

## Duodenal tumors: four case reports

V. PASTA, M. BIFFONI, G. MARTINO, M.S. MALAVENDA, D. SOTTILE, L. MARINACCIO,  
R. MEROLA, M. VERGINE, M.G. BERNIERI, M. MONTI, A. REDLER

SUMMARY: Duodenal tumors: four case reports.

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*We report 4 cases of neuroendocrine tumors of the duodenum. Signs and symptoms were non-specific. The choice of surgery depended on the site and stage of the tumor and any concomitant diseases.*

KEY WORDS: Duodenal carcinoid - Duodenal surgery - Duodenal somatostatinoma - Duodenal cancer

### Introduction

The small intestine comprises about 80% of the total length of the intestinal tract and about 90% of the mucosal surface. Tumors in this area are rare in general, but are most common in the duodenum (1). According to Santoro (2), duodenal tumors make up about 35% of benign and 17% of malignant growths in the small intestine, while 0.3% of all malignant gastrointestinal tract tumors originate in the duodenum.

Benign growths are generally non-symptomatic, with only 25% of cases presenting signs attributable to duodenal diseases. They are often found only during autopsy. The incidence varies considerably according to the caseload, but is likely to rise due to the increased use of diagnostic endoscopic examinations (3).

Moulinier (4) reported 14 benign duodenal tumors in 1000 endoscopies, while Shindo (5) found 3 in 134. Frattòn and colleagues described the endoscopic appearance of 14 duodenal tumors found in 700 duodenoscopies. Two of these were primary tumors, while the others were benign growths, with a strong prevalence of Brunner's gland hyperplasia (6).

The most common benign tumors are leiomyomas

and adenomas. Treatment depends on the site, histological type, extension and patient compliance (7).

Malignant tumors comprise 75% of symptomatic duodenal tumors and, when possible, require surgery.

The monitoring and diversified multidisciplinary treatment of four patients with duodenal tumors led us to make the observations discussed in this article.

### Patients and methods

The cases of four patients (three men and one woman) aged 35 to 73 years and suffering from duodenal growths with different features that required a diversified therapeutic approach (Table 1) are reported herein.

**Case 1** - 58-year-old man with gastroduodenal transit impairment due to duodenal stenosis caused by carcinoma of the first segment of the duodenum. The patient underwent subtotal gastrectomy with resection of the first segment of the duodenum. The carcinoma had not infiltrated the margins and the histological examination suggested carcinoma with unspecified neuroendocrine histological features. The patient was prescribed treatment with somatostatin, but one year later a metastasis was found in the fourth liver segment, verified histologically following metastasectomy. A year later the patient presented additional multiple liver metastases, which led to his death.

**Case 2** - Woman, aged 35 years, with dilation of the main bile duct in the absence of calculi and thickening of the gall bladder walls as in chronic cholecystitis. During open cholecystectomy, cholangiography revealed an obstacle in the papilla. Opening of the duodenum revealed an obstructive growth in the papilla (not found on gastroduodenoscopy) about 1 cm in diameter. Extemporaneous histological examination indicated a carcinoid tumor of Vater's papilla.

TABLE 1 - SCHEME SUMMARIZING THE CASES OF DUODENAL TUMORS.

Patient	Sex	Age (years)	Location	Histological type	Treatment
1	Man	58	First segment	Neuroendocrine carcinoma	Sub-Total gastrectomy
2	Moman	35	Juxtapapillary	Carcinoid	Duodenocephalo-pancreatectomy
3	Man	73	First segment + Gastric carcinoma	Somatostatinoma	Sub-total gastrectomy
4	Man	52	Second segment	Carcinoid	Endoscopic Resection

A duodenocephalopancreatectomy (DCP) was performed, with direct closure of the pancreatic stump. The definitive histological examination confirmed the diagnosis. We followed the patient for at least 10 years after surgery. Apart from her considerable weight loss due to intestinal malabsorption, there were no signs of recurrence (8).

**Case 3 -** Man, aged 73 years, who had been complaining of dyspepsia, asthenia and slight anemia for around a month. Endoscopy revealed an ulcerated growth in the gastric antrum, extending along the distal tract. A hypertrophic plica, covered with normal mucosa, was also found in the first segment of the duodenum. Biopsies detected an intestinal gastric adenocarcinoma (Lauren classification) and a hyperplastic duodenal polyp with diffuse complete intestinal metaplasia. There was also chronic atrophic gastritis in the absence of *Helicobacter pylori* infection.

A total body CT confirmed the gastric tumor and also revealed a vegetative polypoid growth in the first duodenal segment, maximum diameter 2 cm, which had not been found on endoscopic examination. There were 1-cm lymphadenopathies in the celiac tripod and around the emergence of the superior mesenteric artery.

The patient underwent subtotal gastrectomy with lymphectomy and removal of the duodenal polyp. Histological examination confirmed the intestinal type gastric adenocarcinoma (G3), and lymph node metastasis with extranodal extension in just 1 lymph node of 24. The duodenal growth was found to be a carcinoid tumor (cells positive on staining for somatostatin).

Around 2 years after surgery the patient developed liver metastases and peritoneal carcinomatosis, leading to his death (9).

**Case 4 -** Man, 52 years, whose sole symptom was gastroesophageal reflux of recent onset. Endoscopy revealed diffuse hyperemia in the pharynx and larynx, as with acid reflux, cardias mucosa normally located, slight hyperemia of the antrum and a 12-mm polypoid growth with central alteration in the second duodenal segment. Biopsies excluded HP infection and histological examination of the duodenal growth confirmed that it was a well-differentiated neuroendocrine carcinoid tumor.

The growth was removed via endoscopy and definitive histological examination of the polyp confirmed the diagnosis. Removal of the lesion resolved the gastroesophageal reflux, which was thus probably due to the obstruction caused by the duodenal growth.

Subsequent investigations (Octreoscan, CT/PET, US-guided endoscopy and biopsy in the surgical area after six months) confirmed the complete removal of the growth (Figs. 1, 2). There were no signs of recurrence one year later.

## Discussion

Tumors of the small intestine are relatively rare. The



Fig. 1 - Duodenal carcinoid (endoscopic image).

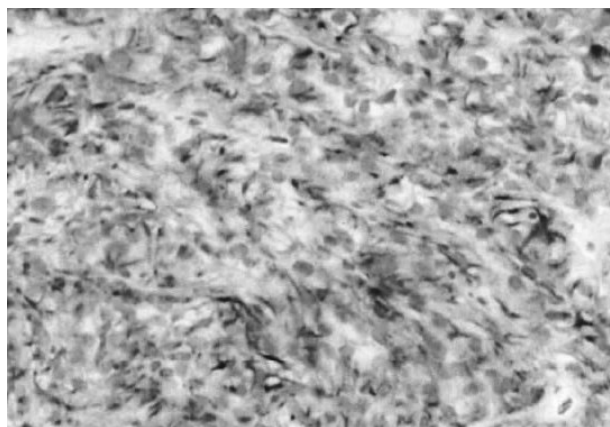


Fig. 1 - Immunohistochemistry demonstrates positivity to cytokeratin AE1/AE3 with predominant expression type "Dot Like" and positive staining for chromogranin A.

duodenum is the most frequently affected tract, comprising 30-50% of cases (3, 6, 10) in comparison with the jejunum (23%) and ileum (16%) (10). The small intestine might be more "resistant" to the onset of tumors for the following reasons:

- fast transit;
- sterile contents, and consequently reduced development of carcinogens;
- high concentration of the microsomal benzopyrene-hydroxylase enzyme system, neutralizing some specific carcinogens that might be present;
- immunoglobulin production;
- rapid mucosa turnover;
- intraluminal alkalinity (11).

The reason for the relatively greater incidence of tumors in the duodenum is unclear. Some authors suggest it may be due to possibly carcinogenic substances in the bile and pancreatic juices, a theory which is supported by the prevalence of these tumors in the peri- and sub-ampulla areas (12, 13). In fact, one of the first topographic classifications of these tumors was proposed in 1894 by Pic (14), who identified three locations:

- supra-ampulla;
- peri-ampulla;
- sub-ampulla.

The first is the least frequent (13, 15), and the peri-ampulla the most frequent.

An even more precise classification, especially for its prognostic implications, differentiates tumors according to the duodenal segment affected (D1, D2, D3, D4, angle of Treitz), as the embryological origin of the various segments differs (11, 13).

The diagnosis and treatment of duodenal tumors has been a constant topic of debate among surgeons. The combination of endoscopy and radiological examination enables diagnosis of 80% of cases of duodenal carcinoma (13). While CT and ultrasound are of little use in the diagnosis of primary disease, they are essential when searching for distant metastases and for staging. Although it is still little used, the first choice of examination should be US-guided endoscopy (11, 13, 16), which enables nearly 100% accuracy in terms of parietal invasion and 66% accuracy with respect to lymph node involvement (12, 15, 16).

The frequent malignancy of polypoid lesions, often in association with syndromes such as celiac disease in adults or Crohn's disease, justifies the careful radiological and endoscopic follow-up of these patients. Where laparotomy is performed for other reasons, intraoperative exploration of the duodenum is advisable, even in patients who are negative for Crohn's disease and celiac disease on endoscopy (17).

Other authors (18) assert that duodenocephalopancreatectomy (DCP) is the only potentially curative method for the treatment of duodenal adenocarcinoma, although long-term survival is influenced by various factors (stage, differentiation and lymph node involvement) (18). Still others (16) report that duodenal segmentec-

tomy following intestinal derotation is a safe, simple procedure for the treatment of primary adenocarcinoma of the third and fourth duodenal segment, preferring it over DCP due to its negligible morbidity and mortality and because it generally enables satisfactory margins of resection and adequate lymphadenectomy (16).

In any case, in operable tumors, the type of procedure varies according to the location, histological type, diffusion, presence of concomitant diseases and the need for more or less extensive lymphectomy (13, 15). In early forms, endoscopic excision should be considered (13, 19, 20); the long-term results are similar to those of traditional surgery, and morbidity and mortality are not affected (13).

The results of chemo- and radiotherapy are discomfoting (12, 21). For this reason, radical surgery should be the objective following early diagnosis.

Our caseload consisted of neuroendocrine tumors, albeit with the co-presence of a gastric adenocarcinoma in two cases. The signs and symptoms of the latter prompted the investigations which led to the detection of the duodenal tumor, that would otherwise have been misrecognized.

In case 1 the endoscopy was diagnostic, while in case 3 it was not exact, and the duodenal growth was only revealed during the CT carried out for preoperative staging. The symptoms were non-specific in all cases, although the slowed duodenal transit in association with GERD in the absence of hiatal hernia in case 4 was the sign which induced us to carry out the gastroscopy which led to the diagnosis.

It is not possible to establish general rules for treatment. We carried out surgical removal of the duodenal tumor plus gastrectomy in the patient with associated gastric growth. In the tumor in Vater's papilla we carried out DCP, while in the fourth case, the early diagnosis enabled us to remove the tumor endoscopically.

## Conclusions

The rarity of duodenal tumors and the anatomic characteristics of this site means that the choice of treatment necessarily varies according to the segment affected, the extension of the tumor and the presence of any concomitant diseases. Early diagnosis can influence subsequent therapeutic choices, even if the initial symptoms are almost always non-specific, scarce or totally absent.

In any case, even non-specific symptoms such as gastroesophageal reflux should not be underestimated and should indicate the need for further diagnostic investigation.

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