

## Double somatostatinoma and double papillectomy in a patient with type 1 neurofibromatosis (von Recklinghausen's disease)

Neurofibromatosis type 1 (NF1) is a rare disorder that is associated with multiple benign and malignant neoplasms [1]. The most common gastrointestinal tumors occurring in patients with NF1 are neurofibromas, gastrointestinal stromal tumors of the small bowel, and ampullary carcinoid tumors or periampullary somatostatinomas. There is controversy about their endoscopic management.

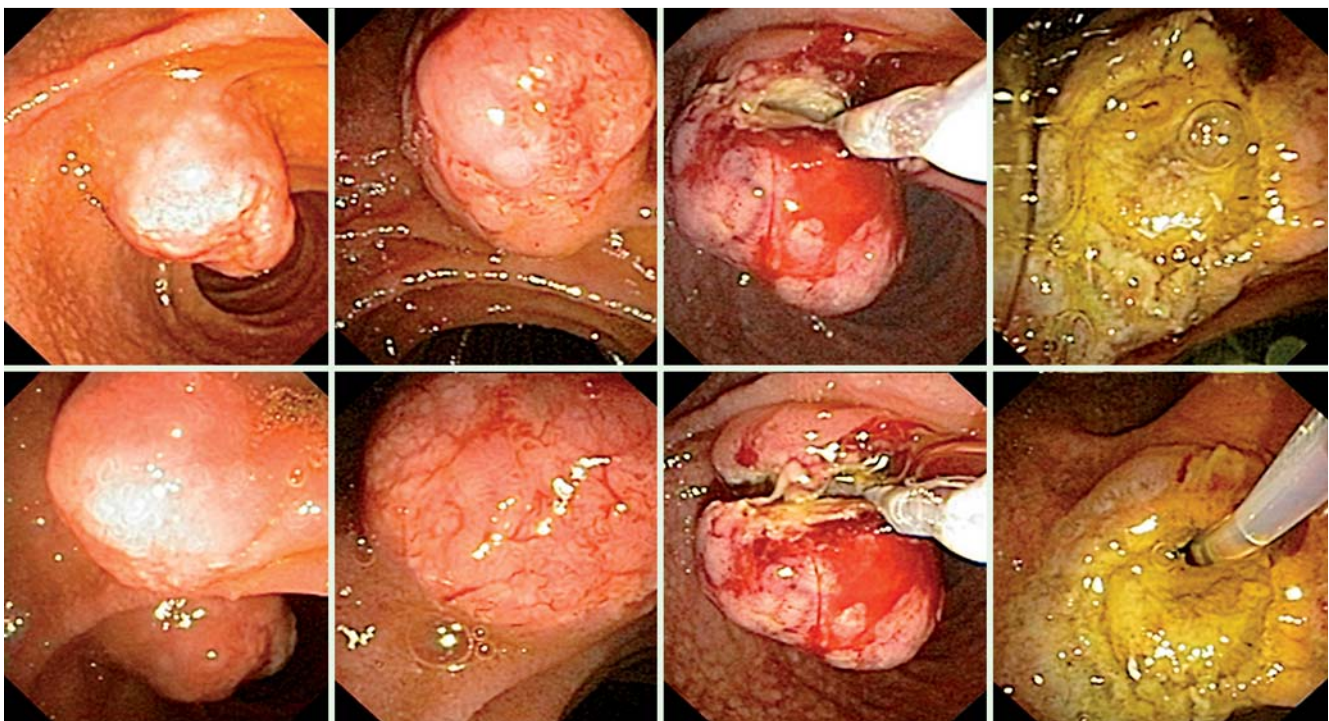
A 26-year-old man was referred for evaluation by endoscopic retrograde cholangiopancreatography (ERCP) of a periampullary mass suggestive of ampulloma. The patient had suffered from loose stools during the preceding 2 years. He had a history of NF1 and morbid obesity (BMI 46). Laboratory analyses revealed the following: serum alanine aminotransferase: 216 IU/L; serum aspartate aminotransferase: 497 IU/L; normal levels of total bilirubin; serum alkaline phosphatase: 429 IU/L; serum  $\gamma$ -glutamyl transpeptidase: 1700 IU/L. Abdominal ultrasound showed dilatation of the intrahepatic and extrahepatic biliary tracts.

Computed axial tomography and magnetic resonance imaging of the abdomen showed three tissue masses with very clear borders located at the major papilla, the minor papilla, and the duodenal wall close to the angle of Treitz. Endoscopic ultrasonography revealed no adenopathies and no muscular layer invasion.

ERCP confirmed the presence of both ampullary tumors. Cannulation of the main duct was not possible so we performed endoscopic papillectomy to remove the major papillary tumor with standard technique without a pancreatic stent (• Fig. 1; • Video 1). Histological study of the specimen revealed a firm neuroendocrine carcinoma measuring  $2.5 \times 2 \times 1$  cm, well differentiated, with neoplastic cells infiltrating the duodenal smooth muscle wall around the ampulla, extending into the overlying duodenal mucosa, but without evidence of vascular and lymphatic invasion. A few psammoma bodies were observed, suggestive of somatostatinoma (• Fig. 2). The patient underwent a second ERCP for minor papillectomy to remove the second tumor (• Fig. 3; • Video 1). Histological

study of the specimen revealed a tumor measuring  $1.3 \times 1.3 \times 1$  cm with the same histology as the tumor in the major papilla. The patient improved clinically. An Octreoscan was performed to evaluate the tumor extent and showed no evidence of pathological cavitation. Two months after the double papillectomy, upper endoscopy was performed and showed no signs of tumor recurrence. The patient then underwent surgical resection of the duodenal tumor. The postoperative course was uncomplicated, and the histological study showed this tumor to be a gastrointestinal stromal neoplasia. Twelve months later, the patient remains asymptomatic with normal liver test results. Tomographic and endoscopic images have revealed no signs of recurrence of tumor or any signs of complications related to surgery.

Patients with NF1 who present with gastrointestinal symptoms should be evaluated for the typical causes, with increased alertness for periampullary and duodenal tumors. Somatostatinoma is a rare functional endocrine tumor. The majority of these tumors arise in the pancreas (68%). Other primary sites include the duodenum, ampulla of Vater, and small intestine. The somatostatin syndrome is most often associated with pancreatic somatostatinomas. Extrapancreatic somatostatinomas tend to display different clinical features related to the local mass effect rather than the systemic effects of somatostatin.



**Fig. 1** Endoscopy revealed a tumor at the major ampulla. Papillectomy was performed with a standard polypectomy snare without a pancreatic stent.

Recently, endoscopic resection of neuroendocrine tumors of the duodenum has increasingly been performed as an alternative to conventional surgery. Most reports state that duodenal neuroendocrine tumors can be treated by endoscopic excision when the diameter is less than 10 mm and there is no invasion of the muscularis propria [2]. Others have shown that the feature associated with metastatic risk is a diameter greater than 2 cm [3], and that patients with a duodenal neuroendocrine tumor more than 10 mm in diameter can be successfully treated endoscopically [4, 5]. In our patient, because of the high sur-

gical risk, it was decided to resect the ampullary tumors endoscopically. To our knowledge, this is the first report of a patient with NF1 diagnosed with double papillary tumors, both of them somatostatin tumors, and treated with double endoscopic papillectomy.

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**Competing interests:** None

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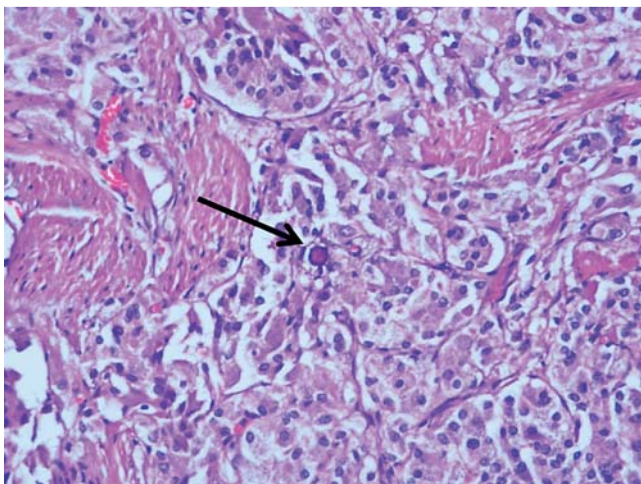
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## References

- 1 Giuly JA, Picand R, Giuly D et al. Von Recklinghausen disease and gastrointestinal stromal tumors. *Am J Surg* 2003; 185: 86–87
- 2 Dalenback J, Havel G. Local endoscopic removal of duodenal carcinoid tumors. *Endoscopy* 2004; 36: 651–655
- 3 Burke AP, Sobin LH, Federspiel BH et al. Carcinoid tumors of the duodenum. A clinicopathologic study of 99 cases. *Arch Pathol Lab Med* 1990; 114: 700–704
- 4 Alberti-Flor JJ, Hernandez ME, Ferrer JP. Endoscopic polypectomy of a large duodenal carcinoid. *Gastrointest Endosc* 1993; 39: 853–854
- 5 Boix J, Lorenzo-Zúñiga V, Moreno de Vega V et al. Endoscopic resection of ampullary tumors: 12-year review of 21 cases. *Surg Endosc* 2009; 23: 45–49

## Video 1

Major and minor papillectomy of the two papillary somatostatinomas.



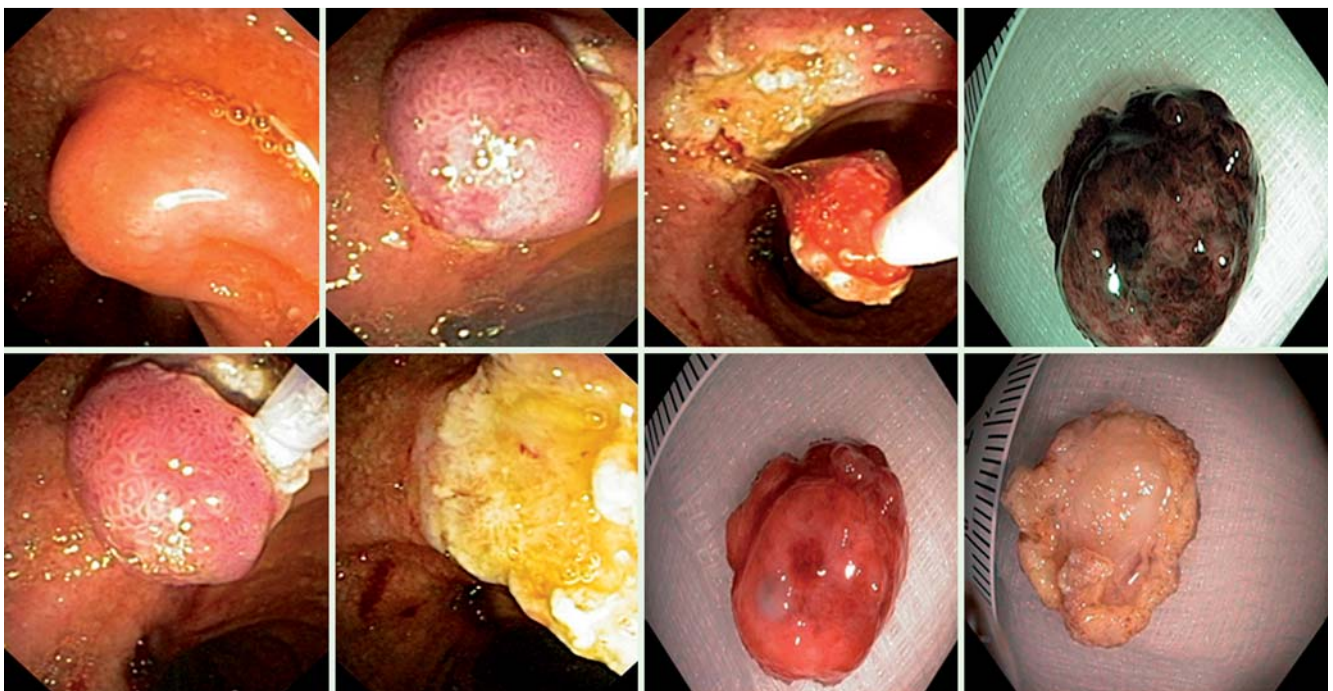
**Fig. 2** Representative photomicrograph of the tumor histology, showing a well-differentiated neuroendocrine carcinoma. A few psammoma bodies were observed, suggestive of somatostatinoma (arrow).

## Bibliography

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**Fig. 3** Minor duodenal papillectomy for tumor.