Case report

CARCINOID TUMOR OF THE APPENDIX (NEUROENDOCRINE TUMOR OF THE APPENDIX) IN THE SETTING OF ACUTE APPENDICITIS: A CLINICAL CASE OF A 19-YEAR-OLD PATIENT

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Background: Appendiceal neuroendocrine tumors (ANETs) are rare and often present alongside acute appendicitis [1, 4]. Tumors larger than 2 cm are associated with high oncological risk according to leading guidelines (NCCN, ENETS) [11, 12]. Determining the extent of surgery (achieving R0 resection status) in emergency settings remains a complex clinical dilemma [7].
Case Presentation: We present the case of a 19-year-old patient diagnosed with acute appendicitis and a concomitant massive mass measuring 81x42 mm with regional lymphadenopathy on CT scan [9]. Based on high intraoperative oncological suspicion, a one-stage right hemicolectomy was performed. Pathomorphology confirmed an appendiceal carcinoid with cecal invasion, achieving R0 resection.
Conclusion: Our case underscores that in the presence of high-risk factors (size ≥2 cm, suspected invasion), one-stage radical surgical intervention is a legitimate and ethically justifiable risk [10].

Key words: Appendiceal Neuroendocrine Tumor, Carcinoid, Acute Appendicitis, Hemicolectomy, R0 Resection, Ki-67.

INTRODUCTION:

Relevance of the Topic and Clinical Challenge

Appendiceal neuroendocrine tumors (ANETs) represent a comparatively rare neoplastic entity [1]. Approximately one-third of these tumors present without specific clinical symptomatology; instead, they produce luminal obstruction of the vermiform appendix, which manifests clinically as the classic presentation of acute appendicitis [8]. This scenario places the operating surgeon in an exceptionally challenging intraoperative situation. During emergency surgery, where inflammatory infiltration obscures the operative field, the clinician must recognize a potentially malignant neoplasm and make an appropriate tactical decision.

In oncological surgery, the primary objective of any resection is to achieve R0 status — a resection margin free of neoplastic cells — to preclude R1 or R2 resection, both of which substantially worsen the patient's oncological prognosis.

AIMS OF THE STUDY

The present study aims to:

1. Drawing upon the most recent international consensus guidelines (NANETS [6], ENETS [12], and NCCN [11]), we delineate the critical determinants of ANET management: tumor size, depth of invasion, and lymphadenopathy.
2. Through detailed analysis of a clinical case, to substantiate the legitimacy of single-stage radical surgical intervention as the preferred approach to achieving R0 resection, notwithstanding diagnostic uncertainty.

LITERATURE REVIEW:

Controversy in the Surgical Management of ANETs

The surgical management of ANETs remains a subject of considerable debate among oncological and colorectal surgeons [4]. The principal challenge lies in the fact that acute appendicitis entirely masks the clinical presentation of appendiceal neuroendocrine tumors, and clinicians frequently identify them as incidental findings. In these circumstances, the surgeon must make intraoperative decisions under significant time constraints [7].

Historically, because clinicians regarded carcinoid tumors as comparatively indolent neoplasms, they often managed them conservatively and considered a simple appendectomy sufficient. However, pathomorphological studies and long-term observational data accumulated over recent decades have demonstrated that the biological behavior of these lesions is not uniform [1, 2]. Consequently, tumor size has emerged as the foremost anatomical criterion in determining the appropriate management strategy.

The Philosophical Dilemma in Surgical Management: Conservatism versus Radicalism

- Conservative approach (appendectomy): Proponents of this strategy argue that the majority of tumors — particularly small lesions (≤1 cm) — carry a minimal risk of metastatic dissemination [2].
- Radical approach (hemicolectomy): This position is grounded in the primacy of oncological safety. Advocates contend that whenever there is any suspicion of invasion or regional lymph node involvement, lym-

phatic dissection is necessary to achieve R0 resection. In their view, the risk of R1/R2 resection is considerably greater and less acceptable from an oncological standpoint [10].

The Two-Stage Management Concept: Risk Factors for Tumors Measuring 1–2 cm

The greatest degree of tactical uncertainty arises in the management of tumors measuring 1-2 cm in greatest dimension [7]. The literature favors a two-stage approach in such cases: initial appendectomy followed by comprehensive pathomorphological examination of the resected specimen.

Additionally, decisive risk factors that justify the performance of hemicolectomy [11] include:

1. **Ki-67 index and tumor grade:** A Ki-67 proliferation index of ≥3% is indicative of aggressive biological behavior [5, 6, 12].
2. **Invasion of the cecal wall:** When the tumor extends to the base of the appendix, appendectomy alone carries a near-certain risk of R1 residual disease [7, 12].
3. **Lymphovascular invasion:** The identification of neoplastic cells within blood vessels or lymphatic channels [5, 11].

International Guidelines and Size-Based Criteria (ENETS, NCCN)

Contemporary international guidelines (NANETS [6], ENETS [12], and NCCN [11]) now provide explicit recommendations based on tumor size and local invasion:

- Low risk (generally <1 cm): The risk of metastatic dissemination in this category is minimal; accordingly, appendectomy alone is considered adequate.
- Intermediate risk (1–2 cm): Requires detailed pathomorphological assessment with evaluation of all three risk factors (Ki-67 index, depth of invasion, and lymphovascular invasion).
- High risk (≥2 cm): At this tumor dimension, the risk of regional lymph node metastasis is substantially elevated [3]. Therefore, right hemicolectomy with lymph node dissection, aimed at guaranteeing R0 status, constitutes the gold standard of care [11, 12].

The Factor of Emergency Surgery: nflammation and Neoplasia

Of particular significance are clinical scenarios analogous to the present case, in which ANETs manifest in the context of acute appendicitis or periappendiceal abscess. In circumstances where the visually estimated tumor dimension clearly exceeds 2 cm and is accompanied by regional lymphadenopathy [9], the prevailing consensus within the oncological community favors single-stage radical resection [10]. This pragmatic approach aims to prevent residual neoplastic disease (R1/R2) and to ensure optimal oncological outcomes for the patient.

CASE REPORT

Diagnostic Findings and Clinical Presentation

Patient: A 19-year-old male presenting with severe abdominal pain and elevated inflammatory markers (leukocytosis, WBC↑).

Computed tomography (CT) of the abdomen demonstrated not only radiological signs consistent with acute appendicitis, but also an 81x42 mm heterogeneous mass in the periappendiceal region, accompanied by regional lymph node enlargement [9]. These findings were strongly indicative of a high probability of malignant neoplasia.

Operative Findings and Specimen Description

Intraoperative examination revealed a massive, indurated inflammatory infiltrate (approximately 8x6 cm), associated with a periappendiceal abscess and high suspicion of cecal wall invasion. Given the elevated oncological risk, the operating surgeon elected to proceed with a single-stage right hemicolectomy.

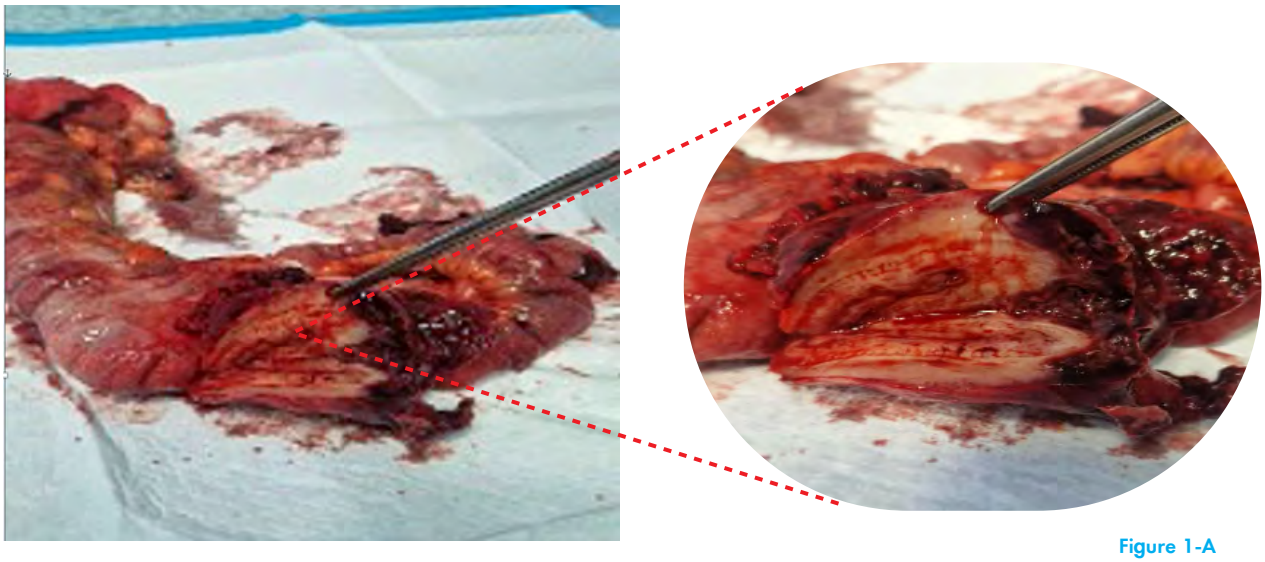

Figure 1-A

The resected specimen constituted a voluminous neoplastic mass. Figure 1A illustrates the gross appearance of the specimen (resected hemicolectomy specimen).

Pathomorphological Confirmation and Final Diagnosis

Pathomorphological examination confirmed the following:

- Carcinoid tumor (neuroendocrine neoplasm).
- Tumor invasion involving all layers of the cecal wall and the adjacent mesentery.
- Resection margins negative for neoplastic involvement (R0 status achieved).
- Figure 1B presents the gross pathological appearance of the specimen on cross-section (resected specimen on cross-section).

Postoperative Course and Outcome

The postoperative course was uneventful and complication-free. The patient was discharged with recommendations for scheduled outpatient oncological surveillance, given that complete (R0) oncological resection had been achieved [10].

DISCUSSION:

Legitimacy of the Surgical Decision from an Oncological Safety Perspective

The present case substantiates the validity of single-stage radical surgical intervention in the setting of intraoperative diagnostic uncertainty [10].

Primacy of R0 Resection in the Setting of Concurrent Infection

The tumor dimensions (≥2 cm) and cecal wall invasion indicated that appendectomy alone would have carried a near-certain risk of R1/R2 resection [7]. Achievement of R0 resection — the benchmark of oncological success — was possible exclusively by means of hemicolectomy.

Ethical and Oncological Justification of the Surgical Approach

The prevailing oncological principle dictates that the risk of oncological inadequacy (R1/R2 residual disease) is substantially greater and considerably less acceptable than the risk of performing an extensive resection for what may prove to be a benign process [10].

CONCLUSION:

Recommendations and Management Algorithm

Practical recommendations:

1. **Single-stage radical resection:** When intraoperative findings unequivocally demonstrate high-risk features — such as a visually estimated tumor size ≥2 cm, cecal wall invasion, or regional lymphadenopathy — the surgeon should perform a right hemicolectomy as a single-stage procedure [3, 10, 11, 12].
2. **Two-stage cautious approach:** When risk factors are intermediate (tumor size 1–2 cm), and no overt signs of invasion are identified, appendectomy followed by definitive histopathological assessment is preferable. In the event of confirmed high-risk features (Ki-67 ≥ 3% and R1 margins), completion hemicolectomy is indicated [5, 7, 12].

Conclusion:

In the management of ANETs, the operating surgeon should adhere to the principle: "An extended resection is preferable to an inadequate one" [10] — a dictum that ensures the patient's durable oncological safety and long-term disease control.

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აპენდიქსის კარცინოიდული სიმსივნე (ჭიანაწლავის ნეიროენდოკრინული სიმსივნე) მწვავე აპენდიციტის ფონზე: 19 წლის პაციენტის კლინიკური შემთხვევა

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რეზიუმე

შესავალი: აპენდიქსის ნეიროენდოკრინული სიმსივნეები (ANETs) იშვიათია და ხშირად მწვავე აპენდიციტის ფონზე ვლინდება [1]. 2 სმ-ზე მეტი ზომის წარმონაქმნები ონკოლოგიურად მაღალი რისკის შემცველია [3, 11]. გადაუდებელ სიტუაციებში ქირურგიის მოცულობის განსაზღვრა (R0 რეზექციის მიღწევა) რჩება რთულ კლინიკურ დილემად.

კლინიკური შემთხვევა: წარმოგიდგენთ 19 წლის პაციენტის შემთხვევას, რომელსაც მწვავე აპენდიციტის ფონზე, კტ-ზე დაუფიქსირდა 81x42 მმ ზომის მასიური წარმონაქმნი ლიმფადენოპათიით. ინტრაოპერაციული მაღალი ონკოლოგიური ეჭვის საფუძველზე, ჩატარდა ერთეტაპიანი მარჯვენამხრივი ჰემიკოლექტომია. პათომორფოლოგიურმა პასუხმა დაადასტურა კარცინოიდი ცეკუმში ინვაზიით, მიღწეული იქნა R0 რეზექცია.

დასკვნა: ჩვენი შემთხვევა ხაზს უსვამს იმას, რომ მაღალი რისკის ფაქტორების (ზომა ≥2 სმ, ინვაზიის ეჭვი) არსებობისას, ერთეტაპიანი რადიკალური ქირურგიული ჩარევა არის ლეგიტიმური და ეთიკურად გამართლებული რისკი [12].

საკვანძო სიტყვები: აპენდიქსის ნეიროენდოკრინული სიმსივნე, კარცინოიდი, მწვავე აპენდიციტი, ჰემიკოლექტომია, R0 რეზექცია, Ki-67.