


CASE REPORT

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# Neuroendocrine tumor of the appendix masquerading as acute appendicitis with a mucocele on CT scan: a rare finding

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## Abstract

**Introduction** Neuroendocrine tumors (NETs) of the appendix are rare and are often discovered incidentally during surgery for acute appendicitis or other unrelated conditions (Modlin et al. in *Gastroenterology* 128:1717–1751, 2005, Alsaad et al. in *Oncol Rep* 16:1105–1109, 2006, Frilling et al. in *Lancet Oncol* 15:e8–e21, 2014). These tumors can range from asymptomatic incidental findings to clinically significant tumors with metastases (Alsaad et al. in *Oncol Rep* 16:1105–1109, 2006, Gomes et al. in *World J Emerg Surg* 10:60, 2015, Paiva et al. in *Eur J Cancer* 38:702–705, 2002, Burke et al. in *Am J Surg Pathol*. 9:661–674, 1985). This case report presents a rare case of a NET of the appendix presenting as acute appendicitis.

**Case description** A 23-year-old male presented with right lower quadrant abdominal pain, nausea, and vomiting for 2 days. A CT scan revealed a mucocoele of the appendix. The patient underwent laparoscopic appendectomy, and the appendix was sent for histopathological examination. The final pathological report confirmed a NET of the appendix with a Ki-67 index of 1% and no lymphovascular invasion. Due to tumor invasion to the cecum and its large size (3–4 cm), the patient underwent right hemicolectomy. The final histopathology report of the resected specimen confirmed the diagnosis of NET of the appendix.

**Discussion** The clinical diagnosis of NETs of the appendix can be challenging due to their rarity and non-specific presentation. Symptoms of NETs of the appendix can mimic those of acute appendicitis, making it difficult to differentiate between the two conditions. Imaging studies, such as CT scans, can provide valuable information about the size and location of the tumor (Gomes et al. in *World J Emerg Surg* 10:60, 2015, Maggard et al. in *Ann Surg* 240:117–122, 2004, Burke et al. in *Am J Surg Pathol*. 9:661–674, 1985, Frilling et al. in *Lancet Oncol* 15:e8–e21, 2014). However, the definitive diagnosis is made through histopathological examination of the resected specimen. The treatment of NETs of the appendix depends on factors such as the size, location, and grade of the tumor. Small tumors confined to the appendix with no lymph-vascular invasion can be treated with appendectomy alone, while larger tumors or those that have spread beyond the appendix may require more extensive surgery, such as right hemicolectomy (Gomes et al. in *World J Emerg Surg* 10:60, 2015, Mestier et al. in *Dig Liver Dis* 52:899–911, 2020, Maggard et al. in *Ann Surg* 240:117–122, 2004, Burke et al. in *Am J Surg Pathol*. 9:661–674, 1985, Frilling et al. in *Lancet Oncol* 15:e8–e21, 2014, Pavel et al. in *Neuroendocrinology* 103:172–185, 2016). In some cases, additional treatments such as chemotherapy or radiation therapy may be recommended.

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**Conclusion** This case report emphasizes the importance of considering NETs of the appendix in the differential diagnosis of acute appendicitis. Imaging studies can provide valuable information, but the definitive diagnosis is made through histopathological examination. The treatment approach for NETs of the appendix depends on various factors and requires a multidisciplinary approach for optimal management.

**Keywords** Mucocele, Appendix, Neuroendocrine tumor, Laparoscopy, Appendectomy, Right hemicolectomy

## Introduction

NETs of the appendix are uncommon and usually found incidentally during surgery for acute appendicitis or other unrelated conditions [1, 2]. They can range from asymptomatic incidental findings to clinically significant tumors with metastases [2, 3]. Here, we present a rare case of a NET of the appendix presenting as acute appendicitis.

## Case description

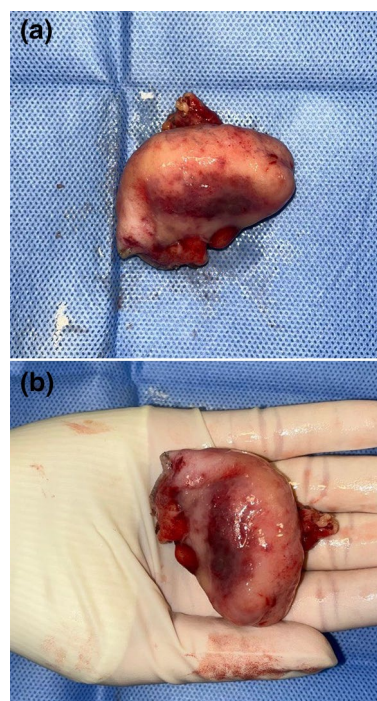
A 23-year-old male presented to the emergency department with a 2-day history of right lower quadrant abdominal pain, nausea, and vomiting. On examination, there was tenderness in the right lower quadrant with rebound tenderness, no guarding with normal vital signs. A CT scan showed a mucocele of the appendix (Fig. 1). The patient underwent laparoscopic appendectomy, and the appendix was sent for histopathological examination. The final pathological report revealed a NET of the appendix with a Ki-67 index of 1% and no lymphovascular invasion, but invasion of the base of cecum with positive margins, size (3–4 cm) and grade.

The patient was subsequently based on histopathological findings the patient was referred for right hemicolectomy. The final histopathology report of the resected specimen rt hemi confirmed the diagnosis of NET of the appendix (Fig. 2).

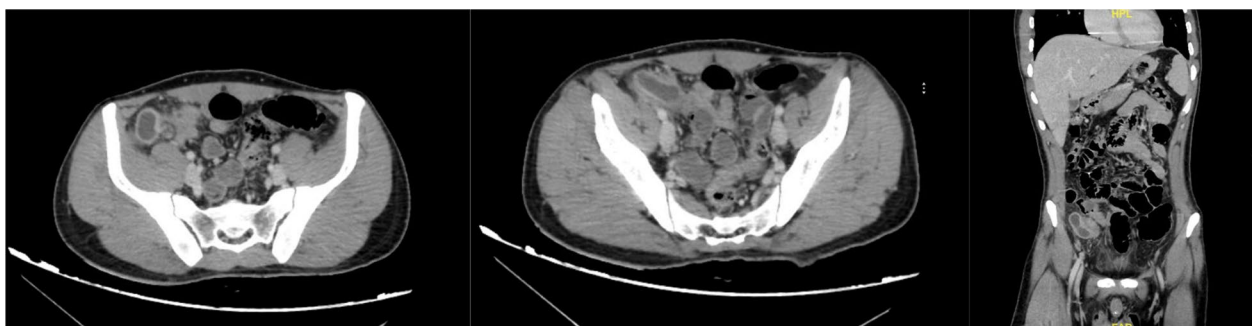
## Intra-operative approach and findings

A 12-mm trocar was placed through the supra-umbilical incision to approach the intraperitoneal cavity using the open Hasson technique. A pneumoperitoneum was made

by the insufflation of carbon dioxide. The table was kept in Trendelenburg position with a 15° left tilt. A 30° telescope was introduced through the umbilical port for diagnostic laparoscopy, and complete abdominal examinations were done. Diagnostic laparoscopy revealed approximately 3–4 cm large mucocele of the appendix.



**Fig. 2** Gross appearance of the resected appendix



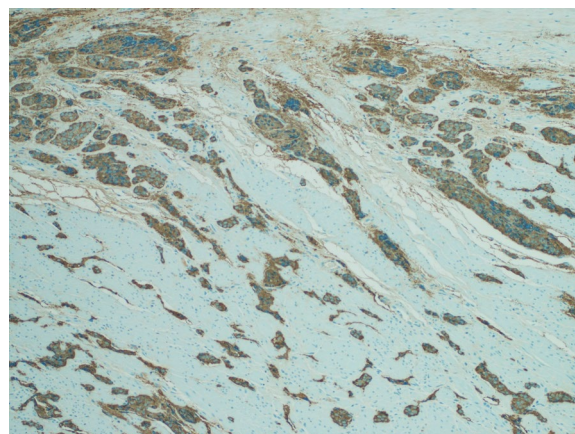
**Fig. 1** Axial, coronal CT appearance of the appendix



Two 5- and 10-mm ports were placed in the left upper quadrant and the suprapubic area. The appendiceal artery was isolated after separating the mesoappendix with the help of bipolar cautery (ligasure). Following this, the base of the appendix was ligated at the ileocecal junction and divided by using an endoscopic stapling device (Multifire Endo GIA, 60 mm) which was used to perform the partial resection of cecum. The appendix was retrieved in a plastic bag through the umbilical port after careful minimal handling. Hemostasis was obtained. The umbilical port site wound was closed with j (needle). The patient tolerated the procedure, he started oral feeding 6 h post-operation and solid food on the next day. He was discharged on the third postoperative day without active complaint. Pathology showed the final report revealed a NET of the appendix with a Ki-67 index of 1% and no lymphovascular invasion, but invaded the base of cecum with positive margins (Figs. 3, 4, 5).

### Discussion

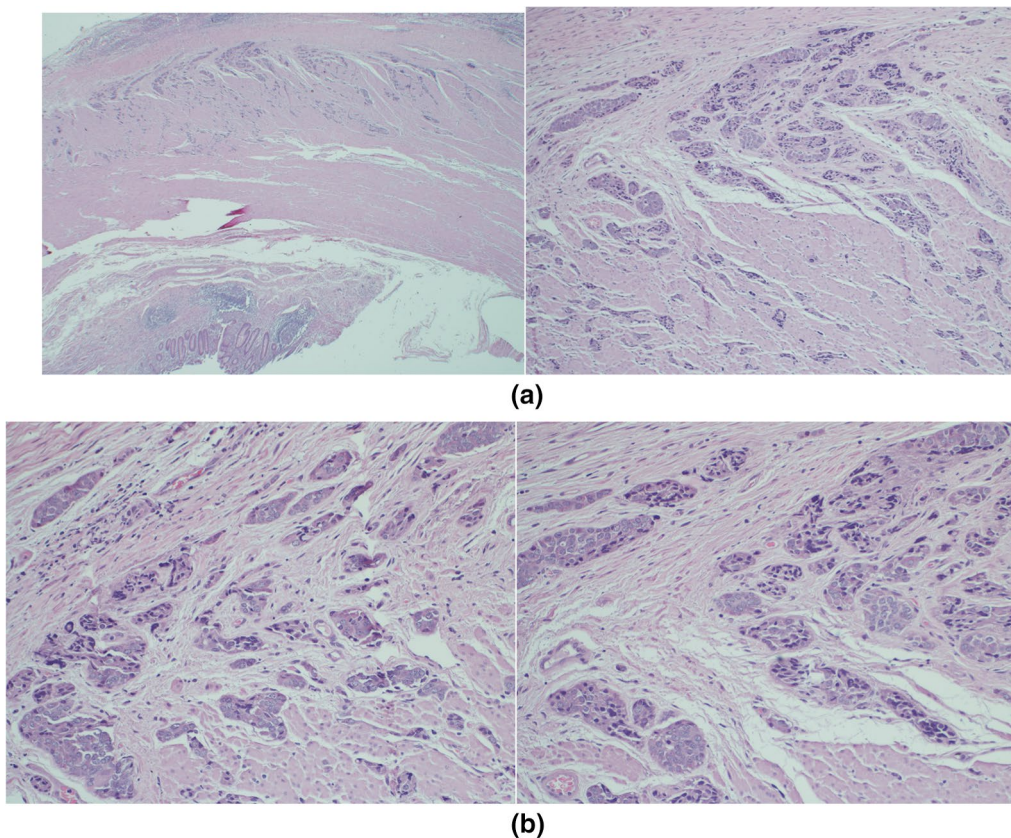
The diagnosis of NETs of the appendix is challenging due to the rarity of these tumors and their non-specific presentation. The symptoms of NETs of the appendix can



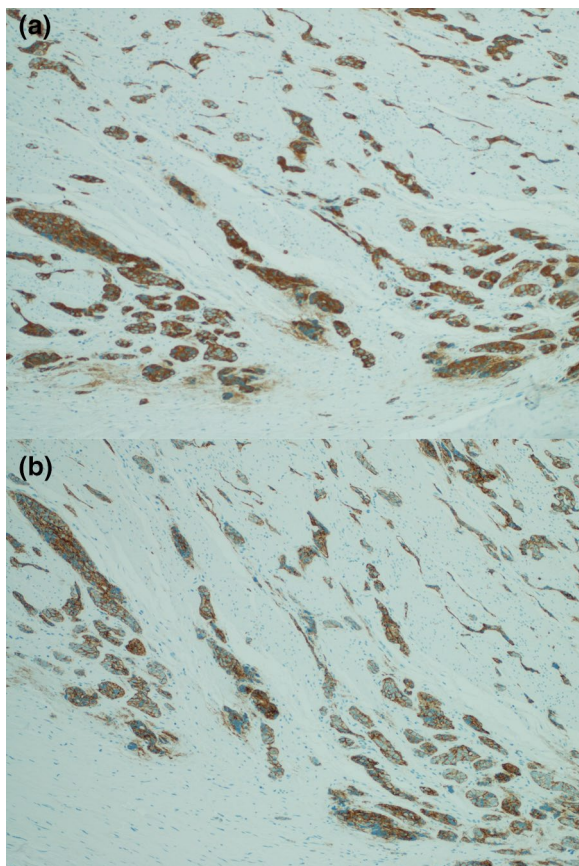
**Fig. 4** Shows diffusely strong positive staining for chromogranin

mimic those of acute appendicitis, making it difficult to distinguish between the two conditions. Imaging studies such as CT scans can provide valuable information regarding the size and location of the tumor [4, 5].

Low-grade appendiceal mucinous neoplasm (LAMN) typically appears as a low-attenuation cystic lesion on CT



**Fig. 3** **A** This high-power view shows the tumor invading muscularis propria. **B** High magnification shows polygonal tumor cells arranged in nests and glandular growth pattern. Cells are monomorphic with round nuclei and finely stippled chromatin



**Fig. 5** a, b Shows diffusely strong positive synaptophysin and diffusely strong positive CD56

scans, often accompanied by calcifications and wall thickening [4–6]. On the other hand, neuroendocrine tumors (NETs) of the appendix may present as solid masses with enhancement on contrast-enhanced CT scans [5, 6]. However, it is important to note that these imaging features are not always definitive for distinguishing between LAMN and NETs. Histopathological examination of the resected specimen remains crucial for accurate diagnosis [1, 2, 5, 6]. However, the definitive diagnosis of NET of the appendix is made by histopathological examination of the resected specimen [7, 8].

The treatment of NETs of the appendix depends on the size, location, and grade of the tumor. Small tumors that are confined to the appendix with no lymphovascular invasion can be treated with appendectomy alone, whereas larger tumors or those that have spread beyond the appendix may require more extensive surgery, such as right hemicolectomy [9, 10]. In some cases, additional treatments such as chemotherapy or radiation therapy may also be recommended [11].

When the appendectomy margins are positive for tumor involvement, it is generally recommended to

perform additional resection to achieve clear surgical margins. However, in this case, the decision for right hemicolectomy was made based on the invasion of the tumor into the cecum and the large size of the tumor (3–4 cm). The aim was to achieve complete resection and ensure adequate oncological clearance. The decision for right hemicolectomy was made in consultation with the multidisciplinary team, considering the extent of tumor involvement and the desire for optimal management [8, 12, 13].

Multidisciplinary treatment is important in cases where the size, location, and grade of the tumor warrant a comprehensive approach. Larger tumors or tumors with evidence of lymph node involvement, distant metastases, or invasion into adjacent structures often require a multidisciplinary team approach [7, 8, 12]. Collaboration between surgeons, oncologists, and radiologists can help determine the most appropriate treatment strategy, which may involve a combination of surgical resection, chemotherapy, and/or radiation therapy. Involving specialists in gastrointestinal oncology can provide valuable input in managing these complex cases [8, 14].

## Conclusion

This case report highlights the importance of considering NETs of the appendix in the differential diagnosis of acute appendicitis. Imaging studies such as CT scans can provide valuable information, but the definitive diagnosis of NET of the appendix is made by histopathological examination of the resected specimen. The treatment of NETs of the appendix depends on the size, location, and grade of the tumor. A multidisciplinary approach is essential for the optimal management of these rare tumors.

## Abbreviations

NETs	Neuroendocrine tumors
LAMN	Low-grade appendiceal mucinous neoplasm

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## Author contributions

All authors were responsible for patient care and writing, editing the manuscript.

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## Availability of data and materials

The datasets used during the current study are available from the corresponding author upon request.



## Declarations

### Ethics approval and consent to participate

Does not require as its case report and all information were de-identified and collected retrospectively.

### Consent for publication

Written informed consent was obtained from the patient for this study. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request. The information in this case report are completely de-identified.

### Competing interests

No competing interests.

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## References

- Modlin IM, Kidd M, Latich I, Zikusoka MN, Shapiro MD. Current status of gastrointestinal carcinoids. *Gastroenterology*. 2005;128(6):1717–51. <https://doi.org/10.1053/j.gastro.2005.03.038>.
- Alsaad KO, Serra S, Schmitt A, et al. Appendiceal adenocarcinomas and carcinoid tumors: clinicopathological review of 184 cases. *Oncol Rep*. 2006;16(5):1105–9. <https://doi.org/10.3892/or.16.5.1105>.
- Gomes CA, Junior CS, Di Saverio S, et al. Acute appendicitis: proposal of a new comprehensive grading system based on clinical, imaging and laparoscopic findings. *World J Emerg Surg*. 2015;10:60. <https://doi.org/10.1186/s13017-015-0055-2>.
- de Mestier L, Walter T, Brix H, et al. Digestive neuroendocrine neoplasms (NEN): French intergroup clinical practice guidelines for diagnosis, treatment and follow-up (SNFGE, GTE, RENATEN, TENPATH, FFCD, GERCOR, UNICANCER, SFCD, SFED, SFRO, SFR). *Dig Liver Dis*. 2020;52(8):899–911. <https://doi.org/10.1016/j.dld.2020.05.010>.
- Maggard MA, O'Connell JB, Ko CY. Updated population-based review of carcinoid tumors. *Ann Surg*. 2004;240(1):117–22. <https://doi.org/10.1097/01.sla.0000129358.51409.8d>.
- McCusker ME, Cote TR, Clegg LX, Sobin LH. Primary malignant neoplasms of the appendix: a population-based study from the surveillance, epidemiology and end-results program, 1973–1998. *Cancer*. 2002;94(12):3307–12. <https://doi.org/10.1002/cncr.10589>.
- Sobin LH, Gospodarowicz MK, Wittekind C. TNM classification of malignant tumours. 7th ed. USA: Wiley-Blackwell; 2010.
- Burke AP, Thomas RM, Elsayed AM, Sobin LH. Carcinoids of the appendix. A clinicopathologic study of 107 cases. *Am J Surg Pathol*. 1985;9(9):661–74. <https://doi.org/10.1097/0000478-198509000-00001>.
- Paiva CE, Marques MP, Silva AR, et al. Ki-67 labelling index and the prognosis of gastrointestinal carcinoid tumours. *Eur J Cancer*. 2002;38(5):702–5. [https://doi.org/10.1016/s0959-8049\(01\)00431-6](https://doi.org/10.1016/s0959-8049(01)00431-6).
- Frilling A, Modlin IM, Kidd M, et al. Recommendations for management of patients with neuroendocrine liver metastases. *Lancet Oncol*. 2014;15(1):e8–21. [https://doi.org/10.1016/S1470-2045\(13\)70362-0](https://doi.org/10.1016/S1470-2045(13)70362-0).
- Pavel M, O'Toole D, Costa F, et al. ENETS consensus guidelines update for the management of distant metastatic disease of intestinal, pancreatic, bronchial neuroendocrine neoplasms (NEN) and NEN of unknown primary site. *Neuroendocrinology*. 2016;103(2):172–85. <https://doi.org/10.1159/0004431711>.
- Pape UF, Perren A, Niederle B, et al. ENETS consensus guidelines for the management of patients with neuroendocrine neoplasms from the jejunum-ileum and the appendix including goblet cell carcinomas. *Neuroendocrinology*. 2012;95(2):135–56. <https://doi.org/10.1159/000335587>.
- Kulke MH, Shah MH, Benson AB, et al. Neuroendocrine tumors, version 1.2015. *J Natl Compr Canc Netw*. 2015;13(1):78–108. <https://doi.org/10.6004/jnccn.2015.0012>.
- Pickhardt PJ, Levy AD, Rohrmann CA Jr, Kende AI. Primary neoplasms of the appendix manifesting as acute appendicitis: CT findings with pathologic correlation. *Radiology*. 2002;224(3):775–81. <https://doi.org/10.1148/radiol.2243011060>.

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