Case Report
Primary retroperitoneal neuroendocrine tumor with concomitant pelvic leiomyoma: a case presentation

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Abstract: Background: Neuroendocrine carcinoma is an aggressive neoplasm arising most commonly from the gastrointestinal tract and the bronchopulmonary tree. We report here a rare case of neuroendocrine tumor (NET) in the retroperitoneal cavity but is not commonly found in the pancreas of retroperitoneum. Case presentation: We report the case of a 54-year-old Chinese male with retroperitoneal neuroendocrine carcinoma and pelvic leiomyoma that initially presented as a large retroperitoneal mass. CT revealed abnormal circular lesions in the horizontal segment of the duodenum (anterior abdominal aorta) and a pelvic space-occupying lesion. The neuroendocrine carcinoma was successfully excised, along with a pelvic leiomyoma. The retroperitoneal tumor was found to be approximately 3.5 cm × 4 cm × 3.5 cm and was located in the retroperitoneal cavity approximately 10 cm from the Treitz ligament and was closely adherent to the jejunum. In addition, an oval mass approximately 8 cm × 6 cm in size was also observed in the pelvic cavity to the left of the bladder. Histopathological and immunohistochemical examinations revealed that the retroperitoneal tumor was a well-differentiated neuroendocrine tumor and that the pelvic mass was a leiomyoma. Conclusions: Histological and immunohistochemical examinations of neuroendocrine tumors are essential for diagnosis. Surgical excision of primary tumors is still one of the main treatments for neuroendocrine tumors. Further studies are needed to elucidate the efficacy of chemotherapy for neuroendocrine tumors in patients with typical and atypical presentations.

Keywords: Neuroendocrine tumor, retroperitoneum, prognosis

Introduction

Neuroendocrine tumors are heterogeneous in nature and have an increasing incidence [1]. They arise from the cells of the diffuse endocrine system and have various clinical and biological behaviours depending on their location, degree of differentiation and secretory activity [2]. They are most commonly found in the gastrointestinal tract and account for 30% of small bowel (SB) neoplasms [3], 17% of rectal tumors [4] and 6% of pancreatic tumors [5]. However, neuroendocrine tumors are also reported in other organs, such as the bladder [6], bronchial tree [7], and thyroid [8]. We report the case of a middle-aged male who was diagnosed with and treated for a neuroendocrine tumor in our hospital. The reported tumor of the retroperitoneal organs is rather rare but is not commonly found in the pancreas of retroperitoneum. The purpose of this case report is to highlight the unusual growth pattern and location of a neuroendocrine tumor and to compare this case with the pertinent English language literature.

Case report

A 54-year-old man from Suzhou, China, was admitted to the Affiliated Suzhou Hospital of Nanjing Medical University due to abdominal discomfort of unknown cause for several months in 2017.

On admission, physical examination showed mild umbilical tenderness on deep palpation. Laboratory examinations showed that the levels of tumor markers (CA199, CA125, CEA and AFP) were normal, and routine blood parameters of liver and kidney function were normal.

After admission, abdominal CT and gastroscopy were performed. CT revealed abnormal circular lesions in the horizontal segment of the duodenum (anterior abdominal aorta), a pelvic space
Retroperitoneal neuroendocrine tumor occupying lesion, and small renal and liver cysts (Figure 1A and 1B). Gastroscopy showed superficial gastritis and a submucosal eminence lesion on the descending part of the duodenum.

As primary neuroendocrine carcinoma in the retroperitoneal organs is extremely uncommon, this mass was initially suspected to be stromal tumor or fibroma. Three days after admission, the patient underwent surgical intervention.

The tumors in the retroperitoneum and pelvic cavity were successfully resected. During the surgery, the retroperitoneal tumor was found to be an approximately 3.5 cm × 4 cm × 3.5 cm, pale and hard encapsulated mass with poor mobility. The retroperitoneal tumor was approximately 10 cm from the Treitz ligament and was closely adherent to the jejunum. In addition, an oval mass measuring approximately 8 cm × 6 cm was also observed in the pelvic cavity to the left of the bladder. The oval mass was pale, hard and smooth, with moderate mobility, and no capsule.

Intra-operative pathological examination showed that the retroperitoneal tumor was a neuroendocrine carcinoma, and the pelvic mass was a leiomyoma. The tumor tissues were further processed for post-operative histopathological examination and immunohistochemistry.

Microscopically, the neuroendocrine carcinoma had immediately sized cells with round nuclei, granular cytoplasm, and coarse clustered chromatin, and karyokinesis was found in less than 2 of 10 cells examined per high power field (2/10), but the typical rosette appearance was not observed, and there was no evidence of necrosis or vascular or perineural invasion. Immunohistochemistry showed that the neuroendocrine carcinoma cells were positive for CgA, Syn, Ki-67, and CD56 but negative for CEA, CD34, and CD117. The Ki-67-positive cells accounted for less than 2% of the stained carcinoma cells (Figure 2). The plasma serotonin and 24-h urine 5-hydroxyindoleacetic acid (5-HIAA) levels were within the normal range. On the basis of histopathological and immunohistochemical findings, the retroperitoneal tumor was definitively diagnosed as a well-differentiated neuroendocrine tumor of the retroperitoneum.

After surgery, adjunctive treatment was not recommended, and the patient was discharged 9 days after surgery. He received regular follow-
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up, and recurrence and metastasis were not observed.

Discussion

In the literature, neuroendocrine tumors (NETs), previously known as carcinoid tumors, were classified by the World Health Organization (WHO) in 2000 [9]. According to the WHO classification, these tumors can be divided into three grades based on their biological and histological characteristics [2]: well-differentiated NETs, moderately differentiated NETs and slightly differentiated NETs.

NETs have been reported in different organs and are mainly found in the gastrointestinal tract. The most common site of NET development is the intestine 60 cm distal to the distal ileum, and NETs account for 25% of all intestinal tumors [10]. Retroperitoneal neuroendocrine tumors have been found in the pancreas [5], duodenum [11], common bile duct [12] and kidney [13]. However, the neuroendocrine tumors of retroperitoneal organs outside the above normal sites as the primary tumor in non-metastatic conditions have rarely been described in the literature. Polikarpova SB et al. showed retroperitoneal neuroendocrine tumors are more common in the elderly, and the clinical symptoms are usually mild. Generally, patients have no manifestations of carcinoid syndrome [14]. These tumors evolve asymptptomatically and are often diagnosed incidentally on an abdominal imaging examination, such as ultrasonography (US) or and computed tomography (CT).

In this report, the patient was 54 years old and was admitted with a several-month history of abdominal discomfort. Abdominal CT showed a retroperitoneal mass between the duodenal wall and the head of the pancreas. Initially, neuroendocrine carcinoma was not considered due to its rarity. The patient underwent surgical intervention, and metastasis was not found in the abdominal cavity. In the operation, we found that this retroperitoneal neuroendocrine tumor did not arise from the small bowel or pancreas, but only adhered to the jejunum. Relatively few cases present with no apparent primary lesion in retroperitoneum.

As in this patient, retroperitoneal neuroendocrine tumors are often non-secreting and have a low incidence of carcinoid syndrome in patients with limited or local forms, and there is a relatively high incidence of carcinoid syndrome in those with liver metastases [14].

Although retroperitoneal neuroendocrine tumors can be found on pre-operative abdominal imaging examinations such as abdominal ultrasonography, CT, or endoscopic ultrasonography, the confirmatory diagnosis is still made based on post-operative histological and immunohistochemical examinations [15].

The treatment for neuroendocrine tumors of the retroperitoneum depends on the location and extent of the tumor. Surgical resection is the primary treatment for retroperitoneal NETs and offers a chance of curing the disease. Non-randomized studies have demonstrated survival benefits from surgery in patients with NETs [16].

Pathological examination showed that the tumor cells were well differentiated. Surgical intervention alone was elected, and the involved jejunum was also resected during surgery.

Post-surgical histological and immunohistochemical examinations for neuroendocrine markers (CgA, Syn, NSE, and CD56) are critical for a confirmatory diagnosis [17].

After surgery, adjunctive therapy was not recommended for this patient due to the absence of metastasis. The available studies indicate that neuroendocrine tumors of the retroperitoneum in most cases are slowly growing and have a limited risk for local invasion and metastasis.

Currently, there are no histological criteria for assessment of the malignant potential of neuroendocrine tumors of the retroperitoneum. Nevertheless, tumor size seems to correlate with the risk for metastasis [15]. On the other hand, lymphovascular invasion is common in these tumors, and quantitative assessment of Ki-67-reactive cells may allow an assessment of prognosis and survival [18].

However, in general, most clinicians agree that curative surgical resection may be a predictor of prognosis in patients with neuroendocrine tumors [16, 17]. Chemotherapy is usually used as first-line therapy for unresectable, poorly differentiated NETs. In our case, lymph node
metastasis was not observed, and thus this patient was only treated surgically without post-operative adjunctive therapy.

In conclusion, extended local excision of the primary tumor is the treatment of choice for local retroperitoneal NETs, but the role of post-operative chemotherapy is still undetermined [19]. Further studies are needed to elucidate the efficacy of chemotherapy for patients with neuroendocrine tumors with typical and atypical presentations.

Conclusions

To our knowledge, excision of primary tumors is one of the main treatments for neuroendocrine tumors. The approach is usually accompanied by lymphadenectomy or regional irradiation in patients with positive lymph nodes or by selective lymph node dissection in patients with negative lymph nodes. This surgical procedure reduces recurrence rates following surgery or at least prolongs the time to recurrence. However, the value of chemotherapy is still uncertain. Thus, our patient was treated with surgical intervention alone. We suggest that further studies are needed to elucidate the true curative potential of chemotherapy for patients with neuroendocrine tumors with typical and atypical presentations.

Acknowledgements

Written informed consent for publication of the clinical details and any accompanying images was obtained from the patient. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Disclosure of conflict of interest

None.

Abbreviations

CgA, Chromogranin A; Syn, synaptophysin; NSE, neuron-specific enolase; Ki-67, antigen Ki67; CD, cluster differentiation; NET, neuroendocrine tumor.

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