

Villous Tumors of the Duodenum

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Villous tumors of the duodenum are rare, but treatment may be problematic because of their association with invasive adenocarcinoma. Two cases of villous tumor of the duodenum are described and 39 other reported cases are reviewed. Presenting symptoms were bleeding 27%; obstruction 24%; jaundice 22% and vague dyspepsia 20%. Diagnosis may be made by radiographic barium contrast evaluation of the duodenum, especially with the addition of air contrast hypotonic studies and by fibro-optic endoscopy. Twenty-seven per cent of villous tumors of the duodenum are associated with adenocarcinoma. Invasive tumor is more common in patients over 50 years old (35%), in tumors of the third and fourth portions of the duodenum (44%) and in tumors over 4 cm in diameter (30%). Local excision is the treatment of choice for benign lesions. Pancreatico-duodenectomy is recommended for tumors which include invasive carcinoma in patients without distal metastases.

ALTHOUGH fewer than 200 cases of epithelial tumors of the duodenum have been reported, recent advances in radiographic and endoscopic diagnostic techniques have led to the earlier and more frequent recognition of these lesions (Fig. 1). They include benign adenomatous polyp, Brunner's gland adenoma, villous tumor, and frank invasive adenocarcinoma. Benign tumors are adequately treated by local excision. Pancreatico-duodenectomy is usually recommended for invasive adenocarcinoma. Most villous tumors are benign, but since areas of frank invasion are frequently observed, the appropriate choice of treatment may be difficult. The present report reviews 39 cases of villous tumor of the duodenum described in the world literature and describes two new cases, with special emphasis on the choice of surgical treatment.

Case Reports

Case 1: J.F., a 52-year-old white woman, was admitted to New York University Hospital on January 7, 1973. She complained of vague dyspepsia for a six-year period and of increasing eructation during the five months

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prior to admission. There had been no melena, hematemesis, vomiting or abdominal pain. Radiographic barium contrast evaluation of the duodenum prior to admission had revealed a smooth filling defect in the second portion of the duodenum. Physical examination was normal. Laboratory data was: hematocrit 36%, white blood cell count 5,900/cu mm, total serum bilirubin 0.3 mg% and alkaline phosphatase 75 mU/ml (normal 30-100).

Celiac angiography performed on January 8, 1973, showed medial displacement of the gastroduodenal artery, with a few irregular vessels noted arising from the retroduodenal portion of that artery. Hypotonic air contrast barium study of the duodenum on January 9, 1973, showed a smooth mass in the second portion of the duodenum adjacent to the ampulla of Vater. Surgical exploration on January 11, 1973 revealed a sessile polypoid tumor, which was generally soft but had several firm areas. Frozen section examination showed villous adenoma with possible invasion. Pancreatico-duodenectomy was performed. The postoperative course was uneventful and the patient was discharged home on January 25, 1973. She has remained well during her one-year followup.

Pathological examination of the operative specimen showed a 4 × 0.7 × 4 cm sessile villous adenoma. The tumor was on the medial wall of the duodenum, 7 cm from the pylorus, but did not involve the ampulla of Vater. There was no evidence of carcinoma on the final sections (Fig. 2).

Case 2: D.G., a 69-year-old white woman, was admitted to New York University Hospital on September 5, 1973. For five years she had experienced epigastric distress, which had been attributed to a hiatal hernia. Barium contrast radiographic evaluation of the upper gastrointestinal tract two years prior to admission demonstrated a large hiatal hernia and a pedunculated lesion of the first portion of the duodenum. A repeat study, three months prior to admission, showed an increase in the size of the duodenal lesion. There were no other gastrointestinal symptoms.

Physical examination was normal. Laboratory data included: hematocrit 37%, white blood cell count 4,500/cu mm, total serum bilirubin 0.6 mg% and alkaline phosphatase 46 mU/ml (normal 30-100).

Laparotomy on September 3, 1973 revealed a 3 cm pedunculated polyp in the first portion of the duodenum and a 15 × 6 cm hiatus hernia sac. The hiatus hernia was repaired and an anterior duodenotomy was then made. The duodenal polyp had a stalk 2 cm in length. The base of the stalk was divided and oversewn and the duodenotomy was closed. The postoperative course was smooth and the patient was discharged on September 21, 1973.

Pathological examination of the specimen showed a mixed epithelial

polyp $3 \times 2 \times 1.5$ cm in size, with villous and adenomatous elements. The stalk was free of villous elements. No carcinoma was noted (Fig. 3).

Discussion

Neoplastic growth in the small intestine is relatively infrequent. Its incidence is one-tenth that of similar lesions of the colon.^{39,50} Furthermore, lesions of the small intestine are frequently benign³⁹ and asymptomatic, and may therefore be recognized only as incidental findings at autopsy.⁴¹ Although the duodenum comprises 8% of the small bowel by length, it harbors 10–22% of small intestinal tumors.^{3,19}

Forty-eight per cent of all duodenal tumors are benign.^{3,8,10,12,15,19,21,24,25,28,39,45,47} They include adenomas (45%), lipomas (18%), myomas (16%), fibromas (10%), angiomas (4%), neurogenic lesions (3%) and less common lesions including polyposis, teratomas and villous tumors.⁴¹

The remaining 52% of duodenal tumors are malignant. They include adenocarcinomas (87%) and sarcomas (13%).²⁶ Carcinoid tumors and lymphomas are also found in the duodenum.¹⁹ Approximately 45% of all small bowel carcinomas occur in the duodenum.²²

One per cent of duodenal tumors are villous in nature. The villous tumor is an epithelial lesion²⁷ arising from or migrating on the surface of the mucus membrane.⁴³ The term "villous" tumor arises from the fact that these lesions have a velvety texture with finger-like projections resembling villi. It has also been called papilloma, papillomatous polyp, papillary tumor, papillary adenoma, papillary polyposis, villous papilloma and villous adenoma.⁴³ On

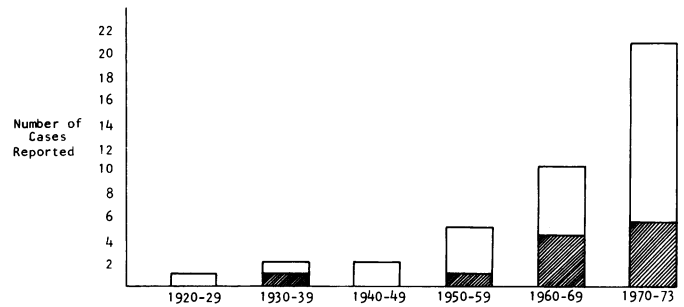


FIG. 1. Number of cases of duodenal villous tumor reported in each decade since 1920. The cross-hatched areas represent villous tumors with invasive carcinoma.

microscopic examination it has frond-like projections of mucosa with branching papillary structure²³ (Figs. 2 and 3).

Villous tumors are seen most frequently in the colon where they comprise 5–28% of all benign tumors.^{17,23,36} Up to 88% of colonic villous tumors are in the distal sigmoid and rectum.³⁵ The reported incidence of invasive adenocarcinoma in villous tumors of the colon and rectum is 30%^{2,35,43} with a range of 5%²³ to 55%.¹⁷ Because of the frequent malignant associations of villous tumors, total excision biopsy is usually recommended.^{2,4}

In the rectum, as in the duodenum, complete excision of the involved bowel results in major morbidity and assessment of the presence or absence of invasive carcinoma in villous tumors assumes increased importance.

A total of 82 villous tumors of the stomach have been reported.^{6,49} Gastric polyps are found in 0.2% of autopsies¹⁸

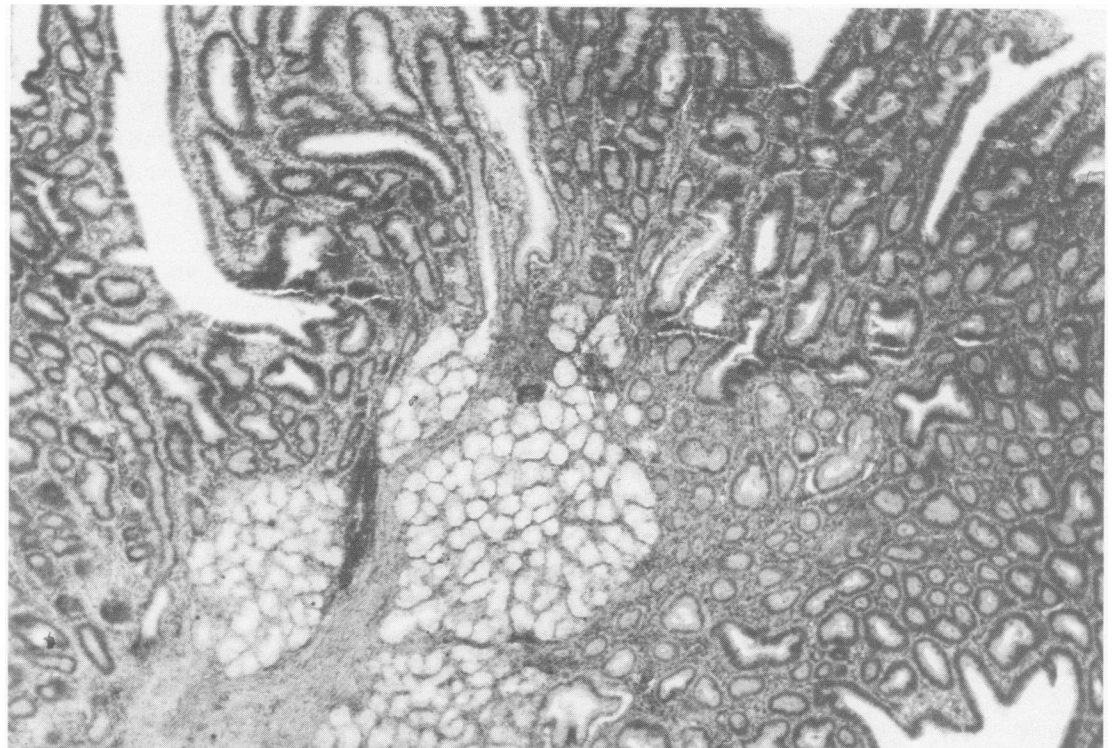


FIG. 2. Photomicrograph of tumor from Case One. (Hematoxylin and Eosin \times 100 original magnification)

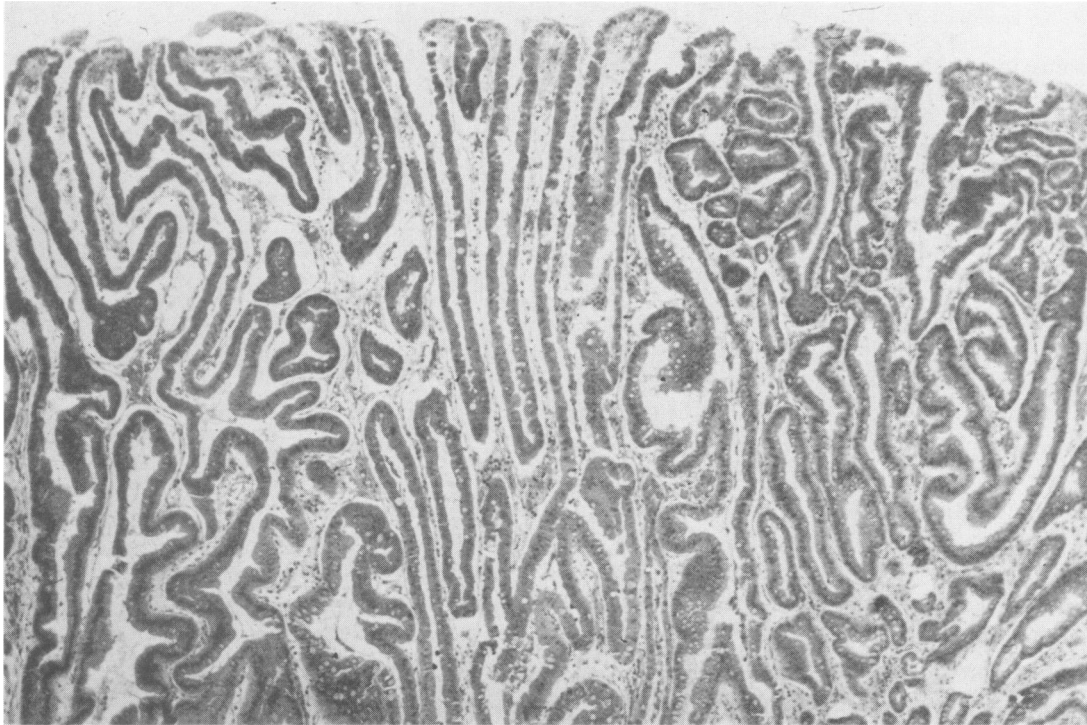


FIG. 3. Photomicrograph of tumor from Case Two. Brunner's glands are seen in the base. (Hematoxylin and Eosin \times 100 original magnification)

and 2% of gastric polyps are villous tumors.⁵² Sixty per cent of reported gastric villous tumors showed areas of adenocarcinoma.⁵²

Three villous tumors of the ileum have been reported and all were benign.⁶ There are eight reported cases in the jejunum,³⁰ three of which had invasive changes.

The present paper adds two cases of villous tumor of the duodenum to the 39 cases previously reported.^{1,5,7,9,11,13,14,18-21,25,29-34,37,38,40,42,44-46,48,50,51,53} Of these 41 cases, 11 or 27% showed evidence of invasive changes.^{13,14,25,32,34,40,44,48,51} This incidence compares closely with the percentage of invasive adenocarcinoma reported in the colon.

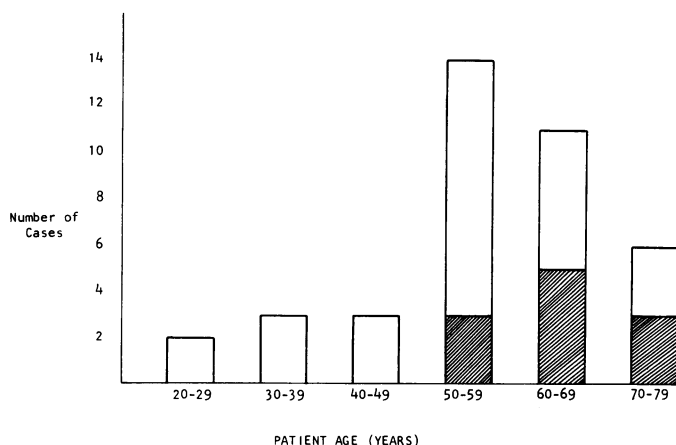


FIG. 4. Patient age in reported cases of villous tumor of the duodenum. The cross-hatched areas represent tumors with invasive carcinoma.

Villous Tumors of the Duodenum

Of the 41 cases reviewed, 24 (56%) were women and 17 (44%) men. The age range was 25–76 years (mean 57 years). Eighty per cent of patients were 50 years or older (Fig. 4).

The duration of presenting symptoms varied from four weeks³² to sixteen years.²⁵ These included: 1) gastrointestinal bleeding of variable degree manifested by anemia, melena or hematemesis. This was present in eleven cases, three of which had major upper gastrointestinal bleeding.^{29,30,33} 2) Obstructive symptoms, including frequent eructation or vomiting, were present in ten cases. 3) Obstructive jaundice was present in nine cases. 4) Vague dyspepsia was the presenting symptom in eight cases. 5) The lesion was an incidental finding on x-ray in two cases^{20,37} and at autopsy in one case (21). Of note is that six patients had undergone previous surgical exploration for these symptoms before the diagnosis was established.^{7,9,30,32,33}

Radiographic barium contrast evaluation of the upper GI tract was performed in 30 cases. These x-rays revealed the lesion in all but two cases.^{30,33} Radiographically, the lesion appears as a filling defect with barium coating the interstices and appearing as a fine network in a palisaded or striated appearance.⁴⁰

Endoscopy was performed in two cases.^{20,46} Both of these lesions were in the first portion of the duodenum. In one case the lesion was red, strawberry-like and polypoid.⁴⁶ In the other case it was said to have the appearance of a colonic villous adenoma.⁵¹

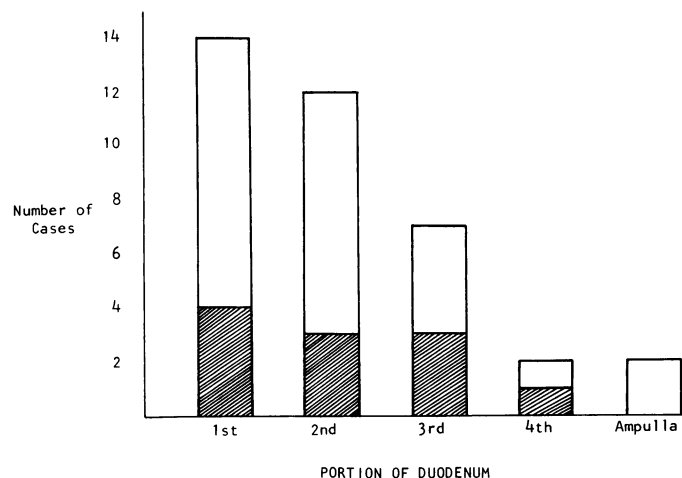


FIG. 5. Location of reported villous tumors of the duodenum. The cross-hatched areas represent tumors with invasive carcinoma.

Hypotonic duodenography may be useful and is the best method for obtaining air-contrast coating of a lesion.⁴⁰ It proved useful in Case 1 of our report. It should be mentioned that angiography in our Case 1 actually proved to be misleading.

Most villous lesions were in the proximal duodenum. There were 14 lesions in the first portion, 12 in the second, seven in the third and two in the fourth. There were also two lesions of the ampulla (Fig. 5). Of the two lesions in the fourth portion, one was an adenocarcinoma,³⁹ and of the seven in the third portion, three showed invasive changes.^{14,37,40} The distal duodenum thus contained only 24% of reported lesions, but 44% of these showed invasive changes. There were four invasive lesions in the first portion^{25,34,44,51} and three invasive lesions in the second portion (25%) (Fig. 5).^{13,32} These figures may be the result of the less

frequent early detection of benign lesions in the less accessible distal area of the duodenum.

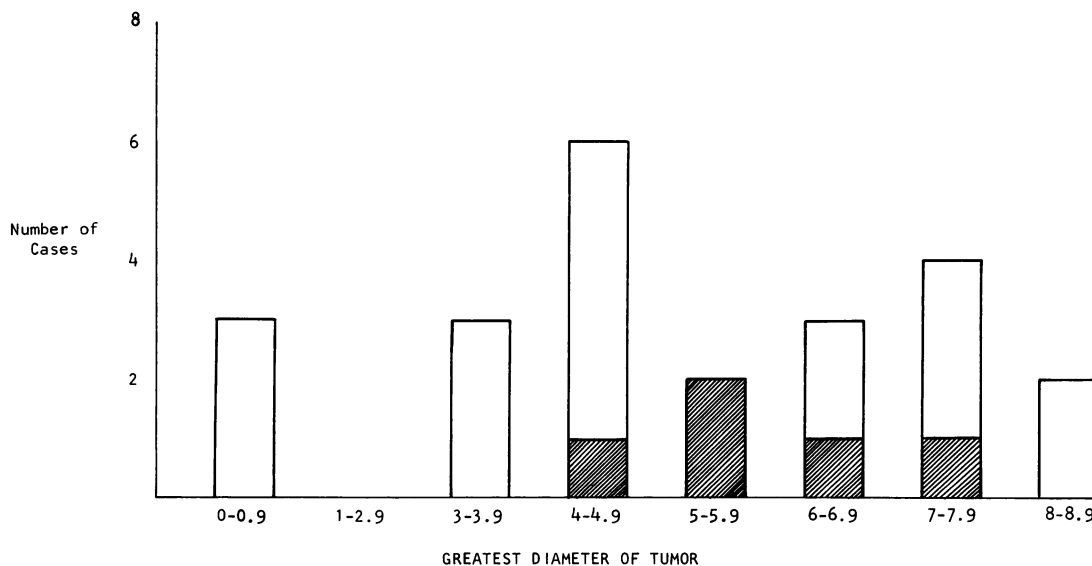
No malignancies were noted in patients under 50 years old. The incidence of invasive changes rises with age (Fig. 4). This contrasts with villous tumors occurring in the colon and rectum. Here, carcinoma has been reported at the age of 30 years¹⁷ and the average age for invasive change in villous tumors is 55.^{2,17}

There was no clear relationship between symptoms and invasive carcinoma. Five of nine patients who were jaundiced did not have invasive tumors.

The size of the lesion was reported in 23 of the 41 cases. This ranged from 4 mm^{9,21} to 8.5 cm.¹⁸ No lesions under 4 cm in size showed evidence of invasion. Of the remaining 17 lesions, five showed invasion (29%) (Fig. 6). In colorectal lesions, it also appears that the larger the lesion, the higher the incidence of malignancy.^{2,36} However, 7-24% of villous tumors of the colon and rectum under 2 cm in size are reported to show invasive changes.^{17,36}

Although rare, villous tumors of the duodenum are being diagnosed more frequently as a result of improved diagnostic techniques (Fig. 1). These tumors may give rise to bleeding, obstructive symptoms or jaundice. Duodenotomy and polypectomy are adequate surgical treatment for benign villous tumors. However, more radical excision may be indicated in the presence of invasive carcinoma. As a result of the development of fibro-optic endoscopy, histological evaluation of the surface of the lesion may be available. The possibility of invasive adenocarcinoma deep within the tumor must, however, always be considered. Although the gross characteristics of the tumor upon inspection and palpation are clearly of primary importance, this review of reported experience indicates that the incidence of invasive carcinoma is especially high in villous tumors of the distal duodenum, in those in patients over 50

FIG. 6. The diameter in centimeters of reported duodenal villous tumors. The cross-hatched areas represent tumors with invasive carcinoma.



years of age and in tumors exceeding 4 cm in diameter. Furthermore, these findings suggest that improved diagnostic techniques, and the removal of these lesions from patients who are younger and whose lesions are smaller, may reduce the incidence of invasive carcinoma.

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