

## Case series

### **Incidental neuroendocrine tumor of the appendiceal base less than 20 mm in diameter: is appendectomy enough?**

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#### **Abstract**

The appendix is the second primary site for neuroendocrine tumors. The management of incidentally discovered neuroendocrine tumor of the appendiceal base less than 20 mm in diameter is still controversial. The aim of this study was to discuss the management of such tumors. Three patients were operated on for acute appendicitis. Histopathologic examination of surgery specimens revealed neuroendocrine tumors of the appendiceal base less than 20 mm in diameter. Since no one presented with poor prognostic factors, no complementary right hemicolectomy was performed. No recurrence was observed. The existence of poor prognostic factors at histopathologic examination should indicate complementary right hemicolectomy for incidental neuroendocrine tumor of the appendiceal base less than 20 mm in diameter.

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## Introduction

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The appendix counts for the second primary site of neuroendocrine tumors occurring in 25 to 30% of the cases [1,2]. These tumors are located in the base of the appendix in only 10% of the cases. They are discovered incidentally after appendectomy for acute appendicitis [3]. The management following appendectomy still controversial especially for tumours of less than 20 mm in diameter. The aim of this study was to discuss the management for incidentally histopathologic discovered neuroendocrine tumors of the appendiceal base with a diameter less than 20mm.

## Methods

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The case series included three patients, two females and one male. The ages at diagnosis were 38 years, 42 years, and 45 years. All three patients were admitted for acute appendicitis. None suffered from the carcinoid syndrome. Appendectomy was performed laparoscopically in two cases and by elective laparotomy in one patient. Per operatively, The macroscopic aspects of the appendix were gangrenous in one patient and phlegmonous in two cases. Appendectomy was performed with uneventful course. The histological analysis of the surgical specimens revealed the neuroendocrine tumors.

## Results

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The three tumors were located at the base of the appendix, well-differentiated, without cellular necrosis nor vascular invasion. The tumor sizes were 5mm, 7mm, and 15 mm. Mitotic figures were 2 mitosis per 10 high-power fields in one patient, 3 in one case, and 4 in the other one. Proliferative activity Ki-67 was nil in two cases and 2% in one patient. Microscopic invasion was limited to the submucosa, the muscosa, and the subserosa in one case each. The surgical margins were negative for tumor cells. No mesoappendiceal involvement was found. Postoperative computerized tomography of the abdomen didn't demonstrate metastases. The somatostatin receptor scintigraphy was normal. The secretion of 5-Hydroxy-Inndole-Acetic Acid measured after a 24-hour collection of urine was normal in all patients. No poor prognostic factors were found in our patients thus

complementary right hemicolectomy wasn't carried out. No relapse was diagnosed after a follow up of 12 months.

## Discussion

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As for our patients, appendectomy seems enough for neuroendocrine tumors of the appendiceal base less than 20 mm in diameter unless poor prognostic factors are present. Our results are in agreement with the literature. The appendiceal neuroendocrine tumors are discovered at a younger age than the other sites with a mean age of 42 years [4] as in our cases. It may be secondary to incidental diagnosis during appendectomy that occurs more frequently in younger patients. They are more frequent in women with a sex-ratio of 0,5 [5-7]. No specific clinical presentation is described [8]. Abdominal pain represents the most common complaint [5,9]. Association with a carcinoid syndrome occurs in only 1% of cases [6]. Several factors are taken into account before deciding to perform complementary surgery. The most considered factor is the tumor size [2]. Since tumors greater than 20 mm in diameter increase the incidence of metastatic spread ranging from 20% to 85% [10,11], they should be managed with a formal right hemicolectomy [2]. For those with a diameter less than 20 mm, appendectomy is enough unless another poor prognostic factor is found. The indications for a complementary surgery include histological evidence of mesoappendiceal extension [12], tumor at the base of the appendix with positive margins or involvement of the caecum [13], high-grade malignant carcinoid tumor with a raised tumor prognostic index as measured by mitotic index and Ki-67 levels [12], lymph node involvement, and cellular pleomorphism with a high mitotic index [14]. The appendiceal neuroendocrine tumors have a good prognosis with 90,3% 5-year disease-specific survival [9].

## Conclusion

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Appendectomy is enough for incidental neuroendocrine tumor of the appendiceal base less than 20 mm in diameter unless poor prognostic factors at histopathologic examination are present.

## Competing interests

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The authors declare no competing interests.

## Authors' contributions

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All authors have read and agreed to the final version of this manuscript and have equally contributed to its content and to the management of the case.

## References

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1. Niederle MB, Hackl M, Kaserer K, Niederle B. Gastroenteropancreatic neuroendocrine tumours: the current incidence and staging based on the WHO and European Neuroendocrine Tumour Society classification: an analysis based on prospectively collected parameters. *Endocrine-Related Cancer*. 2010;17(4):909-918. **PubMed | Google Scholar**
2. Deschamps L, Couvelard A. Endocrine Tumors of the Appendix A Pathologic Review. *Arch Pathol Lab Med*. 2010;134(6):871-875. **PubMed | Google Scholar**
3. Sandor A, Modlin IM. A retrospective analysis of 1570 appendiceal carcinoids. *Am J Gastroenterol*. 1998;93(3):422-428. **PubMed | Google Scholar**
4. Modlin IM, Sandor A. An analysis of 8305 cases of carcinoid tumors. *Cancer*. 1997;79(4): 813-829. **PubMed | Google Scholar**
5. Estrozi B, Bacchi CE. Neuroendocrine tumors involving the gastroenteropancreatic tract: a clinico pathological evaluation of 773 cases. *Clinics*. 2011;66(10):1671-1675. **PubMed | Google Scholar**
6. Plöckinger U, Couvelard A, Falconi M et al. Consensus guidelines for the management of patients with digestive neuroendocrine tumours: well-differentiated tumour/carcinoma of the appendix and goblet cell carcinoma. *Neuroendocrinology*. 2008;87(1):20-30. **PubMed | Google Scholar**
7. Yao JC, Hassan M, Phan A et al. One hundred years after "carcinoid": epidemiology of and prognostic factors for neuroendocrine tumors in 35,825 cases in the United States. *J Clin Oncol*. 2008;26(18):3063-3072. **PubMed | Google Scholar**
8. In't Hof KH, van der Wal HC, Kazemier G, Lange JF. Carcinoid tumour of the appendix: an analysis of 1,485 consecutive emergency appendectomies. *J Gastrointest Surg*. 2008;12(8):1436-1438. **PubMed | Google Scholar**
9. Tsikitis TL, Wertheim BC, Guerrero MA. Trends of incidence and survival of gastrointestinal neuroendocrine tumors in the United States: A seer analysis. *Journal of Cancer*. 2012;3:292-302. **PubMed | Google Scholar**
10. Thompson GB, van Heerden JA, Martin JK Jr, Schutt AJ, Ilstrup DM, Carney JA. Carcinoid tumors of the gastrointestinal tract: presentation, management, and prognosis. *Surgery*. 1985;98(6):1054-1063. **PubMed | Google Scholar**
11. Mac Gillivray DC, Heaton RB, Rushin JM, Cruess DF. Distant metastasis from a carcinoid tumor of the appendix less than one centimeter in size. *Surgery*. 1992;111(4):466-471. **PubMed | Google Scholar**
12. Goede AC, Caplin ME, Winslet MC. Carcinoid tumour of the appendix. *Br J Surg*. 2003;90(11):1317-1322. **PubMed | Google Scholar**
13. Safioleas MC, Moulakakis KG, Kontzoglou K et al. Carcinoid tumors of the appendix. Prognostic factors and evaluation of indications for right hemicolectomy. *Hepatogastroenterology*. 2005;52(61):123-127. **PubMed | Google Scholar**

14. Fornaro R, Frascio M, Sticchi C et al. Appendectomy or right hemicolectomy in the treatment of appendiceal carcinoid tumors? *Tumori*. 2007;93(6):587-590. **Google Scholar**