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Acute Appendicitis Revealing a Neuroendocrine Tumor: A Case Report

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Abstract | **Case Report**

Appendiceal neuroendocrine tumors (ANETs) rarely are first-hand diagnoses made after appendectomy for suspected acute appendicitis. We present the case of a 27-year-old man with classical signs of appendicitis. The histopathological report showed that it was a well-differentiated (G1) neuroendocrine tumor of 7 mm in size with no invasive features. Further immunohistochemistry was carried out to confirm the diagnosis (chromogranin-A+, CD56+, Ki-67 <1%). No further treatment was needed. After three years of clinical follow-up, there is neither recurrence nor metastasis. The taken case underlines the importance of systematic histological analysis and an excellent prognosis of small, non-invasive ANETs.

Keywords: Appendiceal Neuroendocrine Tumor, Acute Appendicitis, Laparoscopic Appendectomy, Well-Differentiated NET, Histopathology, Ki-67 Index.

INTRODUCTION

Appendiceal neuroendocrine tumors (ANETs) are the most prevalent neoplasms of the vermiform appendix. ANETs are often incidental findings during histological examination of appendectomy specimens performed for presumed acute appendicitis, ranging from silent lesions to metastatic tumors [1-3]. We report a rare case of an appendiceal neuroendocrine tumor initially presenting as acute appendicitis.

CASE REPORT

A 27-year-old male patient, with no significant medical history, presented with a 12-hour history of abdominal pain localized to the right iliac fossa, accompanied by vomiting but no other digestive symptoms. On physical examination, the patient was hemodynamically and respiratory stable, afebrile (37.1 °C), with localized tenderness in the right iliac fossa without guarding or abdominal rigidity. No palpable mass was noted.

Laboratory findings revealed a white blood cell count of 12.3 × 10^3/µL, neutrophils at 79%, and C-reactive protein (CRP) at 21 mg/L. Liver, renal, and electrolyte biochemical parameters were within normal limits. Abdominal ultrasound and computed tomography (CT) were consistent with a diagnosis of acute appendicitis.

The patient underwent a laparoscopic appendectomy, and the specimen was submitted for histopathological analysis. The final pathology report confirmed a well-differentiated localized neuroendocrine tumor (NET G1) located at the tip of the appendix, measuring 7 mm at its greatest dimension, without signs of invasion. Surgical margins were clear. Immunohistochemical analysis confirmed the neuroendocrine nature of the proliferation, with positive staining for chromogranin A and CD56, and a Ki-67 index <1%. (pT1, R0, G1)

No additional treatment was required. Clinical follow-up was conducted annually, with no evidence of recurrence or metastasis observed over a three-year period.

DISCUSSION

Appendiceal neuroendocrine tumors (ANETs) are rare neoplasms, accounting for approximately 0.3% to 0.9% of appendectomy specimens in adults and only 0.08% in pediatric populations [4,5]. Their clinical presentation is typically non-specific and often mimics acute appendicitis, making preoperative diagnosis highly challenging [1-6]. In most cases, ANETs are discovered incidentally through histopathological examination of appendices removed for presumed appendicitis.

While CT imaging may occasionally suggest an appendiceal mass, radiologic findings are not reliable for differentiating ANETs from other appendiceal lesions such as mucoceles or adenocarcinomas [4-8]. Thus, definitive diagnosis relies on histological and immunohistochemical analysis of the resected specimen, typically showing positivity for neuroendocrine markers (synaptophysin, chromogranin A, CD56) and a proliferation index (Ki-67) used in the WHO grading system (G1 to G3) [9,10].

Surgical management is based primarily on tumor size, location, histologic grade, and margin status. Appendectomy alone is considered curative for tumors ≤1 cm that are well differentiated (G1), confined to the appendix without mesoappendiceal or vascular invasion, and with negative resection margins [5-11]. In contrast, tumors >2 cm, or those with adverse features (e.g., basal location, mesoappendiceal invasion >3 mm, lymphovascular involvement, or higher grade), generally require a right hemicolectomy to ensure complete oncologic resection and lymphadenectomy [8-12].

Management of intermediate-sized tumors (1–2 cm) remains debated. Multiple studies and international guidelines (ENETS, NANETS, NCCN) advocate for individualized decision-making based not only on size but also on other histopathologic risk factors such as tumor grade and invasion depth [1-13]. Multidisciplinary team involvement is strongly recommended to guide optimal treatment planning.

Postoperative follow-up is similarly risk-adapted. For patients with ANETs ≤1 cm, well-differentiated, and fully resected with clear margins (R0), no routine radiological or biochemical follow-up is necessary [11]. In contrast, patients with tumors >1 cm and additional risk factors, or those with uncertain margin status, require closer surveillance. This may include clinical assessment, serial measurement of chromogranin A, and imaging (CT, MRI, or PET-CT) if recurrence is suspected [6-14].

Despite their generally favorable prognosis-5-year survival rates exceeding 90% for localized disease [8-10], the rarity of ANETs, especially in children, and their non-specific presentation highlight the importance of routine histopathological examination of all appendectomy specimens. Early and accurate diagnosis, along with thorough evaluation of prognostic factors, is critical for curative treatment and to avoid unnecessary secondary surgeries [6-15].

CONCLUSION

Appendiceal neuroendocrine tumors are rare, most often discovered incidentally following appendectomy for suspected acute appendicitis. This case highlights the importance of systematic histopathological examination of all appendectomy specimens and also the excellent prognosis of small, well-differentiated (G1) NETs with no invasive features. In such cases, appendectomy is curative, and patients are followed clinically long-term. Good knowledge of this tumor is important to allow appropriate management and avoid overtreatment.

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