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An Incidental Appendiceal Neuroendocrine Tumor Discovered After Appendectomy: A Case Report and Review of the Literature

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Abstract **Case Report**

Appendicular neuroendocrine tumours (aNETs) are the most common type of appendicular neoplasm and the fifth most common neuroendocrine tumour in the gastrointestinal tract. They are most often discovered incidentally during appendectomy performed for acute appendicitis. They are generally characterised by indolent progression and an excellent prognosis, particularly when they are well differentiated and small in size. We report the case of a 40-year-old female patient who underwent emergency surgery for acute appendicitis. Pathological examination of the surgical specimen revealed a well-differentiated appendicular neuroendocrine tumour, classified as G2 pT2. Postoperative management was discussed taking into account the histopathological characteristics and recent recommendations.

Keywords: Appendicular Neuroendocrine Tumour, Acute Appendicitis, Histological Grade, Prognostic Factors, Surgery.

INTRODUCTION

Appendicular neuroendocrine tumours (aNETs) represent a distinct entity within digestive neuroendocrine neoplasms, characterised by their often incidental discovery, non-specific clinical presentation and generally indolent progression. Although their incidence is low, the increased use of imaging and appendectomy has led to more frequent identification of these lesions, often at an early stage [1, 2]. However, the recognition of prognostic histopathological factors, such as tumour grade, depth of parietal invasion and locoregional spread, has highlighted the need for rigorous assessment in order to adapt staging, therapeutic strategy and post-operative follow-up [2, 3].

CLINICAL CASE

A 48-year-old female patient presented to the emergency department with symptoms suggestive of uncomplicated acute appendicitis. Preoperative history and clinical examination did not reveal any evidence of underlying tumour pathology. No functional symptoms suggestive of hormonal hypersecretion, particularly carcinoid syndrome, were reported. The patient remained apyretic and her general condition remained stable. An emergency appendectomy was performed without complications. Pathological examination of the surgical specimen revealed a well-differentiated grade 2 appendicular neuroendocrine tumour measuring 4 mm in its longest axis. The lesion was pan-parietal with focal infiltration of the subserosa less than 3 mm, corresponding to stage pT2. Immunohistochemistry showed expression of synaptophysin and chromogranin A, with an estimated Ki-67 index of 5%. In addition, the appendix showed lesions of acute suppurative pan-appendicitis with peritoneal involvement. As part of the post-operative assessment, functional imaging by Octreoscanner was performed. It revealed intense fixation of the thyroid gland, predominantly in the left thyroid lobe, consistent with a plunging goitre, as well as a left parauterine tissue mass measuring 5.8 × 5 cm. Further examination revealed no scintigraphic evidence of residual or recurrent disease, nor any visceral bone, cerebral, pulmonary or hepatic metastases. The assessment was completed with a total colonoscopy, which revealed no pathological lesions, in particular no ulceration, erythema, polyps or tumours. The serum chromogranin A level was also within normal limits. The patient then underwent, in separate operations, a left lobectomy, performed after ruling out a medullary thyroid origin due to negative calcitonin levels, and surgical resection of the left lateral uterine mass. Pathological examination, completed by immunohistochemical analysis, found no evidence of neuroendocrine origin, signs of malignancy, or

histological criteria for malignancy in either of these two lesions.

DISCUSSION

Appendicular neuroendocrine neoplasms (aNEN) are relatively common gastrointestinal neuroendocrine tumours, with an approximate annual incidence of 0.15 to 0.6/100,000 inhabitants, preferentially affecting young individuals (average age at diagnosis between 38 and 51 years), with a slight female predominance (2:1) according to certain Western series [1-6]. They are most often discovered incidentally during an appendectomy performed for suspected acute appendicitis, which explains their generally early diagnosis [1, 2].

Their diagnosis is almost always incidental, made during an appendectomy performed for another indication, most often suspected or confirmed acute appendicitis, more rarely during other abdominal surgery, with an estimated incidence of 3 to 5 cases per 1,000 appendectomies [1-7]. Due to this incidental discovery, aNENs are generally asymptomatic. They are preferentially located at the distal end of the appendix in nearly 70% of cases, while 5 to 20% are located in the middle third and less than 10% at the base, which explains the rarity of obstructive phenomena [8]. Furthermore, the hormonal secretion responsible for carcinoid syndrome is exceptional (< 1% of cases) and is only observed in the presence of metastatic disease [9].

Clinically, aNENs are most often asymptomatic and rarely accompanied by functional manifestations; carcinoid syndrome is rare due to the small size of the tumour and the usual absence of liver metastases at diagnosis [1]. The clinical signs observed are therefore generally related to the appendicitis itself rather than to the tumour.

In terms of paraclinical findings, preoperative imaging is less helpful, as the lesions are often only a few millimetres in size and cannot be identified on CT scans. The diagnosis is based primarily on pathological examination of the appendectomy specimen, which allows the tumour size, depth of parietal invasion, extension to the mesoappendix and histological grade according to the World Health Organisation classification to be determined. These parameters are the main prognostic and decision-making factors for staging and subsequent management, in accordance with European recommendations [2, 3].

The staging of NETs differs between the American Joint Committee on Cancer (AJCC)/Union for International Cancer Control (UICC) and European Neuroendocrine Tumor Society (ENETS) classifications in their definitions of T stages (Table 1) [3-10]. According to the National Comprehensive Cancer Network (NCCN) guidelines, patients with a tumour ≥ 2 cm or any size tumour with incomplete resection or positive lymph nodes should undergo multiphase abdominal/pelvic computed tomography (CT) or magnetic resonance imaging (MRI) to rule out locoregional or distant metastasis [11]. Lymph node metastases are reported in approximately 2.5% of tumours < 1 cm, in 31% of tumours > 1 cm but < 2 cm, and in 64% of tumours ≥ 2 cm [12].

Tableau 1: TNM classification for aNET. Changes in pTNM pathological classifications over time (ENETS, AJCC/UICC 2009 seventh edition, AJCC/UICC 2017 eighth edition) [3]

	pTNM ENETS	AJCC/UICC seventh edition	AJCC/UICC eighth edition
pT1	T ≤ 1 cm and submucosa or muscularis propria invasion	T ≤ 2 cm (T1a ≤ 1 cm; T1b > 1–2 cm)	T ≤ 2 cm
pT2	T ≤ 2 cm and submucosa or muscularis propria or mesoappendix/subserosa invasion ≤3 mm	T > 2–4 cm OR Caecal invasion	T > 2–4 cm
pT3	T > 2 cm and/or mesoappendix/subserosa invasion >3 mm	T > 4 cm OR Ileal invasion	T > 4 cm OR Mesoappendix/ subserosa invasion
pT4	Perforates serosa/peritoneum, or invades other neighbouring organs		

Note: T, size in greatest dimension.
Abbreviation: aNET, appendiceal NET.

In terms of surgical management, the ENETS 2023 guidelines recommend isolated appendectomy for well-differentiated aNETs smaller than 1 cm without major unfavourable histopathological criteria. Right hemicolectomy is reserved for tumours ≥ 2 cm in size or those with high risk factors, such as significant extension to the mesoappendix, lymph node involvement or an unfavourable histological grade [3]. For tumours of intermediate size (1–2 cm) or those classified as pT2 solely on the basis of parietal invasion, the recommendations insist on the need for individualised assessment, ideally discussed in a multidisciplinary consultation meeting.

However, the real prognostic value of isolated subserosal infiltration in very small aNETs remains

controversial. Several studies suggest that, in the absence of other poor prognostic factors — notably lymph node involvement, significant extension to the mesoappendix or a high histological grade — the risk of progression remains extremely low, even in cases of pT2 staging [13].

Nevertheless, the reported case highlights a situation frequently encountered in clinical practice: the apparent discordance between a millimetric tumour size and pT2 staging. This classification is based on the depth of parietal invasion, in particular the involvement of the subserosa, regardless of tumour diameter. The ENETS 2023 recommendations reiterate that the staging of aNET should not be limited to size, but should include essential anatomopathological parameters, such as invasion of the appendicular wall and extension to the mesoappendix, which are thought to reflect an increased potential for locoregional dissemination [3].

The follow-up of appendicular neuroendocrine tumours must be individualised according to the type of surgery and the definitive histopathological characteristics. In accordance with the recommendations of the European Neuroendocrine Tumor Society, tumours < 1 cm completely resected by simple appendectomy (R0), as well as tumours > 1 cm treated by right hemicolectomy without lymph node involvement or residual disease, do not require specific follow-up. However, regular follow-up is recommended for tumours measuring 1–2 cm associated with histopathological risk factors (basal location, mesoappendicular invasion > 3 mm, grade 2 or lymphovascular invasion), while tumours > 2 cm, forms with lymph node involvement, metastatic disease or incomplete resection warrant prolonged follow-up. Biological markers and routine imaging are not validated for routine use; MRI is preferred in young patients in order to limit radiation exposure, while CT, possibly combined with somatostatin receptor imaging, is reserved for high-risk situations or in cases of suspected recurrence [2, 3].

Conclusion

Appendiceal neuroendocrine tumours (ANETs) are relatively rare entities, most often discovered incidentally, and are generally associated with a favourable prognosis, particularly when they are well differentiated and small in size. Nevertheless, their malignant potential, although limited, justifies rigorous and systematic pathological analysis of all appendectomy specimens. A thorough histological examination not only allows the identification of lesions that are sometimes subclinical or millimetric in size, but also enables the assessment of essential prognostic parameters such as depth of invasion, histological grade and locoregional extension, which are determining factors for staging and guiding therapeutic management [2, 3].

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