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A case report of neuroendocrine tumor of extrahepatic biliary tract

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ABSTRACT

Background: A rare form of tumor is a neuroendocrine tumor (NET). According to the 2015 National Comprehensive Cancer Network (NCCN) Clinical Practice Guidelines in Oncology, all endocrine tumors of the digestive system should be treated with neuroendocrine tumors of the gastrointestinal system. The incidence of NET is 2.5 cases per 100,000 people annually on average. Since NET has a variety of clinical symptoms, it can be divided into functionally active and inactive types. This study aimed to present a neuroendocrine tumor of the extrahepatic biliary tract.

Case presentation: An 18-year-old female with a lump in the upper right abdomen with intermittent pain that had been felt in the last 8 months. There was evidence of previous weight loss and an easy appetite. Physical inspection revealed a mass with a fixed, smooth, flat edge and solid consistency. A vesica felea enlargement and a normal wall were discovered on the CT scan. Stone and sludge were not detected. The choleducus and cystic duct were normal-looking, and a 0.5 cm spherical stone was present. Bile fluid was found to move smoothly from the ductus cysticus to the distal and into the ampulla of Vater. Our patient was diagnosed early with an intra-abdominal tumor. An intraoperative cystic tumor measuring 9x7x8 cm and attached to the duodenum was discovered at the pancreatic cap. The pancreatic uncinata process underwent tumor excision. The patient was managed post-op in a stable condition, and the tumor's histology revealed a carcinoid tumor.

Conclusion: NET is a rare type of tumor, but two third of it occurs in the gastrointestinal system. It is hard to detect because of its heterogeneous nature and various clinical presentation. Surgery is the first-line therapy in this disease, followed by chemotherapy, radiotherapy, and tumor debulking as palliative management.

Keywords: neuroendocrine tumor, carcinoid tumor, extrahepatic biliary tract.

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INTRODUCTION

Neuroendocrine tumor (NET) is a rare type of tumor. It has other names, including carcinoid tumor, gastro-entero-pancreatic tumor (GEP tumor), islet cell tumor, endocrine tumor, and neuroendocrine carcinoma. "Carcinoid" term was used earlier, and it indicated a type of malignant tumor arising in the small intestine that formed metastases but had a slow-growing ability. It was introduced by Oberndorfer in 1902. However, the term "neuroendocrine tumor of the gastrointestinal and pancreaticobiliary system" was preferred to refer to all endocrine tumors of the digestive system because they all originate from the diffuse neuroendocrine system, according to the National Comprehensive Cancer Network's (NCCN) Clinical Practice Guidelines in Oncology published in 2015.^{1,2}

NET are found mostly in the gastrointestinal tract; approximately two-

thirds of NETs at any location are found in the gastrointestinal (GI) tract, one quarter occurs in the lung, with the remainder arising in other endocrine tissues. According to the main tumor's origin, the type of neuroendocrine cells involved, and the pathological characteristics, NET of the pancreaticobiliary system are heterogeneous with varying biological and clinical activity. Eastern and Western people have varied GI tract NET dispersion patterns. Most tumors among American patients were discovered in the small intestine (38%), followed by the rectum (34%), colon (16%), stomach (11%), and an undetermined location (1%). The incidence is 2.5 cases per 100,000 people annually on average. There have been fewer than 200 reported occurrences of NET since 1961, according to Pliz First. Compared to other ethnic groups, African-Americans have a higher prevalence of NET. According

to Surveillance Epidemiology and End Results (SEER) data, the incidence of NET among white men is 2.47 per 10,000 people, whereas it is 4.48 per 10,000 people among African Americans. 64 years old is the typical diagnosis age for NET.²⁻⁴

The clinical manifestations of NET are various. Functionally active NET presents with specific hormone overproduction syndromes, such as insulinoma, gastrinoma, and glucagonoma syndrome. Functionally inactive NET typically presents with abdominal cramps (68%-78%), weight loss (32%-50%), jaundice because of biliary obstruction or metastases process (21%-50%), or nausea and vomiting (36%). Metastases are found in nearly 19%-22% of patients. Nonfunctional gastropancreatic NET usually shows a more severe clinical presentation, so the frequency of metastases found when the NET is first diagnosed is higher. The most common

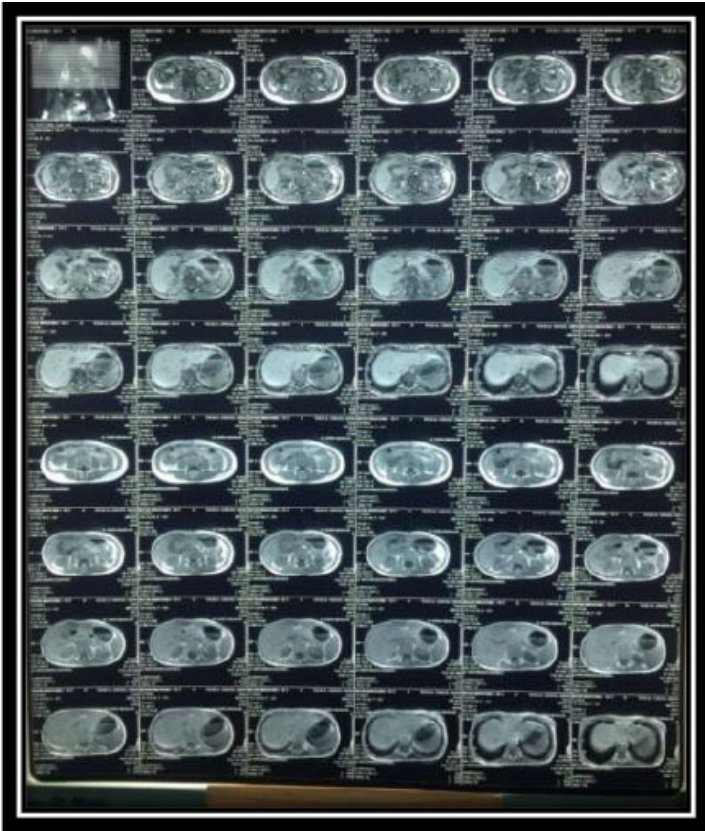


Figure 1. CT scan result of the patient found an enlargement on the vesica felea.

Figure 2. CT scan result, which concludes the diagnosis of cholelithiasis.

metastases site is the liver.³ This study aimed to present a NET of the extrahepatic biliary tract.

CASE PRESENTATION

An 18-year-old female came with a chief complaint of a lump in the upper right abdomen that had been felt for the last 8 months. The initial lump was small and was getting bigger. The patient also complained of pain in the lump occasionally. A history of weight loss was found. A history of loss of appetite and easily satiated was found. History of nausea, bloody stool, and light-colored stool was denied. A history of fever was found occasionally, often followed by pain on the lump. History of trauma was denied.

Upon physical inspection, we discovered a fixed solid mass with a smooth surface, a flat edge, and no signs of pain. A leucocyte count of 5,400 cells/ μ l, total bilirubin level of 0.69 mg/dl, direct bilirubin level of 0.30 mg/dl, and indirect bilirubin level of 0.39 mg/dl were all found

in the blood examination. A vesica felea enlargement and a regular and normal wall were discovered on the CT scan (Figure 1). Stone and sludge were not detected. The choleducus and cystic duct were normal-looking, and a 0.5 cm spherical stone was present. Bile fluid moved smoothly from the ductus cystic to the distal and into the ampulla of Vater. According to a CT scan, the finding conclusion of this case was cholelithiasis (Figure 2). Our patient was diagnosed early with an intra-abdominal tumor. When an intraoperative cystic tumor measuring 9x7x8 cm was discovered in the pancreas cap and stuck to the duodenum, we performed tumor excision on the pancreatic uncinata process (Figure 3).

After surgery, the patient was given regular care and histology results showed that the tumor was a carcinoid tumor (NET). During post-operative care, there was not any clinical complaint found. The patient was discharged on day 6. The patient came to the surgery polyclinic on days 10 and 15 and was in good condition.

DISCUSSION

NET is a type of tumor that contains multipotential cells that can secrete various hormones such as vasoactive, serotonin, gastrin, somatostatin, glucagon, and insulin. NET is a rare case. This tumor is symptomatic, and the clinical manifestation arises along with the development of tumor size, the invasive properties of the tumor, the metastases process, and the released hormone by the tumor. NET is hard to detect preoperatively. Functionally inactive patients do not typically present clinical syndrome of hormone overproduction. Patients are usually diagnosed during examination when they experience unexplained complaints of upper abdominal jaundice because of a big tumor in the pancreas cap or recurrent abdominal cramps because of pseudo-obstruct by NET growth in the jejunum and ileum.^{3,5,6}

One study showed that NET is more dominant in women (61.5%) than men. Another study showed that the average age of patients when the tumor was first

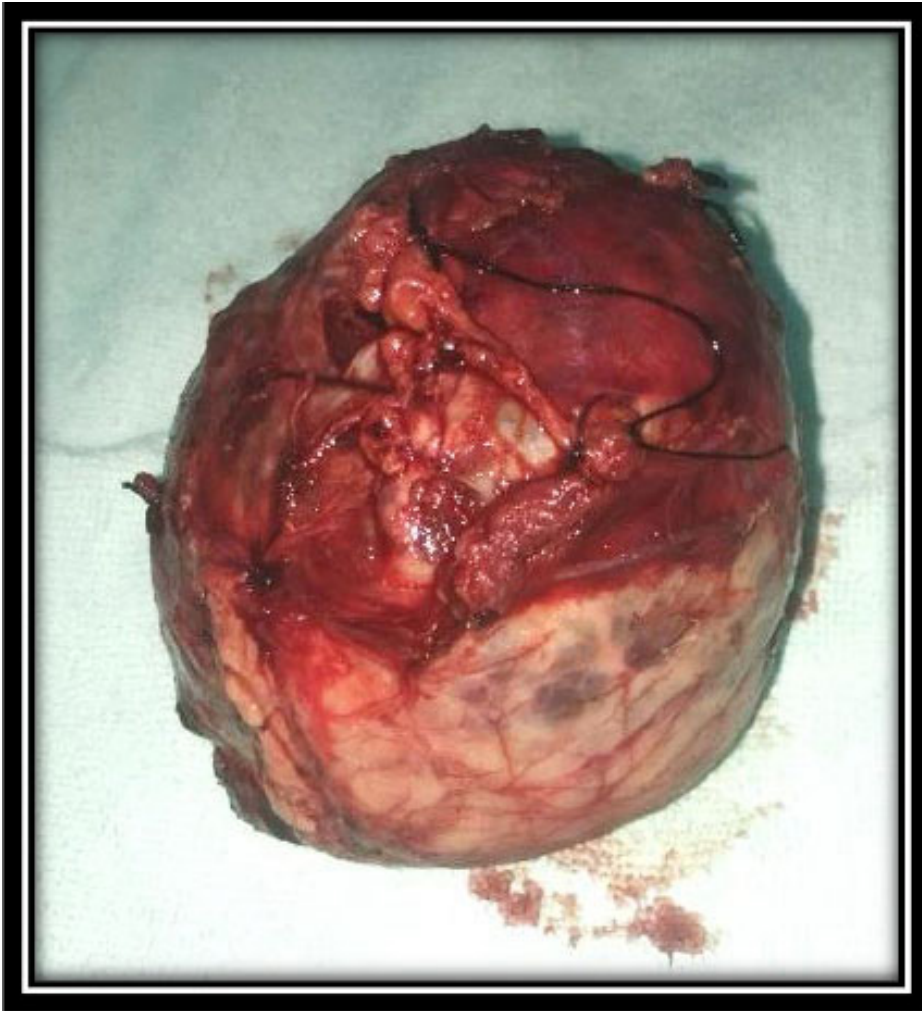


Figure 3. An intraoperative cystic tumor in the pancreas cap with a cyst size of 9x7x8 cm.

diagnosed was 64 years old. It is more common in the black-colored skin population than any other ethnicities. Except for insulinoma, most NETs are malignant. However, these tumors have low mitotic activity and grow slowly. NET often metastases to the liver, followed by lymph nodes, bone, lungs, and brain, and rarely to other organ.^{3,7}

Our patient underwent a CT scan to evaluate the pancreaticobiliary system, and the result indicated cholelithiasis. Because of its heterogeneous nature, it is hard to diagnose and make an algorithm for the NET. However, a CT scan is the most common initial work-up for patients with suspected NET. In diagnosing it, we need an imaging strategy to detect the mass. A combination of anatomic and functional imaging has been proven accurate. Anatomic imaging includes

CT scans, magnetic resonance imaging (MRI), transabdominal ultrasonography, GI endoscopy, and ultrasonographic endoscopy. MRI has a sensitivity of 85%, whereas transabdominal USG is not sensitive enough to detect primary tumors; it has a sensitivity of 33%. Functional imaging is important to evaluate the stage of the tumor and the metastases process. It targets to evaluate the receptors, uptake pathways, and the metabolism process of NET. The available tests include somatostatin receptor scintigraphy (SRS), positron emission tomography (PET), and tumor biopsy. Serologic tests are usually done, especially in functionally active patients, such as chromogranin A, CA19-9, and carcinoembryonic antigen, along with other laboratory tests for specific hormones according to patients' clinical presentation.^{3,8-10}

Our patient underwent exploration laparotomy surgery and found a cystic tumor with a size of 9x8x7 cm stuck to the duodenum. Then we did tumor excision on the uncinata process of the pancreas. The preferred therapy for resectable NET patients is surgery. However, since the high rate of metastases, additional palliative management, such as tumor debulking, chemotherapy, and radiotherapy, is often needed. Chemotherapy is not the first-line therapy of NET, because the used regimens (streptozocin and 5-fluorouracil [5-FU]) are not as effective as surgery. The use of somatostatin analogs also becomes the mainstay therapy for most symptomatic patients.^{3,11}

The more aggressive the surgical intervention and using somatostatin analogs, the better the prognosis. Nearly all patients with metastatic disease will experience a recurrence in 7 years, even if the previous metastatic disease was completely cured. The favorable prognostic factor for patients with NET includes resected primary tumor, the absence of liver metastases, or aggressive management for liver metastases. Whereas the bad indicators are the involvement of lymph nodes, metastases were detected when the tumor first diagnosed, and the presence of lymph and vascular invasion of the tumor.^{3,11}

CONCLUSION

NET is a rare type of tumor, but two-thirds of it occurs in the GI system. It is hard to detect because of its heterogeneous nature and various clinical presentation. Surgery is the first-line therapy in this disease, followed by chemotherapy, radiotherapy, and tumor debulking as palliative management. Using somatostatin analogs has also become a mainstay therapy for functionally active patients.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient to publish this case report and accompanying images.

COMPETING INTERESTS

The authors declare that they have no competing interests.

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AUTHOR CONTRIBUTIONS

All authors have contributed equally to this manuscript.

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