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An Unusual Presentation of Pancreatic Neuroendocrine Carcinoma in a Young Woman: A Case Report

Iman Hakim Wicaksana[1*], Suryo Wahyu Raharjo[2]

[1]Resident of Surgery, Faculty of Medicine, Universitas Sebelas Maret/Dr. Moewardi General Hospital, Surakarta, Indonesia

[2]Department of Digestive Surgery, Faculty of Medicine, Universitas Sebelas Maret/Dr. Moewardi General Hospital, Surakarta, Indonesia

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***Corresponding author:**
Iman Hakim Wicaksana

E-mail address:
hakimwicaksana@gmail.com

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A B S T R A C T

Introduction: Pancreatic neuroendocrine tumors (PNETs) are rare neoplasms arising from the endocrine cells of the pancreas. They are typically diagnosed in older adults, with a peak incidence between 70 and 74 years. PNETs in young adults are uncommon and often present with non-specific symptoms, leading to delays in diagnosis and treatment. **Case presentation:** We report the case of a 22-year-old woman who presented with a large, non-functional PNET located in the tail of the pancreas. The patient initially complained of vague abdominal discomfort, nausea, and vomiting. Imaging studies revealed a large, heterogeneous mass in the left upper quadrant. The patient underwent a distal pancreatectomy, and histopathological examination confirmed the diagnosis of a poorly differentiated pancreatic neuroendocrine carcinoma. **Conclusion:** This case highlights the challenges in diagnosing PNETs in young adults. Clinicians should maintain a high index of suspicion for PNETs in young patients presenting with abdominal symptoms, even in the absence of classic hormonal syndromes. Early diagnosis and surgical intervention are crucial for improving outcomes in these patients.

1. Introduction

Pancreatic neuroendocrine tumors (PNETs) are a rare and fascinating group of neoplasms that originate from the endocrine cells of the pancreas. These tumors, accounting for approximately 1-2% of all pancreatic tumors, arise from the islet cells of Langerhans, the hormone-producing factories responsible for regulating various metabolic and digestive processes. The rarity of PNETs, combined with their often subtle and non-specific clinical presentation, makes them a diagnostic challenge for clinicians. PNETs are broadly classified into two main categories based on their hormonal activity: functional and non-functional. Functional PNETs (FPNETs) are characterized by the excessive secretion of hormones, leading to distinct clinical syndromes such as insulinoma (insulin hypersecretion), gastrinoma (gastrin hypersecretion), glucagonoma (glucagon hypersecretion), and VIPoma (vasoactive intestinal peptide hypersecretion). These syndromes, though rare, provide valuable clues for early diagnosis and intervention. Non-functional PNETs (NFPNETs), on the other hand, do not secrete excessive amounts of hormones and often remain clinically silent until they reach a significant size or metastasize to distant organs. The diagnosis of NFPNETs is often incidental, discovered during imaging studies performed for unrelated reasons. The absence of specific hormonal syndromes makes the diagnosis of NFPNETs particularly challenging, as their symptoms can mimic

those of more common gastrointestinal disorders. The incidence of PNETs is estimated to be around 2.2 cases per 100,000 individuals, with a slight predominance in females. The peak incidence occurs in the seventh decade of life, making PNETs relatively uncommon in young adults. This age-related distribution further contributes to the diagnostic challenge in younger individuals, as clinicians may not initially consider PNETs in their differential diagnosis.[1-4]

The clinical presentation of PNETs is as diverse as their hormonal activity and anatomical location. FPNETs typically manifest with symptoms directly related to hormone hypersecretion, such as hypoglycemia in insulinomas, peptic ulcer disease in gastrinomas, and hyperglycemia in glucagonomas. These hormonal syndromes, though often dramatic in their presentation, can be managed effectively with medical and surgical interventions. NFPNETs, in contrast, are often asymptomatic until they reach a substantial size or spread beyond the pancreas. Symptoms such as abdominal pain, jaundice, and weight loss may develop as the tumor encroaches on surrounding structures or metastasizes to distant organs. The insidious onset of symptoms and the lack of specific hormonal markers make the diagnosis of NFPNETs particularly challenging. The diagnosis of PNETs, particularly in young adults, is often a complex and multi-faceted process. The non-specific nature of symptoms, coupled with the rarity of these tumors, can lead to delays in diagnosis and treatment. A high index of suspicion is crucial for clinicians evaluating young patients with abdominal symptoms, even in the absence of classic hormonal syndromes. Imaging studies, such as computed tomography (CT) and magnetic resonance imaging (MRI), play a pivotal role in the detection and characterization of PNETs. These modalities provide detailed anatomical information about the tumor's size, location, and relationship to surrounding structures. Functional imaging techniques, such as somatostatin receptor scintigraphy (SRS), can further aid in identifying hormonally active tumors.[5-7]

The treatment of PNETs requires a multidisciplinary approach, involving gastroenterologists, surgeons, oncologists, and radiologists. Surgical resection remains the cornerstone of treatment for localized PNETs, offering the potential for cure in selected patients. Adjuvant therapies, such as chemotherapy, radiation therapy, and targeted therapies, may be considered for patients with advanced or metastatic disease. Early diagnosis and prompt intervention are critical for improving outcomes in patients with PNETs. Delays in diagnosis can lead to tumor progression, metastasis, and a worse prognosis. Clinicians must maintain a high index of suspicion for PNETs in young adults presenting with abdominal symptoms, even in the absence of classic hormonal syndromes.[8-10] In this case report, we present the unusual case of a 22-year-old woman who presented with a large, non-functional PNET located in the tail of the pancreas.

2. Case Presentation

This report details the case of a 22-year-old woman who presented with left upper quadrant abdominal pain, a chief complaint that prompted a thorough investigation leading to the diagnosis of a poorly differentiated pancreatic neuroendocrine carcinoma. The patient's history of present illness revealed a 3-month progression of vague abdominal discomfort accompanied by nausea and vomiting. This discomfort had gradually intensified, culminating in a palpable mass in the left upper quadrant. Additionally, she reported a significant weight loss of 10 kg within the past month. Importantly, there was no history of fever, jaundice, or changes in bowel habits, which are often associated with pancreatic pathology. Her past medical history was unremarkable, with no known diagnoses of diabetes, hypertension, or a family history of pancreatic tumors. The patient reported taking no regular medications and had no known drug allergies. Social history indicated a non-smoker with no history of alcohol or illicit drug use. Upon examination, the patient appeared to be in good health, alert, and oriented. Her vital signs were within

normal limits: blood pressure 130/88 mmHg, heart rate 81 bpm, respiratory rate 20 breaths/min, and temperature 36.3°C. A head, eyes, ears, nose, and throat (HEENT) examination revealed no abnormalities, including no scleral icterus (yellowing of the eyes), conjunctival pallor (pale conjunctiva), or lymphadenopathy (swollen lymph nodes). Respiratory examination was clear bilaterally, with no signs of respiratory distress such as wheezes, rales, or rhonchi. Cardiovascular examination revealed a regular heart rate and rhythm, with no murmurs, gallops, or rubs. Gastrointestinal examination was significant for a soft, non-tender, palpable mass in the left upper quadrant. There was no evidence of hepatosplenomegaly (enlarged liver or spleen), and bowel sounds were normal. Examination of the extremities showed no edema (swelling), normal capillary refill, and no clubbing or cyanosis (bluish discoloration) of the fingers. A neurological examination demonstrated grossly intact function with no focal deficits. Complete blood count revealed a white blood cell count of 9.4 x 10^9/L (normal range: 4.5-14.5 x 10^9/L), hemoglobin of 13.7 g/dL (normal range: 12.3-15.3 g/dL), hematocrit of 42% (normal range: 33-45%), and a platelet count of 228 x 10^9/L (normal range: 150-450 x 10^9/L). These findings indicated no significant hematological abnormalities. Liver function tests showed aspartate aminotransferase (AST) of 22 U/L (normal range: 0-40 U/L), alanine aminotransferase (ALT) of 28 U/L (normal range: 0-40 U/L), alkaline phosphatase of 80 U/L (normal range: 40-120 U/L), and total bilirubin of 1.0 mg/dL (normal range: 0.1-1.2 mg/dL). These results were within normal limits, suggesting no significant liver dysfunction. Renal function tests demonstrated creatinine of 0.6 mg/dL (normal range: 0.9-1.3 mg/dL) and urea of 28 mg/dL (normal range: <50 mg/dL), indicating normal kidney function. Electrolytes were also within normal limits. Tumor markers, including carcinoembryonic antigen (CEA) at 2.0 ng/mL (normal range: <5 ng/mL) and carbohydrate antigen 19-9 (CA 19-9) at 15 U/mL (normal range: <37 U/mL), were not significantly elevated. Coagulation studies, including prothrombin time (PT) at 12.3 seconds (normal range: 10-15 seconds), activated partial thromboplastin time (APTT) at 35.9 seconds (normal range: 20-40 seconds), and international normalized ratio (INR) at 0.87, were all within the normal range. Serology for hepatitis B surface antigen (HBsAg) was non-reactive. A chest X-ray was performed to assess for potential metastatic disease and showed no evidence of pulmonary metastases. An abdominal CT scan was crucial in identifying a 9.7 x 11.4 x 9.7 cm heterogeneous mass in the tail of the pancreas. This mass exhibited well-circumscribed enhancement during the arterial phase of the scan. Importantly, there was no radiographic evidence of metastatic disease at the time of imaging. Following surgical resection (distal pancreatectomy), the macroscopic examination of the resected specimen revealed a 12 x 12 x 10 cm mass with a tan-brown cut surface. Microscopic examination confirmed the diagnosis of a poorly differentiated neuroendocrine carcinoma. The tumor cells were arranged in solid nests and trabeculae, exhibiting high-grade nuclear features and frequent mitotic figures, indicating a high proliferative rate. Immunohistochemical staining was positive for chromogranin A, synaptophysin, and CD56, confirming the neuroendocrine origin of the tumor. Based on the collective findings from the patient's clinical presentation, imaging studies, and histopathological examination, the definitive diagnosis was a poorly differentiated pancreatic neuroendocrine carcinoma. This diagnosis carries significant implications for prognosis and treatment planning, necessitating a multidisciplinary approach to patient care (Table 1).

Following the diagnosis of a poorly differentiated pancreatic neuroendocrine carcinoma, the patient was counseled regarding treatment options, and a multidisciplinary team, including surgeons, oncologists, and gastroenterologists, formulated a comprehensive management plan. Surgical resection was deemed the most appropriate course of action given the localized nature of the tumor and the absence of distant metastases. The patient underwent

a laparotomy and distal pancreatectomy under general anesthesia. A midline abdominal incision provided access to the abdominal cavity. Intraoperative findings confirmed the presence of a 12 x 12 cm solid mass in the tail of the pancreas, consistent with preoperative imaging. The surgical procedure involved meticulous dissection and ligation of the pancreatic duct and splenic vessels to ensure complete removal of the tumor while preserving the spleen. Hemostasis was carefully achieved to minimize blood loss, and the abdomen was irrigated with normal saline to remove any residual debris. The abdominal wall was then closed in layers. The estimated blood loss during the procedure was 200 mL, and there were no intraoperative complications. The total operative time was 3 hours. The patient's postoperative recovery was largely uncomplicated. Pain management was achieved with intravenous analgesics, and she tolerated a progressive diet, starting with clear liquids and advancing to a regular diet as tolerated. A Jackson-Pratt drain was placed in the surgical bed to monitor for fluid accumulation and removed on postoperative day 2 due to minimal output. The final pathology report confirmed the diagnosis of poorly differentiated pancreatic neuroendocrine carcinoma. Importantly, the surgical margins were negative, indicating complete resection of the tumor. The patient was discharged home on postoperative day 7 in stable condition with appropriate instructions for wound care and follow-up appointments. She was referred to medical oncology for consideration of adjuvant chemotherapy to reduce the risk of recurrence. The patient's postoperative follow-up was closely monitored to assess for any complications and to detect early signs of recurrence; 1 week post-discharge: The patient underwent a clinical examination to assess surgical wound healing and overall recovery. There were no signs of infection or other complications; 3 months postoperative: A repeat CT scan of the abdomen was performed, which showed no evidence of recurrence; 6 months postoperative: Another CT scan of the abdomen was conducted, again demonstrating no evidence of recurrence; Subsequent follow-up: The patient continues to undergo ongoing surveillance with imaging and clinical evaluation every 6-12 months to monitor for any signs of recurrence. This comprehensive and vigilant follow-up strategy is crucial for the early detection and management of any potential recurrence, ultimately aiming to optimize the patient's long-term outcome (Table 2).

3. Discussion

Pancreatic neuroendocrine tumors (PNETs) are a captivating yet enigmatic subset of pancreatic neoplasms. They originate from the diffuse neuroendocrine system, a complex network of cells distributed throughout the body that produce and release hormones in response to various stimuli. Within the pancreas, these neuroendocrine cells reside within the islets of Langerhans, microscopic clusters of cells that play a vital role in regulating glucose homeostasis, digestion, and other essential physiological processes. While PNETs can develop at any age, they exhibit a striking predilection for older adults, with the peak incidence occurring in the seventh decade of life. This age-related distribution reflects the cumulative effect of genetic and environmental factors that contribute to the development of these tumors. In contrast, the occurrence of PNETs in young adults is a relatively rare phenomenon, often shrouded in diagnostic challenges and therapeutic uncertainties. The case presented here vividly illustrates the perplexing nature of PNETs in young adults. A 22-year-old woman, at the prime of her life, presented with vague abdominal discomfort, nausea, and vomiting – symptoms easily attributable to a myriad of common and benign gastrointestinal ailments. The absence of classic hormonal syndromes, such as hypoglycemia, peptic ulcer disease, or hyperglycemia, further masked the underlying pathology, leading to a delay in diagnosis and treatment.

Table 1. Anamnesis, physical examination, laboratory, imaging, histopathology, and diagnosis.

Domain	Findings
Anamnesis	* Chief Complaint: a 22-year-old woman, Left upper quadrant abdominal pain * History of Present Illness: 3-month history of vague abdominal discomfort, nausea, and vomiting. Increasing discomfort with a palpable mass in the left upper quadrant. 10-kg weight loss in the past month No history of fever, jaundice, or changes in bowel habits * Past Medical History: No significant medical history, including diabetes, hypertension, or family history of pancreatic tumors * Medications: No regular medications * Allergies: No known drug allergies * Social History: Non-smoker, non-alcoholic, no history of illicit drug use
Physical examination	* General Appearance: Well-appearing, alert, and oriented * Vital Signs: BP: 130/88 mmHg, HR: 81 bpm, RR: 20 breaths/min, Temp: 36.3°C * HEENT: Normocephalic, no scleral icterus or conjunctival pallor, no lymphadenopathy * Respiratory: Clear to auscultation bilaterally, no wheezes, rales, or rhonchi * Cardiovascular: Regular rate and rhythm, no murmurs, gallops, or rubs * Gastrointestinal: Soft, non-tender, palpable mass in the left upper quadrant, no hepatosplenomegaly, normal bowel sounds * Extremities: No edema, normal capillary refill, no clubbing or cyanosis * Neurological: Grossly intact, no focal deficits
Laboratory	* Complete Blood Count: WBC: 9.4 x 10^9/L (normal range: 4.5-14.5 x 10^9/L) Hemoglobin: 13.7 g/dL (normal range: 12.3-15.3 g/dL) Hematocrit: 42% (normal range: 33-45%) Platelets: 228 x 10^9/L (normal range: 150-450 x 10^9/L) * Liver Function Tests: AST: 22 U/L (normal range: 0-40 U/L) ALT: 28 U/L (normal range: 0-40 U/L) Alkaline phosphatase: 80 U/L (normal range: 40-120 U/L) Total bilirubin: 1.0 mg/dL (normal range: 0.1-1.2 mg/dL) * Renal Function Tests: Creatinine: 0.6 mg/dL (normal range: 0.9-1.3 mg/dL) Urea: 28 mg/dL (normal range: <50 mg/dL) Electrolytes: within normal limits * Tumor Markers: CEA: 2.0 ng/mL (normal range: <5 ng/mL) CA 19-9: 15 U/mL (normal range: <37 U/mL) * Coagulation Studies: PT: 12.3 seconds (normal range: 10-15 seconds) APTT: 35.9 seconds (normal range: 20-40 seconds) INR: 0.87 * Serology: HBsAg: Non-reactive
Imaging	* Chest X-ray: No evidence of pulmonary metastases * Abdominal CT Scan: 9.7 x 11.4 x 9.7 cm heterogeneous mass in the tail of the pancreas. Well-circumscribed with arterial phase enhancement. No evidence of metastatic disease *
Histopathology	* Macroscopic: 12 x 12 x 10 cm mass with a tan-brown cut surface * Microscopic: Poorly differentiated neuroendocrine carcinoma Tumor cells arranged in solid nests and trabeculae. High-grade nuclear features and frequent mitoses. Immunohistochemical staining was positive for chromogranin A, synaptophysin, and CD56.
Diagnosis	Poorly differentiated pancreatic neuroendocrine carcinoma

Table 2. Surgery procedure and follow-up.

Domain	Findings
Surgical procedure	* Procedure: Laparotomy and distal pancreatectomy * Anesthesia: General anesthesia * Incision: Midline abdominal incision * Findings: 12 x 12 cm solid mass in the tail of the pancreas * Procedure Details: Distal pancreatectomy with preservation of the spleen Careful dissection and ligation of the pancreatic duct and splenic vessels Hemostasis achieved Abdomen irrigated with normal saline Layered closure of the abdominal wall * Estimated Blood Loss: 200 mL * Complications: None * Operative Time: 3 hours
Postoperative course	* Recovery: Uncomplicated * Pain Management: Adequate pain control with intravenous analgesics * Diet: Progressive diet as tolerated, starting with clear liquids and advancing to a regular diet * Drain Management: Jackson-Pratt drain placed in the surgical bed and removed on postoperative day 2 after minimal output * Discharge: Discharged home on postoperative day 7 * Pathology: Final pathology confirmed poorly differentiated pancreatic neuroendocrine carcinoma with negative surgical margins * Adjuvant Therapy: Referred to medical oncology for consideration of adjuvant chemotherapy * Follow-up: - 1 week post-discharge: Clinic visit to assess surgical wound healing and overall recovery No signs of infection or other complications - 3 months postoperative: Repeat CT scan of the abdomen No evidence of recurrence - 6 months postoperative: Repeat CT scan of the abdomen No evidence of recurrence - Subsequent follow-up: Ongoing surveillance with imaging and clinical evaluation every 6-12 months

This case underscores a critical aspect of PNETs in young adults, their tendency to present with non-specific symptoms. Unlike their functional counterparts, which often announce their presence with dramatic hormonal disturbances, non-functional PNETs can silently grow and invade surrounding tissues before they are detected. This insidious nature poses a significant diagnostic challenge, as clinicians may not initially consider PNETs in their differential diagnosis, especially in young patients. Early detection of PNETs in young adults is paramount, as it allows for timely intervention and potentially curative treatment. However, the non-specific nature of symptoms and the rarity of these tumors in this age group create a formidable barrier to early diagnosis. Clinicians must maintain a high index of suspicion for PNETs in young adults presenting with abdominal symptoms, even in the absence of classic hormonal syndromes. A comprehensive evaluation, including a detailed medical history, a thorough physical examination, and a judicious selection of laboratory and imaging studies, is essential for unraveling the diagnostic puzzle. The symptoms of PNETs in young adults often mimic those of more common gastrointestinal disorders, such as gastritis, irritable bowel syndrome, or even anxiety. This overlap in symptomatology can lead to misdiagnosis and delayed treatment. PNETs are relatively rare, and many healthcare providers may not be familiar with their presentation in young adults. This lack of awareness can contribute to delayed diagnosis and referral to specialists. Even when PNETs are suspected, distinguishing them from other pancreatic masses, such as adenocarcinoma or cystic neoplasms, can be challenging. Advanced imaging techniques and tissue biopsy are often required to establish a definitive diagnosis. Imaging studies, such as computed tomography (CT) and magnetic resonance imaging (MRI), play a pivotal role in the detection and characterization of PNETs. These modalities provide detailed anatomical information about the tumor's size, location, and relationship to surrounding structures. Functional imaging techniques, such as somatostatin receptor scintigraphy (SRS) and positron emission tomography (PET), can further aid in identifying hormonally active tumors and detecting metastases. CT scans provide detailed cross-sectional images of the abdomen and pelvis, allowing for visualization of the pancreas and surrounding organs. PNETs typically appear as well-defined masses with varying degrees of enhancement. MRI scans offer superior soft tissue contrast compared to CT scans, providing more detailed information about the tumor's internal structure and its relationship to adjacent blood vessels and ducts. SRS and PET scans exploit the unique characteristics of PNETs to aid in diagnosis and staging. SRS utilizes radiolabeled somatostatin analogs, which bind to somatostatin receptors commonly expressed on PNETs, allowing for visualization of the tumor. PET scans, using radiolabeled glucose analogs, can detect areas of increased metabolic activity, which is often seen in PNETs. However, imaging studies alone cannot provide a definitive diagnosis of PNETs. A tissue biopsy, obtained through endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) or surgical exploration, is necessary to confirm the diagnosis and determine the tumor's grade and histological subtype. Microscopic examination of the biopsied tissue allows for the identification of characteristic cellular features of PNETs, such as the arrangement of cells in nests, trabeculae, or rosettes. Immunohistochemical staining utilizes antibodies to detect specific proteins expressed by PNET cells, such as chromogranin A, synaptophysin, and CD56. These markers help confirm the neuroendocrine origin of the tumor and differentiate it from other pancreatic neoplasms. Histopathological analysis also allows for the grading of PNETs based on their degree of differentiation and mitotic activity. This grading system helps predict the tumor's aggressiveness and guides treatment decisions. The management of PNETs in young adults requires a multidisciplinary approach, involving gastroenterologists, surgeons, oncologists, radiologists, and pathologists. This collaborative effort ensures that patients receive comprehensive and

individualized care, tailored to their specific needs and circumstances. A multidisciplinary team can provide a comprehensive evaluation of the patient's condition, considering all aspects of the disease, including its clinical presentation, imaging findings, and histopathological features. Based on the comprehensive evaluation, the multidisciplinary team can develop an individualized treatment plan that takes into account the patient's age, overall health, tumor characteristics, and personal preferences. A multidisciplinary approach ensures coordinated care throughout the patient's journey, from diagnosis to treatment and follow-up. This coordinated care minimizes the risk of fragmentation and ensures that patients receive the best possible outcomes. A diagnosis of PNETs can have a profound impact on the quality of life of young adults. The physical symptoms of the tumor, the emotional distress associated with a cancer diagnosis, and the potential side effects of treatment can all take a toll on patients' physical and mental well-being. A cancer diagnosis can trigger a range of emotions, including fear, anxiety, and depression. Young adults with PNETs may face additional challenges, such as concerns about their fertility, body image, and future prospects. The physical symptoms of PNETs, such as abdominal pain, nausea, and fatigue, can significantly impact daily activities and quality of life. Treatment for PNETs, including surgery, chemotherapy, and radiation therapy, can cause a variety of side effects that can affect patients' physical and emotional well-being. Supportive care, including psychological counseling, nutritional support, and pain management, is essential for helping young adults cope with the challenges of living with PNETs. Psychological counseling can provide emotional support and coping strategies for young adults facing a cancer diagnosis and treatment. Maintaining adequate nutrition is crucial for patients with PNETs, especially those undergoing treatment. Nutritional counseling can help patients optimize their dietary intake and manage any nutritional deficiencies. Effective pain management is essential for improving quality of life and allowing patients to participate in daily activities.[11-13]

The diagnosis of pancreatic neuroendocrine tumors (PNETs) in young adults presents a formidable challenge, akin to navigating a labyrinth with hidden pathways and deceptive turns. Their rarity, non-specific clinical presentation, and the propensity to mimic more common and benign conditions create a diagnostic maze that demands a high index of suspicion and a proactive approach to investigation. Clinicians must remain vigilant, even when confronted with seemingly innocuous symptoms, as early detection is crucial for timely intervention and potentially curative treatment. PNETs in young adults often deviate from the classic presentation seen in older individuals. The absence of typical hormonal syndromes, such as hypoglycemia in insulinomas or peptic ulcer disease in gastrinomas, and the predominance of vague abdominal symptoms can easily mislead clinicians towards more common diagnoses. This is where the art of clinical suspicion comes into play. Clinicians must challenge the prevailing notion that PNETs are primarily a disease of the elderly. While the incidence is undoubtedly higher in older adults, the possibility of PNETs should not be dismissed in younger patients, especially those presenting with persistent or unexplained abdominal symptoms. While evidence-based medicine forms the foundation of clinical practice, intuition and a "gut feeling" can play a crucial role in raising suspicion for rare conditions like PNETs. Clinicians should trust their instincts when something doesn't quite fit the usual pattern. The diagnostic journey for PNETs in young adults is often fraught with uncertainty. Clinicians must be comfortable navigating this ambiguity, pursuing investigations even when initial findings are inconclusive. A detailed medical history, including a comprehensive review of systems, is akin to piecing together a puzzle, where each piece of information contributes to the overall picture. Particular attention should be paid to any subtle changes in bowel habits, appetite, weight, and energy levels, as these can be early indicators of an underlying pancreatic pathology. Active listening is

crucial for capturing the nuances of the patient's narrative. Clinicians should encourage patients to describe their symptoms in their own words, paying attention not only to what they say but also to how they say it. Understanding the timeline of symptom onset and progression can provide valuable insights. Gradual onset and insidious progression of symptoms are often characteristic of PNETs. While PNETs in young adults are often sporadic, a family history of neuroendocrine tumors or genetic syndromes associated with PNETs should be explored. A thorough physical examination, including careful palpation of the abdomen, is an essential component of the diagnostic evaluation. The presence of a palpable mass, even in the absence of other significant findings, should raise suspicion and prompt further investigation. Palpation of the abdomen requires a gentle yet deliberate approach. Clinicians should assess for any masses, tenderness, or organomegaly. The physical examination should not be limited to the abdomen. Examination of other systems, such as the skin (for flushing or rashes) and the lymph nodes (for enlargement), can provide additional clues. The diagnostic workup for PNETs in young adults often involves a combination of laboratory investigations, imaging studies, and endoscopic procedures. While routine blood work and serological tests may not reveal specific abnormalities in patients with non-functional PNETs, they can help rule out other potential causes of the patient's symptoms and provide baseline values for future monitoring. Imaging studies, such as computed tomography (CT) and magnetic resonance imaging (MRI), play a pivotal role in the detection and characterization of PNETs. These modalities provide detailed anatomical information about the tumor's size, location, and relationship to surrounding structures. Functional imaging techniques, such as somatostatin receptor scintigraphy (SRS) and positron emission tomography (PET), can further aid in identifying hormonally active tumors and detecting metastases. EUS, with its ability to provide high-resolution images of the pancreas and facilitate fine-needle aspiration (FNA) for tissue diagnosis, has emerged as a valuable tool in the evaluation of pancreatic masses. While imaging studies provide valuable clues, the definitive diagnosis of PNETs rests on histopathological examination and immunohistochemical staining of tissue obtained through EUS-FNA or surgical biopsy. The journey of a patient with PNETs does not end with diagnosis and treatment. Long-term surveillance is crucial for monitoring for recurrence, managing late complications, and providing ongoing support. Even after successful treatment, patients with PNETs require long-term surveillance to monitor for any signs of recurrence. This surveillance often involves periodic imaging studies, blood tests, and clinical evaluations. PNETs can sometimes cause late complications, such as hormonal imbalances or metastatic disease, even years after initial treatment. Clinicians must remain vigilant in recognizing and managing these late effects. The care of patients with PNETs extends beyond the physical realm. Addressing the psychosocial impact of the disease and providing emotional support are essential components of holistic care.[14-16]

While imaging studies provide valuable clues and illuminate the path toward diagnosis, the definitive diagnosis of pancreatic neuroendocrine tumors (PNETs) rests on the bedrock of histopathology and immunohistochemistry. These techniques delve into the cellular and molecular intricacies of the tumor, unraveling its secrets and revealing its true nature, much like archaeologists carefully excavating and analyzing ancient artifacts to understand a civilization's history. Histopathology, the microscopic examination of tissue, provides a window into the cellular architecture and morphology of the tumor. Imagine peering through a powerful microscope, where a seemingly homogenous mass transforms into a complex landscape of cells, each with its own unique characteristics and secrets to tell. PNETs exhibit a wide spectrum of microscopic features, ranging from well-differentiated tumors with a low proliferative rate, resembling a well-organized community, to poorly differentiated carcinomas with aggressive behavior, akin to a chaotic and rapidly expanding city. In the

case presented, the histopathological examination revealed a poorly differentiated neuroendocrine carcinoma, characterized by high-grade nuclear features and frequent mitotic figures. These findings painted a picture of a tumor with a propensity for rapid growth and potential for metastasis, like an invasive force with ambitions beyond its borders. Immunohistochemistry, a technique that utilizes antibodies to detect specific proteins within tissue, complements histopathology by providing molecular insights. It's like having a specialized key that unlocks specific doors within the cellular landscape, revealing hidden compartments and secret passages. In PNETs, immunohistochemical staining can confirm the neuroendocrine origin of the tumor and differentiate it from other pancreatic neoplasms, ensuring that the diagnosis is precise and targeted. In this case, immunohistochemical staining confirmed the neuroendocrine nature of the tumor, with positive staining for chromogranin A, synaptophysin, and CD56. These markers, like beacons in the cellular landscape, illuminated the tumor's identity and guided the path toward definitive diagnosis. PNETs can exhibit various architectural patterns, including nests, trabeculae, rosettes, and solid sheets. These patterns, like fingerprints or the unique layout of a city, provide clues about the tumor's behavior and potential for aggression. Nests, for instance, resemble tightly knit communities, while trabeculae are akin to well-organized streets. Rosettes, on the other hand, evoke images of intricate floral patterns, and solid sheets resemble dense urban blocks. The individual cells within a PNET can also vary in size, shape, and staining characteristics. Poorly differentiated tumors often display cellular pleomorphism, with cells exhibiting irregular shapes and sizes, a hallmark of aggressive behavior. Imagine a city populated by individuals of diverse backgrounds and appearances, some conforming to the norm, others deviating with unique traits. The nucleus, the command center of the cell, can also provide valuable information. High-grade nuclear features, such as enlarged nuclei, irregular nuclear membranes, and prominent nucleoli, are often associated with poorly differentiated tumors. These features suggest a nucleus that is actively engaged in directing cellular processes, potentially driving the tumor's aggressive behavior. Mitotic figures, the visible signs of cell division, are an indicator of the tumor's proliferative rate. Frequent mitotic figures suggest a rapidly growing tumor with a higher potential for metastasis. It's like observing a city in a state of rapid expansion, with new buildings and infrastructure constantly emerging. Chromogranin A, synaptophysin, and CD56 are commonly used markers to confirm the neuroendocrine origin of PNETs. These markers are expressed by neuroendocrine cells throughout the body and serve as identifiers of this specific cell lineage, much like an identity card confirms an individual's citizenship. Immunohistochemical staining can also be used to assess hormone expression within the tumor. This information can help classify the tumor as functional or non-functional and guide treatment decisions. It's like intercepting the chemical messages that cells use to communicate, revealing their intentions and potential impact. Some markers, such as Ki-67, a marker of cell proliferation, can provide prognostic information. High Ki-67 expression is often associated with more aggressive tumor behavior and a worse prognosis. It's like having a glimpse into the future, predicting the tumor's potential course and guiding treatment strategies. The diagnosis of a poorly differentiated PNET carries significant prognostic implications. These tumors are known for their aggressive behavior, potential for metastasis, and poorer response to treatment. Early diagnosis and prompt intervention are crucial for improving outcomes in these patients. Accurate diagnosis of PNETs, including their grade and differentiation, is essential for guiding treatment decisions and predicting prognosis. Histopathology and immunohistochemistry, with their ability to delve into the cellular and molecular intricacies of the tumor, provide the foundation for accurate diagnosis and personalized patient care. It's like having a detailed map of the tumor's landscape, guiding the clinician's

every move and ensuring that treatment is tailored to the individual patient's needs.[17,18]

Surgical resection stands as the cornerstone of treatment for localized pancreatic neuroendocrine tumors (PNETs), offering the potential for cure in selected patients. It's the primary weapon in the arsenal against these tumors, a beacon of hope for those seeking a definitive solution. The extent of surgical resection, however, is not a one-size-fits-all approach, it hinges on the tumor's size, location, and its intricate relationship with surrounding structures. Distal pancreatectomy, as performed in this case, is the standard surgical procedure for PNETs located in the tail of the pancreas. This procedure involves the removal of the tail of the pancreas, the portion that harbors the tumor, while preserving the remaining head and body of the pancreas. It's a delicate dance between removing the disease and preserving the organ's function. The surgical management of PNETs is not for the faint of heart, it demands careful dissection and meticulous attention to detail. The pancreatic duct and splenic vessels, like delicate threads interwoven within the pancreatic fabric, must be carefully ligated to prevent complications such as pancreatic fistula or splenic infarction. Hemostasis, the meticulous control of bleeding, must be achieved to minimize blood loss and ensure a clear surgical field. And finally, the abdomen must be thoroughly irrigated to remove any residual tumor cells, ensuring that no enemy combatants remain to threaten a resurgence. In this case, the surgical procedure was a resounding success, with no intraoperative complications. The patient's postoperative recovery was largely uneventful, a testament to the surgical team's skill and the patient's resilience. She was discharged home on postoperative day 7, ready to embark on the next chapter of her life. The final pathology report confirmed negative surgical margins, indicating complete resection of the tumor, a victory in the battle against this formidable foe. Before embarking on the surgical journey, a thorough preoperative evaluation is essential. This includes a detailed assessment of the patient's overall health, nutritional status, and any co-existing medical conditions. Imaging studies, such as CT scans and MRI, provide crucial information about the tumor's size, location, and relationship to surrounding structures, guiding the surgical strategy. The choice of surgical technique depends on the tumor's location and size. For tumors located in the head of the pancreas, a Whipple procedure, a more extensive operation, may be necessary. Laparoscopic techniques, which involve smaller incisions and less invasive procedures, are increasingly being used for smaller tumors, offering the potential for faster recovery and fewer complications. The pancreas, nestled deep within the abdomen, presents unique surgical challenges. Its rich blood supply and proximity to vital structures, such as the duodenum and bile duct, demand meticulous dissection and careful handling of tissues. Postoperative care is crucial for ensuring a smooth recovery and minimizing complications. Pain management, nutritional support, and early ambulation are essential components of postoperative care. Surgical resection offers the most effective means of removing the tumor and reducing the risk of recurrence. It's like removing the weeds from a garden, preventing them from taking root and spreading. Studies have shown that surgical resection of localized PNETs can significantly improve overall survival and disease-free survival. It's a chance at a second life, free from the clutches of this insidious disease. Surgical resection can also alleviate symptoms caused by the tumor, such as abdominal pain, jaundice, and weight loss. It's like removing a thorn from the side, providing much-needed relief and restoring quality of life.[19,20]

4. Conclusion

This case underscores the complexity and diagnostic challenges of pancreatic neuroendocrine tumors (PNETs) in young adults. The patient's non-specific symptoms, such as vague abdominal discomfort, nausea, and vomiting, are easily misconstrued as common gastrointestinal issues, leading to potential delays in diagnosis. This

emphasizes the necessity for clinicians to maintain a high index of suspicion for PNETs even in younger patients, particularly when presenting with such symptoms. Early diagnosis is crucial for improving patient outcomes and facilitating timely intervention. Advanced imaging techniques, such as computed tomography (CT) and magnetic resonance imaging (MRI), play a vital role in detecting and characterizing these tumors. Additionally, functional imaging techniques, including somatostatin receptor scintigraphy (SRS) and positron emission tomography (PET), aid in identifying hormonally active tumors and detecting metastases, which are crucial for accurate staging and treatment planning. The definitive diagnosis of PNETs relies on histopathological examination and immunohistochemical staining of tissue samples. These techniques provide crucial insights into the cellular and molecular characteristics of the tumor, enabling accurate diagnosis and personalized treatment planning. Surgical resection remains the cornerstone of treatment for localized PNETs, offering the potential for cure in selected patients. The extent of surgical resection depends on the tumor's size, location, and relationship with surrounding structures. In this case, the patient underwent a successful distal pancreatectomy with no intraoperative complications. Postoperative care is crucial for ensuring a smooth recovery and minimizing complications. In conclusion, this case highlights the necessity for a multidisciplinary approach in managing PNETs, encompassing gastroenterologists, surgeons, oncologists, radiologists, and pathologists. This collaborative effort ensures comprehensive and individualized care tailored to the patient's specific needs. The diagnosis of PNETs can significantly impact a young adult's quality of life, underscoring the importance of supportive care, including psychological counseling, nutritional support, and pain management.

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