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# An atypical type I gastric neuroendocrine tumor

ANDRADA-VIORELA GHEORGHE[1], MIHAI RIMBAS[1], OCTAV GINGHINA[2], ANDRADA SPANU[2], THEODOR ALEXANDRU VOIOSU[1]

[1]Department of Gastroenterology, "Colentina" Clinical Hospital, Bucharest, Romania
[2]Department of General Surgery, "Sfantul Ioan" Hospital, Bucharest, Romania

**Background**. Gastric neuroendocrine tumors (GI-NETs) are rare lesions, usually discovered incidentally during endoscopy. Based on their pathology, there are 4 types of GI-NETs. Type I are multiple small polypoid lesions with central ulceration located in the gastric body or the fundus, associated with atrophic gastritis usually noninvasive and very rarely metastatic. We report on a rare case of a gastric NET arising from the muscularis propria layer of the pyloric ring.

**Case report**. We present the case of a 65-year old woman with a history of alcoholic cirrhosis, investigated for melena. Upper endoscopy revealed a 30 mm submucosal pedunculated polypoid lesion located on the pylorus protruding in the duodenum, with normal overlying mucosa, fundic gastric atrophy and multiple small polyps at this level, with no active bleeding. CT scan did not reveal any distant metastases. An ultrasound endoscopy was performed, and a round hypoechoic heterogeneous solitary mass, evolving from the pyloric muscle was described. Considering a 30-mm tumor evolving from the gastric muscle layer in the absence of local invasion and with no distant metastases we decided against an endoscopical resection and we referred the patient to surgery. A laparoscopic wedge resection was performed. The pathology report described a 30/25 mm well-differentiated neuroendocrine tumor invasive in the muscularis mucosa (pT3).

**Conclusions**. Usually, type I neuroendocrine tumors are located in the body or the fundus of the stomach without submucosal invasion. The interesting feature in our case was that the tumor originated from the pylorus, making it an atypical presentation for a neuroendocrine tumor.

**Key words**: ultrasound endoscopy, gastric neuroendocrine tumor, surgery.

## INTRODUCTION

Gastric neuroendocrine tumors (GI-NETs) are rare lesions with a reported incidence of only 1 to 2 cases/1000000 [1]. Most gastric NETs are discovered incidentally during upper endoscopy [3]. In a study conducted on 13715 carcinoid tumors, during a 50 years' analysis, there was an increase in the incidence of gastric NETs from 2.25% in the first decades to 5.85% in the last decade of the study [2].

In 2010 World Health Organization released a new classification of gastrointestinal and pancreatic neuroendocrine tumors, dividing them based on the number of mitoses and expression of Ki67 index into low-grade (G1) NET, intermediate grade (G2) NET and high grade neuroendocrine carcinoma [4].

## CASE REPORT

A 65-year old woman with a history of alcoholic Child A cirrhosis was admitted in our unit with melena. Besides pallor, the clinical examination was normal. Laboratory studies showed severe anemia (Hb 6.1 g/dL), mild cholestasis (ALP 1.7xN, GGT 7xN) and an albumin level of 3.3 g/dL. The rest of the liver tests were within normal range. Following admission an upper endoscopy was performed and revealed a pedunculated polypoid submucosal lesion located on the pylorus protruding in the duodenum, with normal overlying mucosa (Figure 1).

The lesion was 30 mm in diameter and no active bleeding source was found. The rest of the antrum was normal, but the gastric fundic mucosa was atrophic and multiple small polyps (< 10 mm) were observed at this level. Multiple biopsies from fundic polyps and from the main lesion were taken. The initial pathology report revealed benign atrophic gastritis, with no evidence of *H. pylori.*

A CT scan of the abdomen revealed a mass involving the pylorus protruding into the duodenum bulbus, with perihepatic lymph nodes and no distant metastases (Figure 2). Endoscopic ultrasound (EUS) identified the same round hypoechoic heterogeneous solitary mass, with well-defined margins, evolving from the pyloric muscle (Figure 3), with no local invasion and an 18 mm perihepatic lymph node.


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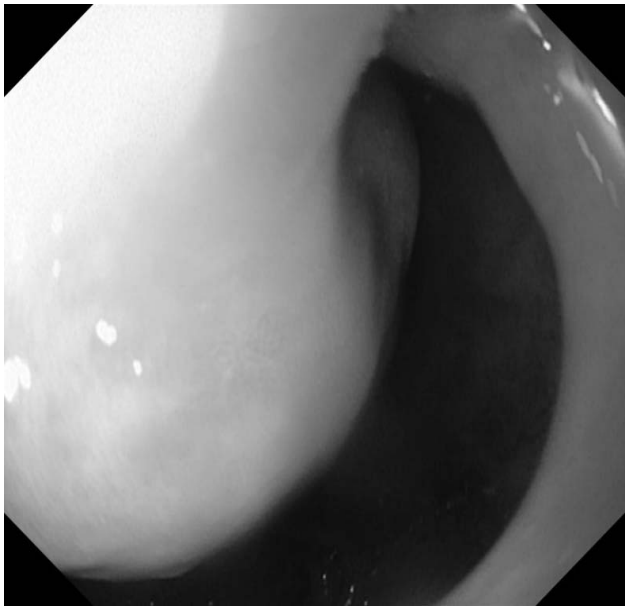

Figure 1. Pyloric submucosal tumor.

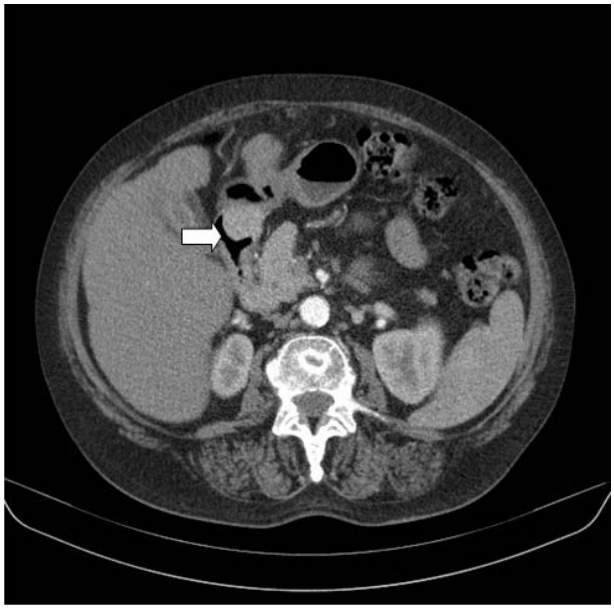

Figure 2. CT scan showing a pyloric lesion (arrow).

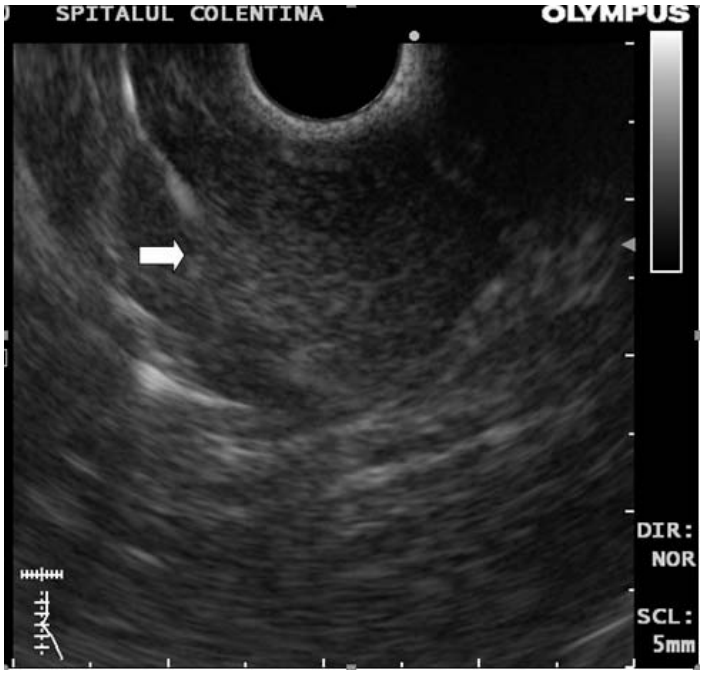


Figure 3. EUS aspect of the lesion (arrow).

Considering a 30-mm tumor evolving from the gastric muscle layer in the absence of local invasion and with no distant metastases we decided against an endoscopical resection and we referred the patient for surgery. A laparoscopic wedge resection was performed with no complications. The patient was discharged 3 days after the procedure. The pathology report described a 30/25 mm well-differentiated neuroendocrine tumor invasive in the muscularis mucosa (pT3).

## DISCUSSION

Based on their pathophysiology, there are 3 main categories of NETs: well differentiated gastrin-dependent, non-gastrin dependent or sporadic and poorly differentiated. There are 2 subgroups of gastrin-dependent neuroendocrine tumors: type I, commonly found in patients with atrophic gastritis and type II, associated with Zollinger-Ellison syndrome (ZES). Type III NETs

are well-differentiated carcinoid tumors that do not develop in the setting of atrophic gastritis or ZES [3]. Type IV are poorly differentiated neuroendocrine carcinomas [6].

Of all GI-NETs, type I is the most common, reaching almost 80% [5]. These tumors usually present as multiple small polypoid [5] or sessile [7] lesions with a central ulceration located in the gastric body or the fundus, associated with atrophic gastritis [5]. Type I tumors are usually noninvasive and very rarely metastatic [3]. For multiple small lesions (< 10-20 mm) with no evidence of infiltration of the muscular wall or angioinvasion endoscopic surveillance is the best strategy [8], otherwise endoscopic resection of the tumor or surgical resection are indicated depending on the size, lymph node invasion or number of lesions [5, 9].

In our case the localization of the tumor on the pyloric ring and the absence of *H. pylori* infection is unusual for a type I gastric NET, however the gastric mucosa was highly atrophic and multiple small fundic polyps were described at the upper endoscopy.

NETs are mainly diagnosed at upper GI endoscopy. EUS staging is becoming more and more popular among endoscopists, who choose to perform it before endoscopical mucosal resection (EMR) or endoscopical submucosal dissection [12]. EUS usually shows uniform, well-defined, hypoechoic tumors located in the deeper mucosa or the submucosa.

As discussed above, both types I and II can be managed endoscopically, either by EMR or by ESD depending on size. A study was conducted on 62 patients with type I gastric carcinoid tumors comparing the usefulness of ESD *versus* EMR with results that showed a higher resection rate for the ESD technique over EMR [13]. This may be because most gastric neuroendocrine tumors invade the submucosal layer [14]. Surgical resection with lymph node dissection is applied for sporadic well-differentiated and poorly-differentiated neuroendocrine tumors [5], in association with chemotherapy. In our case, because of the size of the tumor and since it was evolving from the muscle layer, we opted for surgical treatment.

## CONCLUSIONS

There is a significant increase in the incidence of gastric neuroendocrine tumors during the past years, highlighting the importance of understanding their nature and precisely chose the type-adapted treatment. Usually, type I neuroendocrine tumors are located in the body or the fundus of the stomach, lacking submucosal invasion. The interesting feature in our case was that the tumor originated from the pylorus, making it an atypical neuroendocrine tumor.

**Conflicts of interest:** No conflict to declare.

**Authors' contribution**: A.V.G wrote the manuscript. T.V. and M.R. performed the ultrasound endoscopy and revised the article. O.G. and A.S. performed the surgery.

***Introducere***. *Tumorile gastrice neuroendocrine (GI-NETs) sunt leziuni rare, de obicei descoperite incidental în timpul endoscopiei. În funcție de histopatologie, sunt descrise 4 tipuri de GI-NETs. Tipul I se caracterizează prin multiple mici leziuni polipoide cu ulcerații centrale localizate în corpul gastric sau în fornix, asociate cu gastrita atrofică, de obicei noninvazive şi foarte rar metastatice. Vom prezenta un caz rar de tumoră gastrică neuroendocrină cu origine din musculara proprie a inelului piloric.*

***Prezentarea de caz***. *O pacientă în vârstă de 65 de ani cunoscută cu ciroză etanolică, se prezintă pentru investigarea unui episod de melenă. Endoscopia digestivă superioară identifică o leziune submucoasă polipoidă, pediculată cu diametrul de 30 mm, localizată la nivelul pilorului, protruzionând în duoden, cu mucoasa de acoperire normală, cu atrofie la nivelul fornixului şi multipli mici polipi la acest nivel, fără a se decela vreo sursă activă de sângerare. Tomografia computerizată nu a evidențiat metastaze la distanță. S-a efectuat ecoendoscopie şi s-a descris o formațiune solitară, rotundă, heterogenă, hipoecogenă, cu origine de la nivelul stratului muscular al pilorului. Luând în considerare diametrul de 30 mm al tumorii şi originea din stratul muscular al tumorii, în absența unei invazii locale sau a metastazelor la distanță, am decis împotriva unei rezecții endoscopice*

*şi am îndrumat pacientul către chirurgie. A fost efectuată o rezecție laparoscopică. Examenul histopatologic a descris o tumoră neuroendocrină bine diferențiată de 30/25 mm invazivă în muscularis mucosa (pT3).*

***Concluzii**. De obicei, tipurile I de tumori neuroendocrine sunt localizate la nivelul corpului şi fornixului gastric fără invazie submucoasă. Caracteristica interesantă a cazului prezentat a fost faptul că tumora origina de la nivelul pilorului, fiind o prezentare atipică pentru o tumoră neuroendocrină.*

**Correspondence to**: Theodor Alexandru Voiosu, MD, PhD
Department of Gastroenterology, "Colentina" Clinical Hospital, Bucharest, Romania
E-mail: theodor.voiosu@gmail.com

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