus and was diagnosed to have hypertension for 3 months. Physical examination was normal. Laboratory data didn’t reveal abnormal values. Tumor markers (CEA, CA19-9, CA125, CA15-3, AFP) were in normal ranges. Abdominal ultrasonography (USG) and computed tomography (CT) detected minimal dilatation of intrahepatic bile ducts with a 10 mm wide choledochus, and a hypoechoic mass of 20x25 mm located in the head of the pancreas. Magnetic resonance imaging (MRI) revealed intrahepatic bile duct dilatation and heterogeneity in the uncinate process of the pancreas. Esophagogastroduodenoscopy demonstrated a minimally elevated, rough mucosa with an infiltrative appearance on the second part of the duodenum, on the Vater’s ampulla. The esophagus and stomach were intact. The pathological diagnosis of the endoscopic biopsy of this mass was adenocarcinoma. Based on these findings, the patient underwent pylorus preserving pancreaticoduodenectomy and paraaortic and hepatoduodenal lymph node dissection. There were no complications observed in the postoperative period.

The postoperative pathological diagnosis was villous adenoma with high grade dysplasia centered in the ampulla of Vater. Grossly, a soft, sessile, papillary mass was seen projecting into the duodenal lumen in the ampulla. It was 2.5 cm in size. Microscopically adenomatous epithelium growing in a villous arrangement was seen (Figure 1).

Fig 1. Villous adenoma showing adenomatous epithelium growing in a finger like arrangement. Haematoxylin and eosin staining (H&E; x20)
In some areas, the crypts showed irregular branching and cytologically the nuclei were enlarged and vesicular with prominent nucleoli. The nuclei were stratified and reached the cell surface. Mitotic figures were prominent at the surface. Those changes were considered high grade dysplasia or carcinoma in situ (Figure 2). There was no stromal invasion. All of the lymph nodes were reactive.

After having the diagnosis of villous adenoma of the duodenum, the patient underwent gastrointestinal surveillance. The passage graphs of the small intestine, double contrasted barium colonography and total colonoscopy didn’t reveal a coexistent polyp. And the patient didn’t have a Familial Adenomatous Polyposis (FAP) history either.

**Discussion**

Villous tumor of the duodenum was first described in 1893 as a broad based mass. In 1928 the first definitive case of villous adenoma of the duodenum was published. Since then 73 cases have been reported in a 1981 review (1). After that, VTD have been reported with increasing frequency over the last 20 years, probably because of the increasing use of upper gastrointestinal endoscopy for evaluation of patients with gastrointestinal complaints (2).

Incidence of neoplastic growth in the small intestine is usually one-tenth that of the similar lesions of the colon (1). Although the duodenum comprises 8% of the small bowel by length, it harbors 10%-20% of small intestinal tumors. Similarly villous tumors of the small intestine occur most frequently in the duodenum. However, they account for only 1% of all duodenal tumors (1).
Despite being relatively rare, VTD remain the most common benign periampullary neoplasm (1,3). VTD appear to behave in a manner similar to villous adenomas of the colon and rectum (1). VTD possesses a high risk of malignant development (4,5). Carcinomatous changes occur in 30 to 60% of duodenal villous adenomas and much less in tubulovillous and tubular adenomas (6). In fact, in Hoyuela’s report (4), villous adenoma was associated with adenocarcinoma in 50% of cases. Besides, Farnell et al. (7) pointed out in his report that villous tumors contained carcinoma in situ in 4% and invasive carcinoma in 22% of patients. In Galandiuk’s series (2), the incidence of malignancy was 47%.

VTD may occur in the majority of patients with Gardner’s syndrome and FAP (5,8). Polyps in the duodenum occur in 40-91% of FAP patients, and are true dysplastic lesions at risk of cancer. Periampullary carcinoma is now the leading cause of death after proctocolectomy in patients with FAP which is affecting up to 12% of patients. For these reasons patients with FAP should have routine esophagogastroduodenoscopy (1). Based on this data our patient was evaluated and no FAP was found.

VTD cause symptoms due to their location, usually near the ampulla of Vater. The most common presentation of such tumors is clinical and biochemical evidence of obstructive jaundice (9). The other common presenting symptoms are; pancreatitis, bleeding, duodenal obstruction, intussusception, anemia and vague abdominal complaints (1).

As small VTD can easily be missed by barium swallow examinations, today the most useful and accurate diagnostic tool is probably fiberoptic endoscopy with full visualization of the duodenum which enables both visual identification and biopsy of the tumor. Due to generally large lesion size and the small sample taken via the endoscopic biopsy forceps, inaccurate diagnoses are common, giving false negative benign results in 40% to 60% of cases (1). In fact, Menzel et al. (10) reported 62% of overall accuracy for endoscopic forceps biopsies. Because of these, excluding associated malignant disease prior to resection of an adenoma is not always possible (4). In our case, it was thought that the endoscopic biopsy was taken from the high grade dysplasia site of the polyp and diagnosed as adenocarcinoma.

The other methods that can be used in diagnosis and staging are; endoscopic retrograde cholangiopancreatography, USG, CT and endoscopic USG.

Although there is uniform agreement that VTD should be resected, opinions differ as to the optimal method of resection. The treatment options are; endoscopic snare excision or ablation, local submucosal excision (with or without sphincteroplasty and pancreatic duct septotomy in the case of periampullary VTD), full thickness excision, pancreas sparing duodenectomy and pancreaticoduodenectomy (PD). Regardless of the type of procedure, complete excision is mandatory (1).

Although PD (preferentially the pylorus preserving) remains the procedure of choice in treating VTD containing cancer, the selection of an appropriate procedure for benign VTD remains controversial (1). The problem of occult malignancy and false negative results confounds surgical management of VTD. In fact, up to 60% of cases may harbor carcinoma at the time of diagnosis (1).
Recurrence of benign villous tumors after local excision is common and may be malignant (7). Indeed the highest recurrence was associated with local excision (1,2). In a recent study from the Mayo Clinic, 24% of all recurrences were adenocarcinoma (1). Therefore, endoscopic or local excision should be considered only in patients who are unfit to operative excision (1,4,11).

In contrast with the colon which has no lymphatics above muscularis mucosa, the small bowel mucosa contains lymphatics that course through the normal villi extending near the luminal surface and thus even an intramucosal carcinoma arising in a VTD may theoretically metastasize before invading the muscularis mucosa, which suggests for PD and lymph node dissection (1).

Because of increasing experience in improving operative mortality (<4%) following PD, some surgeons strongly advocate PD as the initial surgical treatment in medically fit patients. PD achieve good results and it appears to be the procedure of choice in order to treat villous adenomas with proved presence of carcinoma, carcinoma in situ, severe dysplasia or in selected patients with benign villous tumors (4,7). Especially tumors located near the papilla, PD seems to be the best treatment (12). Presentation with jaundice and weight loss or hard areas on palpation, an ulcerated tumor, dilatation or extensive involvement of the common bile duct and/or pancreatic duct on preoperative imaging, severe dysplasia should be considered as suggestive of underlying malignancy and PD should be considered for the fit patients especially when an associated polyposis syndrome is present (1).

The 2- and 5- year survival rates for patients with adenomas containing invasive cancer were 22% and 0% respectively, compared to 87% and 87% respectively for benign adenomas (including those with carcinoma in situ) (2).

As a result, the presence of a VTD is an indication for surgical treatment. Selection of the method of treatment requires discerning surgical judgement based on clinical presentation, presence of an associated polyposis syndrome, information from preoperative diagnostic evaluation and intraoperative findings. It is important to realize that malignancy will not be diagnosed preoperatively in a significant proportion of the patients. PD prevents recurrence, is indicated for VTD containing cancer, carcinoma in situ, dysplasia and may be considered as a valuable alternative for selected, surgically treated patients with presumably benign VTD.

We performed pancreaticoduodenectomy in our case with no complications. Our patient is still alive with no complaints and evidence of recurrent disease for 16 months postoperatively. We recommend that, considering the false negative benign results of endoscopic biopsies, pancreaticoduodenectomy is an acceptable treatment option for villous tumors of the duodenum if the clinician is experienced enough.
References