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NEUROENDOCRINE TUMOR OF THE APPENDIX: CASE REPORT

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Abstract: Neuroendocrine tumors (NETs) of the appendix are rare neoplasms, and in most cases, they are diagnosed incidentally in appendectomy specimens. The occurrence of NETs in the appendicular stump, especially after a long interval since surgery, is an exceptionally uncommon condition. In this report, we present the case of a 67-year-old patient who had undergone appendectomy at age 16 and who, during initial investigation for endometrial thickening, presented with a subepithelial lesion in the appendicular stump orifice identified by colonoscopy. Histopathological diagnosis confirmed that it was an appendicular stump NET. The patient underwent laparoscopic right colectomy, with satisfactory postoperative evolution. Clinical, diagnostic, and therapeutic aspects of the case are discussed, contextualized in light of the most recent literature.

Keywords: Neuroendocrine tumor; Appendicular stump; Colectomy;

Introduction

Neuroendocrine tumors (NETs) of the appendix represent approximately 0.2% to 0.7% of appendectomy specimens, being detected incidentally in most cases in appendices removed due to suspected acute appendicitis[1,2]. The identification of NET in the appendicular stump years after appendectomy is an extremely rare condition, with few reports in the international literature[4] . This report describes the case of an ENT of the appendicular stump diagnosed incidentally during a colonoscopy performed to investigate gynecological pathology in an elderly patient, highlighting the approach adopted and reviewing the most recent evidence on the management of these tumors.

Case Report

A 67-year-old female patient, hypertensive and diabetic, with a history of conventional appendectomy performed at age 16, with no available pathological report from that time. During investigation of endometrial thickening, a colonoscopy was requested, which revealed an oval subepithelial nodular lesion, approximately 20 mm in size, located in the appendicular stump region (Figure 1 and Figure 2).

An endoscopic biopsy of the lesion was performed, and histopathological examination revealed a well-differentiated neuroendocrine tumor (grade 1), with positive immunoreactivity for CD56, chromogranin A, synaptophysin, and a Ki-67 cell proliferation index of less than 3%. Subsequently, staging was performed with computed tomography of the chest, abdomen, and pelvis, with no evidence of metastases or lymphadenopathy.

Given the diagnosis and the size of the lesion on colonoscopy, a right oncological colectomy by laparoscopy was performed without complications. The patient had a good postoperative recovery and was discharged on the fifth day of hospitalization.

The pathological examination of the surgical specimen showed a well-differentiated neuroendocrine tumor (grade 1), measuring 14 x 11 mm, extending to the subserosa and perineural invasion. Thirteen lymph nodes were resected, all free of neoplasia. Pathological staging was defined as pT3 pN0, according to the 9th edition of the AJCC.

Figure 1

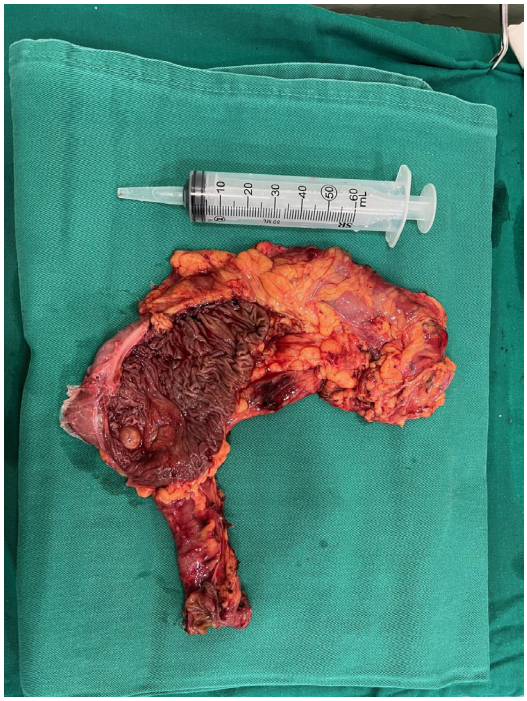

Figure 1: Surgical specimen after right videolaparoscopic colectomy, showing ileocolic segment and appendicular stump with nodular lesion in the cecal mucosa.

Figure 2

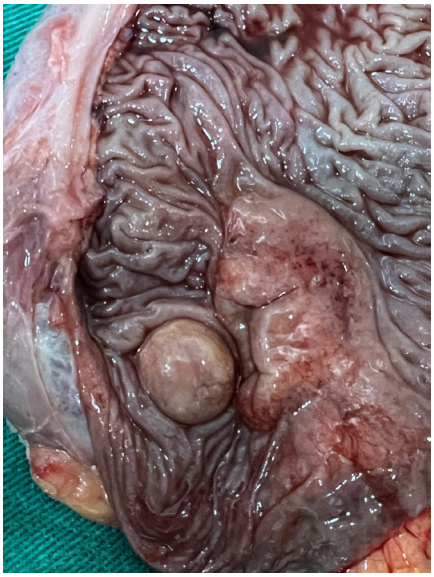

Figure 2: Detail of the mucosal surface of the cecum, showing a subepithelial nodular lesion in the appendicular stump.

Discussion

Neuroendocrine tumors of the appendix are mostly well differentiated and have a good prognosis, especially when they are smaller than 2 cm[1,2]. Location in the appendicular stump is uncommon and may result from appendicular residue inadvertently preserved during appendectomy or from tumor development in residual neuroendocrine tissue [(3,4)] .In the case in question, the diagnosis occurred incidentally during a colonoscopic examination, reinforcing the relevance of complete endoscopic evaluation in the investigation of endometrial changes, especially in elderly and asymptomatic patients, in addition to routine screening for colorectal cancer starting at age 45.

The recommended treatment for appendicular NETs with a diameter ≥ 2 cm or with histopathological risk factors comprises right colectomy with associated lymph node resection [4] . According to the guidelines of the European Neuroendocrine Tumor Society (ENETS) 2023, right hemicolectomy is indicated for tumors > 2 cm, compromised surgical margins (R1/R2), or the presence of aggressive features, with the procedure being optional for tumors between 1 and 2 cm based on additional factors, such as angiolymphatic invasion, high histological grade, or subserosal invasion (4) .

Recent studies, including series by Muñoz de Nova et al. (2022) and Holmager et al. (2021), corroborate the importance of adequate staging and complete resection in these cases, showing that optimized surgical management favors survival and locoregional control of the disease [5,6] . Additionally, contemporary publications, such as those by Butz , et al. (2024), and Ritter et al. (2025), discuss the effectiveness of the laparoscopic

approach and the possibility of individualizing the approach in tumors measuring 1–2 cm, according to risk profile and shared multidisciplinary decision [7,8]. In the present case, the tumor size of 2 cm associated with the presence of perineural invasion fully justified the performance of laparoscopic hemicolectomy, with adequate postoperative evolution.

Conclusion

Although rare, the possibility of a neuroendocrine tumor in the appendicular stump should be considered when incidental lesions are identified in the cecal region, even in patients who have previously undergone appendectomy. Appropriate individualized surgical treatment, with careful assessment of the need for oncological resection and lymph node evaluation, is essential to ensure a favorable prognosis. In addition, periodic follow-up is essential, considering the potential for late recurrence in residual sites. This report reinforces the importance of thorough endoscopic investigation and appropriate staging in the management of unusual cases such as this one.

References

1. Modlin IM, Lye KD, Kidd M. A 5-decade analysis of 13,715 carcinoid tumors. Cancer. 2003;97(4):934–959. doi:10.1002/cncr.11105

2. Pape UF, Niederle B, Costa F, Gross D, Kelestimur F, Kianmanesh R, et al. ENETS Consensus Guidelines for the management of patients with neuroendocrine neoplasms of the appendix. Neuroendocrinology. 2016;103(2):144–152. doi:10.1159/000443168

3. Gustafsson BI, Helland MJ, Tommeras K, Nordrum IS, Osnes T, Waldum HL. Appendiceal neuroendocrine neoplasms: incidence, survival and prognostic factors. Cancer. 2008;113(5):993–1001. doi:10.1002/cncr.23611

4. Kaltsas G, Walter T, Knigge U, Toumpanakis C, Santos AP, et al. European Neuroendocrine Tumor Society (ENETS) 2023 guidance paper for appendiceal neuroendocrine tumours (aNET). J Neuroendocrinol. 2023;35(10):e13332. doi:10.1111/jne.13332

5. Muñoz de Nova JL, Vázquez A, de la Portilla F, Santoyo J, et al. Management of incidentally discovered appendiceal neuroendocrine tumors after an appendicectomy. World J Gastroenterol. 2022;28(22):2413–2425. doi:10.3748/wjg.v28.i22.2413

6. Holmager P, Lykke J, Schultz NA, Qvist N, Mortensen MB, Achiam MP, et al. Neuroendocrine neoplasms of the appendix: characterization of 335 patients and evaluation of prognostic factors. Eur J Surg Oncol. 2021;47(4):917–922. doi:10.1016/j.ejso.2020.10.014

7. Butz F, Stremmel C, Köhler F, Tschuor C, Kahlert C, Ulrich A, et al. Current treatment options in neuroendocrine tumors of the appendix. Visc Med. 2024;40(1):16–23. doi:10.1159/000543185

8. Ritter AS, Meijer LL, van Eeden S, Boerma D, de Herder WW, et al. Novel surgical initiatives in gastroenteropancreatic neuroendocrine tumours: are we overtreating aNETs? Curr Oncol Rep. 2025;26(3):128–136. doi:10.1007/s11912-024-01632-4