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CARCINOID TUMOR OF THE DUODENUM: a rare tumor at an unusual site. Case series from a single institution

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ABSTRACT - ***Context*** - Duodenal carcinoids are extremely rare, and their characteristics and biological behavior have not been fully elucidated. ***Objective*** - To analyze the clinicopathological characteristics of patients with resected duodenal carcinoids. ***Methods*** - Twenty patients (12 females and 8 males) were investigated. Their average age was 66.4 ± 5.8 years old (43 to 88 years old). The data corresponding to the clinical picture, diagnosis, treatment, and prognosis of patients with duodenal carcinoid tumors subjected to resection over a period of 18 years (1993-2011) were analyzed. ***Results*** - The most common symptoms were dyspepsia (50%) and epigastric pain (45%) followed by weight loss (10%) and vomiting (5%). Carcinoid syndrome was not observed in any patient. The lesion was located on the first part of the duodenum in 15 (75%) patients, the second part in 4 (20%) patients, and the third part in 1 (5%) patient. The diagnosis of a carcinoid tumor was established through an endoscopic excision biopsy in 19 (95%) patients and an histopathological examination of the surgical specimen in 1 (5%) patient. The average tumor size was 1.1 cm ± 0.4 cm (0.3 cm to 6.0 cm). Nineteen (95%) patients were initially treated by endoscopic resection of the duodenal lesion. One patient (5%), whose tumor was on the third part of the duodenum underwent a duodenectomy of the third and fourth duodenal parts and duodenojejunal anastomosis. The duodenal carcinoid resection margin was involved in four (20%) patients. Four (20%) patients were subjected to a partial gastrectomy to fully remove the lesion. The tumor was restricted to the submucosal layer in 16 (80%) cases, and it penetrated into the muscular layer in 4 (20%) cases. All patients exhibited positive chromogranin A, neuron-specific enolase, and/or synaptophysin immunostaining. The average duration of the follow-up period was 39.6 months (3 to 96 months). Twelve (60%) of the 20 cases in this series are alive without any evidence of active disease. Only one (5%) patient died due to liver metastases of the duodenal carcinoid. ***Conclusions*** - Duodenal carcinoids are rare and indolent tumors usually associated with a benign progression. Duodenoscopy, computerized tomography, and endoscopic ultrasound should be performed to evaluate the tumor size, the level of wall invasion, and the presence of regional or distant lymphatic metastases. Endoscopic removal of tumors smaller than 1.0 cm without periampullary localization or evidence of muscular propria layer invasion assessed by histology and/or endoscopic ultrasound is recommended. The endoscopic resection with a carcinoid tumor size between 1.0 cm and 2.0 cm can be incomplete and require new endoscopic resection or even surgical removal. Duodenal carcinoid larger than 2.0 cm require full-thickness resection and concomitant lymphadenectomy.

HEADINGS – Intestinal neoplasms. Duodenum. Carcinoid tumor. Carcinoma, neuroendocrine. Apudoma.

INTRODUCTION

Carcinoid tumors are neuroendocrine neoplasias most commonly found in the gastrointestinal tract, ovaries, lungs, bronchia, testicles, and larynx[1, 14, 24]. Carcinoid tumors are usually more benign than the epithelial carcinomas at the same anatomical sites, although the term "carcinoid" covers a wide group of neoplasms that arise from a variety of neuroendocrine cells characterized by varying degrees of aggressiveness[1, 18].

Carcinoid tumors represent 1.2%-1.5% of all gastrointestinal tract neoplasms[14, 18, 24]. Within the gastrointestinal tract, carcinoid tumors are most commonly found in the appendix followed by the ileum, rectum, and stomach[3, 14, 15, 21, 22, 24]. Carcinoid tumors represent 1/3 of all malignant tumors that affect the small intestine[15, 18]. These tumors rarely involve the duodenum, but they are increasingly recognized as a consequence of the now generalized use of an upper gastrointestinal endoscopy[15, 18]. These tumors are usually solitary, small, and restricted to the duodenal submucosa[15].

Important features of the natural history, the ideal extent of surgical treatment, and the prognosis of this

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neoplasm have not yet been elucidated due to their rarity and to the inclusion of patients with ampullary carcinoid tumors and gastrinoma in the case series, which often express somatostatin and gastrin, respectively, but exhibit different biological behaviors compared with duodenal carcinoids[2, 5, 18].

The present study performed a retrospective analysis of the clinical, diagnostic, histopathological, therapeutic, and prognostic features of patients with duodenal carcinoid tumors who were treated at our institution.

METHODS

This study was conducted in accordance with the regulations of the Ethics in Human Research Committees at our institution and in accordance with the ethical principles of the Declaration of Helsinki.

A total of 20 patients with a primary duodenal carcinoid confirmed by histopathological and immunohistochemical exams were treated at our institution over a period of 18 years (1993-2011).

Diagnosis of carcinoid tumors was performed according to the World Health Organization's (WHO) histological classification of tumors[23]. Patients with periampullary carcinoid tumors or gastrinoma were excluded from this study. The anatomopathological reports were reviewed, and some features such as location and the number of lesions, macroscopic appearance, size, depth of wall invasion, presence of angiolymphatic invasion, type of resection, status of the resection margin, presence of metastases, and the immunohistochemical expression of neoplasms, were recorded.

The biodemographic and clinical data (e.g., clinical aspects, diagnosis, presence of metastases, presence of synchronic tumors, treatment, length of follow-up, relapse, and survival) of the patients were obtained from the hospital records and interviews with the patients or their relatives. The survival length was estimated from the time of the endoscopic or surgical resection and the last follow-up visit or death.

The diagnosis was made after histopathological and immunohistochemical examinations of tissues obtained by an upper digestive endoscopy or from surgical specimens. Tumor sections were analyzed to assess the level of invasion of the duodenal wall, histological pattern, lymphatic, vascular, or neural invasion, mitotic index, and the status of the tumor margin. Immunohistochemical investigation by the avidin-biotin technique was performed in all cases.

Preoperative staging was performed based on the data of the physical examination, upper digestive endoscopy, biopsy, endoscopic ultrasound (EUS), and computerized tomography (CT) of the abdomen and thorax.

Quantitative variables are represented as absolute frequency (N), relative frequency (%), and means with standard deviation.

RESULTS

The average time between onset of symptoms and treatment was 3.3 ± 1.2 months, ranging from 1 to 6 months. The most common symptoms were dyspepsia (50%) and epigastric pain (45%), followed by weight loss (10%) and vomiting (5%). Physical examinations did not reveal significant findings. No other malignant neoplasias were found in this case series. No patient was diagnosed with neurofibromatosis (Von Recklinghausen disease), gastrinoma, or multiple endocrine neoplasia type 1 (MEN-1). Fifteen (75%) of 20 patients were receiving treatment with proton-pump inhibitors prior to diagnosis.

The urine concentration of 5-hydroxyindoleacetic acid (5-HIAA) was measured in one patient (5%), and it was normal (<6 mg/24 h). Carcinoid syndrome was not observed in any of the patients. The serum levels of chromogranin A were not measured.

A carcinoid tumor diagnosis was established by an endoscopic excision biopsy of 19 (95%) patients, and a histopathological examination of the surgical specimen of one (5%) patient. The characteristics of all 20 patients are summarized in Tables 1 and 2.

TABLE 1. Characteristic of patients and tumors in 20 cases of duodenal carcinoids

Characteristics	Number of patients (%)
Gender	
Male	8 (40)
Female	12 (60)
Average age	66.4 ± 5.8 years old (43 to 88 years old)
Symptoms	
Dyspepsia	10 (50)
Epigastric pain	9 (45)
Weight loss	2 (10)
Vomiting	1 (5)
Location*	
D1	15 (75)
D2	4 (20)
D3	1 (5)
Macroscopic appearance	
Polypoid	10 (50)
Sessile	10 (50)
Number of lesions	
Single	18 (90)
Multiple	2 (10)
Size	Mean 1.1 ± 0.4 cm (0.3 to 6.0 cm)
<1 cm	9 (45)
1 to 2.0 cm	10 (50)
>2 cm	1 (5)

*D1, D2 and D3 = 1st, 2nd and 3rd parts of the duodenum

TABLE 2. Treatment and characteristics of tumors in 20 cases of duodenal carcinoid tumors

Treatment	Number of patients (%)
Endoscopic resection	15 (75); re-resection 3 (15)*
Endoscopic resection plus partial gastrectomy	4(20)
Duodenal segmental resection	1 (5)
Depth of invasion	
Submucosa	16 (80)
T1	1 (5)
T2	1 (5)
T3	1 (5)
Status of resection margin	
Free	16 (80)
Involved	4 (20)
Immunohistochemistry	
Chromogranin A+	15/20 (80)
Neuron-specific enolase+	13/20 (65)
Synaptophysin+	12/20 (60)

* = Involved margin;
+ = Positive

The lesion was located on the first part of the duodenum in 15 (75%) patients, on the second part in 4 (20%) patients, and on the third part in 1 (5%) patient. The lesion was solitary in 18 (90%) patients and multiple in 2 (10%) patients. The macroscopic appearance was polypoid (Figure 1) in 10 (50%) patients, and sessile or intramural in the other 10 (50%) patients. The average tumor size was 1.1 cm ± 0.4 cm (0.3 cm to 6.0 cm). The diameter of the duodenal carcinoid was <1.0 cm in nine (45%) patients, 1.0 cm to 1.9 cm in 10 (50%) patients, and ≥2.0 cm in one (5%) patient. CT of the abdomen and thorax was normal in all patients except one with carcinoid tumor with 6.0 cm in length in which a mass in the third portion of the duodenum was identified. EUS (Figure 2) available recently in our institution was performed only in the last five patients in this series and lymph node involvement was not found.

Nineteen (95%) patients with duodenal carcinoid <2.0 cm of tumor size were initially treated with an endoscopic resection of the duodenal lesion. A negative resection margin was achieved in 16 (80%) patients. The resection margin of the duodenal carcinoid was involved in four (20%) cases, three of which exhibited tumors larger than 1.5 cm in diameter. Three patients were subjected to a second endoscopic resection of the duodenal lesion, and a free resection margin was achieved in all of them. One of these patients was subjected to a partial gastrectomy because this patient was evaluated with the potential risk of duodenal perforation in a new attempt endoscopic resection.

In this series, the lesions that invaded deeply into the muscular layer of the duodenum, the presence of multiple and sessile lesions, the occurrence of involved margins, in which the attempt of a new endoscopic resection was assessed as high risk of duodenal perforation or even technically not feasible, or lesions larger than 2.0 cm in length were removed by surgery.

Four (20%) patients with a previous endoscopic resection of a duodenal carcinoid located on the first part of the duodenum were subjected to a partial gastrectomy to remove the lesion due to the depth of duodenal wall invasion, to the presence of multiple and sessile lesions, or the occurrence of involved margins. One patient with a 6.0 cm duodenal carcinoid located on the third part of the duodenum was subjected to a duodenectomy of the third and fourth parts of the duodenum and duodenojejunal anastomosis. Anatomopathological examination identified carcinoid tumor metastases in the resected lymph nodes, and this patient developed liver metastases. This patient underwent chemoembolization with an intravenous infusion of 5-fluorouracil and adriamycin in the hepatic artery, but he died 6 months after this surgery.

All of the tumors exhibited the typical carcinoid histology: monomorphic islands of small cells with round nuclei surrounded by scarce cytoplasm. The tumor cells exhibited

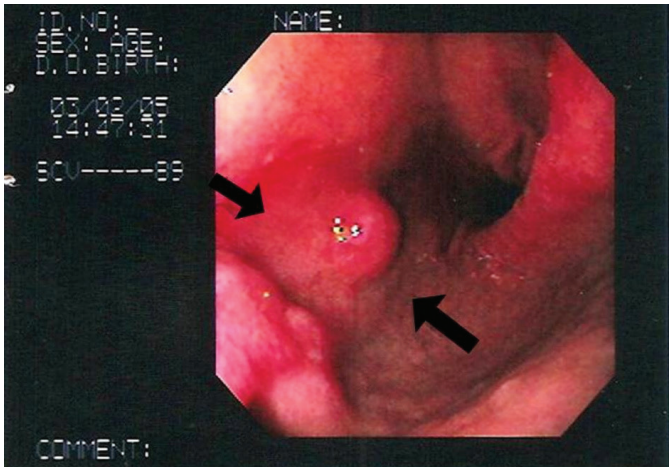

FIGURE 1. Upper gastrointestinal endoscopy photograph showing an intraluminal submucosal polyp (carcinoid tumor) (arrows), in the inferior duodenal bulb

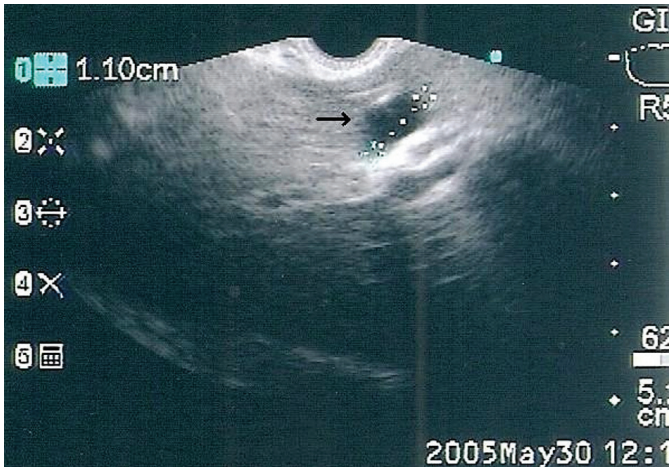

FIGURE 2. Endoscopic ultrasonography (EUS) demonstrating a duodenal hypoechoic mass (carcinoid tumor) (arrow) originating from the deep submucosa

a uniform eosinophilic cytoplasm and a fine granular chromatin pattern with a trabecular or insular arrangement. Mitosis and nuclear atypia were rarely observed (Figures 3 and 4). Carcinoid tumors were limited to the submucosal layer in 16 (80%) patients, and there was invasion of the muscular layer in four (20%) patients; three of these tumors were larger than 1.0 cm in diameter. None of the patients in the present case series exhibited vascular, lymphatic, and/or neural invasion. The mitotic index was less than two mitotic figures per high-power field in 19 (95%) patients, but it was higher in the patient who had a lesion located on the third part of the duodenum.

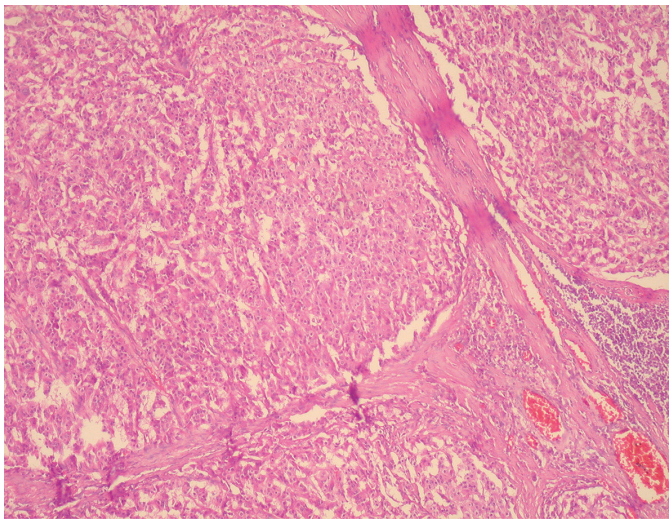

FIGURE 3. Photomicrograph of duodenal carcinoid revealed a well--circumscribed tumor located in the submucosal layer adjacent to the muscularis propria, solid nests of uniform cuboidal cells separated by fine fibrovascular septae, and fascicular proliferative pattern of spindle cells with diffuse perinuclear vacuolization (hematoxylin-eosin, 100x)

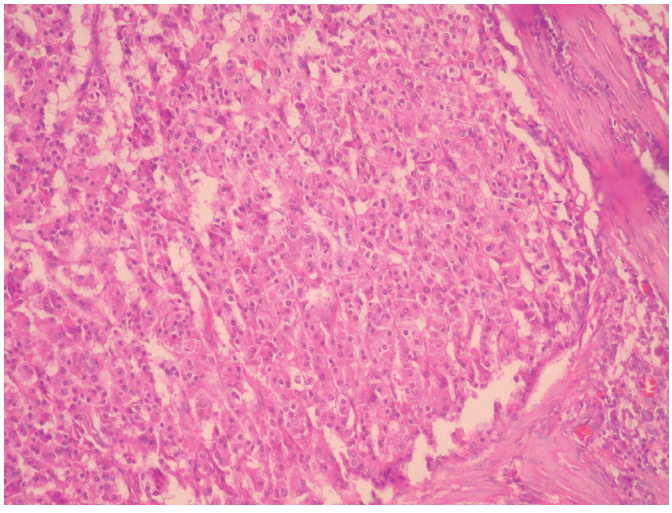

FIGURE 4. Photomicrograph of duodenal carcinoid. The tumor cells had round or oval nuclei and proliferated in a trabecular and microglandular pattern without mitotic figures (hematoxylin-eosin, 200x)

Histological samples from all 20 patients exhibited positive diffuse and occasionally focal immunostaining for chromogranin A (Figure 5), neuron-specific enolase (NSE), and/or synaptophysin. Chromogranin A was observed in 15 (75%) carcinoids, NSE in 13 (65%) carcinoids, and synaptophysin in 12 (60%) carcinoids.

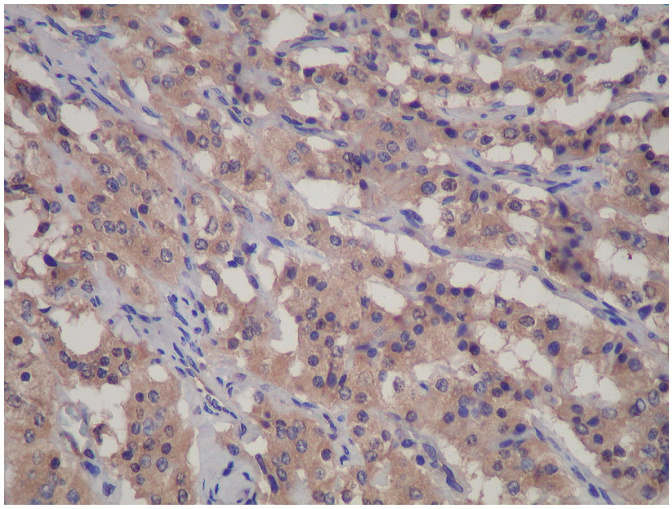

FIGURE 5. Photomicrograph of duodenal carcinoid. A strong cytoplasmatic and nuclear diffuse positivity reactivity for chromogranin A is observed in endocrine neoplastic cells. (immunoperoxidase, 400x)

The average follow-up period was 39.6 months (3.0 to 96.0 months). None of the patients exhibited any other primary neoplasias during the follow-up period. Eleven (55%) patients are still alive without evidence of active disease. Seven (35%) patients died; six (30%) of whom died from causes unrelated to the duodenal carcinoid. Only one (5%) patient died due to an advanced disease and liver metastases, and one (5%) patient did not followed-up.

DISCUSSION

The WHO classifies neuroendocrine ampullary and duodenal tumors into a single category[2], but studies have shown that these tumors are different lesions[7, 12]. The biological behavior of ampullary carcinoids is more aggressive and bears no relationship to the size or mitotic activity of the carcinoids. Ampullary carcinoids express somatostatin, which is seldom observed in duodenal carcinoids[7]. Furthermore, 25% of ampullary, but not duodenal, carcinoid tumors are associated with neurofibromatosis[2, 7, 12].

Duodenal carcinoid tumors are most commonly found on the first part of the duodenum, and their frequency progressively decreases towards the distal parts of the duodenum[2, 3, 7, 22], and this data was confirmed in the present case series. These tumors are indolent, especially when they are small and limited to the submucosa, and the symptoms are generally unspecific[5, 15]. The most common symptoms are abdominal pain, gastroesophageal reflux, dyspepsia, and

gastrointestinal bleeding[8, 12]. The predominant symptoms in the present case series were dyspepsia and epigastric pain. Patients may also be asymptomatic, and the diagnosis can be accidental[5, 8, 15]. The neuroendocrine markers, chromogranin A and 5-HIAA, in serum and urine, respectively, may be useful for the diagnosis[2, 15, 18]. In contrast to the carcinoid tumors of the middle intestine, duodenal carcinoids that originate in the anterior intestine are seldom associated with carcinoid syndrome[2, 18]. Duodenal carcinoids are associated with specific clinical syndromes, such as Zollinger-Ellison syndrome, MEN-1[2, 16], although none were observed in the present case series.

Upper digestive endoscopy is the most useful method for the preoperative diagnosis of these tumors. Duodenal carcinoids appear as intraluminal polypoid lesions or as submucosal masses that are small and spherical with a smooth, or rarely ulcerated, surface. Consequently, deep and multiple biopsies are required to improve the diagnostic index of this neoplasia[1, 8, 15]. CT and magnetic resonance imaging (MRI) do not usually identify small primary tumors, but these techniques may indicate liver and/or mesenteric metastases[11]. A pattern of early and increased contrast-enhancement on CT during the initial venous phase is suggestive of carcinoid tumors[11]. EUS detects small tumors, which appear as homogeneous, well-defined, and mildly hypoechoic or isoechoic masses arising from the second layer[10, 16, 20]. EUS can define the tumor size, level of wall invasion, and the presence of regional lymphatic metastases[11, 13]. Therefore, EUS is necessary to define endoscopic resectability. EUS was performed on the last five patients in the present case series as this technique only recently became available at our institution.

The metastases of carcinoid tumors express a high density of somatostatin-specific receptors, which exhibit a high affinity for the somatostatin analog, octreotide[10]. Therefore, these lesions may be visualized in vivo using labeled octreotide, which detects carcinoid lesions and effectively predicts the therapeutic effect of somatostatin analogs against these tumors[10].

The new classification of carcinoids suggested by the WHO classifies the duodenal, ampullary, and upper jejunal carcinoids as well-differentiated endocrine tumors, well-differentiated endocrine carcinomas, or poorly differentiated endocrine carcinomas[16, 23]. Carcinoid tumors are classified as typical or atypical based on their mitotic activity and the presence of necrosis[15, 23]. Typical carcinoid tumors exhibit fewer than two mitotic figures per 10 high-power fields and an absence of necrotic areas. In contrast, atypical carcinoid tumors exhibit more than two mitotic figures per 10 high-power fields or the presence of necrotic foci. Only one patient in the present case series, whose tumor was larger than 6 cm and located on the third part of the duodenum with T3 invasion, exhibited an atypical carcinoid tumor. This patient developed liver metastases and died 6 months after a duodenal carcinoid resection.

An immunohistochemical investigation is crucial for the accurate diagnosis of duodenal carcinoids. Unspecific neuroendocrine markers, such as chromogranin A, NSE, and synaptophysin, are identified alone or in association in most cases[4, 18, 23]. All patients in the present case series showed immunostaining for at least two of these unspecific neuroendocrine markers. The following hormones are expressed by tumor cells in a decreasing order of frequency: cholecystokinin, gastrin, somatostatin, and serotonin[4]. However, hormonal production is of little or no clinical relevance, except gastrin, which may be clinically significant in a small fraction of patients with duodenal carcinoid and Zollinger-Ellison syndrome[4, 7, 15].

The differential diagnosis of a duodenal carcinoid includes Brunner's gland hamartoma, heterotopic pancreatic and gastric tissues, adenoma, adenocarcinoma, gangliocytic paraganglioma, gastrointestinal stromal tumor (GIST), lymphoid hyperplasia, polyposis syndrome, metastatic disease, and mesenchymal neoplasias, such as neurofibromas and schwannomas[1, 11, 15, 17, 18].

The ideal treatment for duodenal carcinoids is an endoscopic or a radical surgical excision[2, 6, 15]. Tumors in the submucosa outside of the periampullary area that are up to 1 cm without lymph nodes metastases on EUS or CT and no mitotic figures exhibit indolent behavior and no metastases. In these cases, endoscopic removal is the suitable treatment[6, 9, 25]. An endoscopic resection of the lesion was the initial treatment in nine (45%) patients who had a tumor smaller than 1.0 cm. The resection margin was involved by the neoplasm in two of these patients, and both underwent a successful second endoscopic resection.

The treatment of duodenal carcinoid tumors larger than 1.0 cm is controversial. Transduodenal full-thickness resection using laparotomy or laparoscopy[20] is considerate the best treatment for 1.0 cm to 2.0 cm tumors when the tumor invasion of the duodenal wall goes beyond the submucosal layer[15, 16, 18, 19, 26]. Full-thickness excision avoids the possibility of involved margins and the potential for a local relapse. Endoscopic resection of the full thickness of duodenal carcinoid tumors with complete closure of the duodenal defect is also possible[20]. Zyromski et al.[26] conducted a retrospective study with 27 patients with duodenal carcinoid tumors and found that none of the patients with tumors smaller than 2.0 cm treated with local excision exhibited metastases or recurrence. In the present case series, 10 (50%) patients exhibited tumors that measured 1.0 cm to 2.0 cm. The lesions were endoscopically resected in all of these patients, and in 4 out of 10 underwent surgical resection with lymphadenectomy due to the lesion depth assessed by histology and/or EUS or to an involved margin that could not be treated with another endoscopic resection. The lymph nodes were not involved in any of these patients.

Surgical resection may be performed when the carcinoid tumors are larger than 2.0 cm, especially when the mitotic index is higher than two mitotic figures per high-power field, EUS reveals deep wall invasion and/or peritumoral lymph node involvement, and CT and/or MRI indicate the suspicion of lymph node involvement[5, 8, 15, 26]. A full-thickness resection with a regional lymphadenectomy is recommended in these cases. One patient in the present case series underwent a

duodenectomy of the third and fourth parts of the duodenum with a lymphadenectomy due to the presence of a large lesion (6.0 cm) that invaded up to the retroperitoneum and exhibited a high mitotic index. Examination of the lymph nodes that were included in the surgical specimen revealed neoplastic involvement.

Burke et al.[5] identified three pathological characteristics of duodenal carcinoid tumors that are independent risk factors for metastases: invasion of the muscular layer, a size greater than 2.0 cm, and the presence of more than two mitotic figures per high-power field.

In cases of liver metastases, a surgical resection and/or cytoreductive techniques, such as radiofrequency ablation and chemoembolization, may improve carcinoid syndrome symptoms that are mediated by hormones produced by the tumor, in order to improve the quality of life and increase survival[5, 8, 15, 18]. Somatostatin analogs may induce symptomatic and biochemical responses that stabilize the disease and reduce the growth of metastases via cytostatic effects[2, 8, 15, 18].

Serum chromogranin A reflects the tumor load of carcinoids, and it is a useful tumor marker for the monitoring of disease response to treatment and disease progression, particularly relapse. Chromogranin A is also an independent predictor of survival in patients with gastrointestinal carcinoids[8, 16, 24].

The global survival of patients with duodenal carcinoids is excellent in 80% to 90% of cases with well-differentiated tumors[2, 14, 15, 18, 24]; however, survival is lower among patients with less differentiated neoplasms. Most patients with duodenal carcinoid tumors die from another cause[14, 18, 24], a fact that was observed in the present case series and confirms the indolent nature of this disease.

CONCLUSION

In conclusion, duodenal carcinoids are rare and indolent tumors usually associated with a benign progression. Duodenoscopy, computerized tomography, and endoscopic ultrasound should be performed to evaluate the tumor size, the level of wall invasion, and the presence of regional or distant lymphatic metastases. Endoscopic removal of tumors smaller than 1.0 cm without periampullary localization or evidence of muscular propria layer invasion assessed by histology and/or endocopic ultrasound is recommended. The endoscopic resection with a carcinoid tumor size between 1.0 cm and 2.0 cm can be incomplete and require new endoscopic resection or even surgical removal. Duodenal carcinoid larger than 2.0 cm require full-thickness resection and concomitant lymphadenectomy.

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RESUMO – ***Contexto*** - Carcinoides duodenais são extremamente raros e as características e o comportamento biológico dessa neoplasia permanecem indefinidos. ***Objetivo*** - Analisar as características clinicopatológicas de doentes com carcinoide duodenal ressecado. ***Métodos*** - Vinte doentes (12 mulheres e 8 homens) foram estudados. A média de idade dos doentes foi de 66,4 ± 5,8 anos (43 a 88 anos). Os dados do quadro clínico, diagnóstico, tratamento e prognóstico dos doentes com tumor carcinoide do duodeno submetidos a ressecção da lesão no período de 18 anos (1993-2011) foram analisados. ***Resultados*** - Os sintomas mais frequentes foram dispepsia (50%) e epigastralgia (45%), seguidos por perda de peso (10%) e vômitos (5%). Não foram observados doentes com síndrome carcinoide. A lesão estava localizada na primeira porção do duodeno em 15 (75%) pacientes, na segunda porção em 4 (20%) e na terceira porção em 1 (5%). O diagnóstico de tumor carcinoide foi estabelecido pela biopsia endoscópica excisional em 19 (95%) pacientes e pelo exame histopatológico da peça cirúrgica em um (5%). O tamanho médio dos tumores foi de 1,1 cm ± 0,4 cm (0,3 cm a 6,0 cm). Dezenove (95%) doentes foram tratados, inicialmente, por ressecção endoscópica da lesão duodenal e um (5%) com lesão na terceira porção duodenal foi submetido a duodenectomia da terceira e quarta porções do duodeno e duodenojejunoanastomose. A margem de ressecção do carcinoide duodenal estava comprometida em quatro (20%) casos e em quatro (20%) pacientes foi realizada gastrectomia parcial para retirada completa da lesão. O tumor estava limitado à camada submucosa em 16 (80%) casos e penetrava a camada muscular própria em 4 (20%). Todos os pacientes apresentaram imunomarcação positiva para cromogranina A, enolase neurônio-específica ou sinaptofisina. A média do período de seguimento foi de 39,6 meses (3 a 96 meses). Dos 20 casos desta série, 12 (60%) permanecem vivos e sem evidência de doença ativa e apenas 1 (5,0%) faleceu por metástase hepática do carcinoide duodenal. ***Conclusões*** – Carcinoides duodenais são tumores raros e indolentes normalmente associados a bom prognóstico. Duodenoscopia, tomografia computadorizada e ultrassonografia endoscópica devem ser realizadas para avaliar o tamanho do tumor, o nível de invasão da parede e a presença de metástases linfáticas regionais e/ou distantes. Remoção endoscópica de tumores menores que 1,0 cm, sem localização periampolar ou evidência de invasão da camada muscular própria avaliada pela histologia e/ou ultrassonografia endoscópica é recomendada. A ressecção endoscópica de tumor carcinoide com tamanho entre 1,0 cm e 2,0 cm pode ser incompleta e requerer nova ressecção endoscópica ou mesmo remoção cirúrgica. Carcinoides duodenais maiores que 2,0 cm necessitam de ressecção com espessura total e linfadenectomia concomitante.

DESCRITORES – Neoplasias intestinais. Duodeno. Tumor carcinoide. Carcinoma neuroendócrino. Apudoma.

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