

Somatostatinoma of the minor duodenal papilla associated with pancreas divisum treated by endoscopic papillectomy

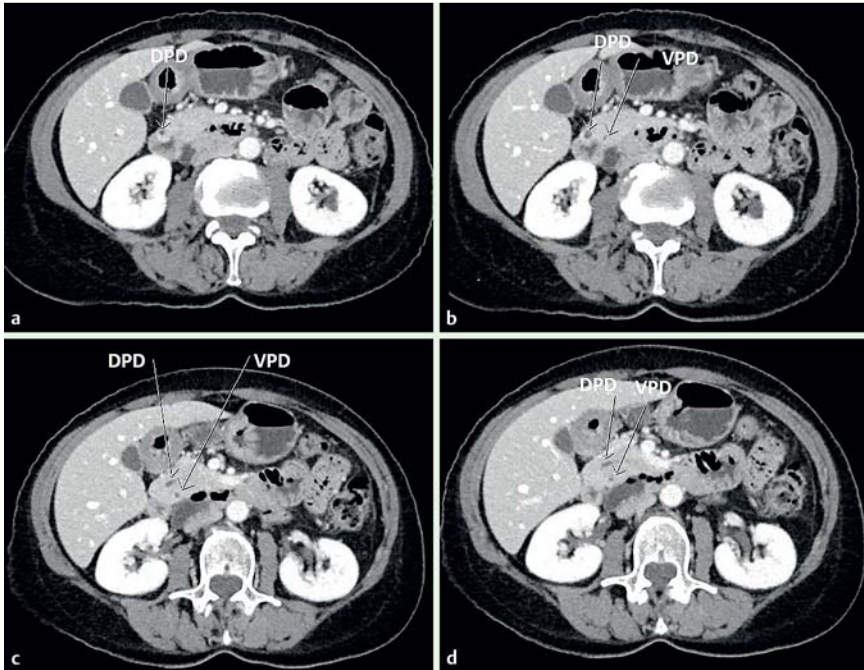


Fig. 1 Computed tomography (CT) scan showing: **a** the dorsal pancreatic duct (DPD) emerging from the minor duodenal papilla; **b** below of the DPD, the ventral pancreatic duct (VPD) emerging from the major duodenal papilla; **c** discontinuity of the DPD and the VPD; **d** the isthmus of the pancreas with the DPD predominant, while the VPD is linked to the uncinate process.

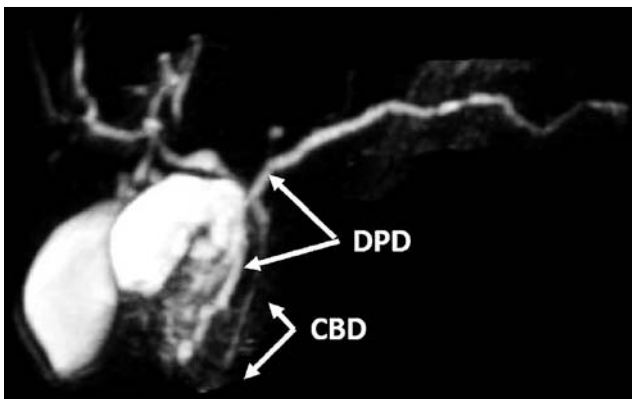
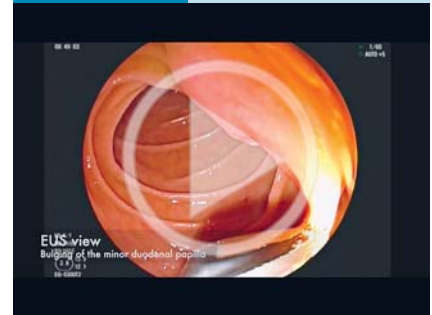


Fig. 2 Magnetic resonance cholangiopancreatography (MRCP) image showing pancreas divisum, with the dorsal pancreatic duct (DPD) emerging through the minor duodenal papilla (MiDP) and the common bile duct (CBD) discharging via the major duodenal papilla (MaDP).

Somatostatinomas are found in the major duodenal papilla (MaDP), but their diagnosis is difficult because of their absence of symptoms and small size, and because the tumor emerges from the submucosa [1]. Somatostatinomas in the minor duodenal papilla (MiDP) are extremely rare [2]. Most MiDP tumors are asymptomatic, while those of the MaDP more often cause jaundice and pain [3].

Pancreas divisum is a common congenital anatomic variant of the pancreas and occurs when the ventral pancreatic duct (VPD) and the dorsal pancreatic duct (DPD) do not fuse during embryogenesis [4]. This disunity causes the exocrine pancreatic secretions to drain via the MiDP, increasing the pressure within the DPD. Pancreatoduodenectomy is the option of choice for curative treatment of neuroendocrine tumors (NETs), but the morbidity

Video 1



Imaging showing pancreas divisum (PD) with predominance of the dorsal pancreatic duct (DPD) over the ventral pancreatic duct (VPD) and a somatostatinoma of the minor duodenal papilla (MiDP) that was resected en bloc by endoscopic papillectomy (EP).

NET, neuroendocrine tumor; WHO, World Health Organization; MRCP, magnetic resonance cholangiopancreatography; CT, computed tomography; CBD, common bile duct; EUS, endoscopic ultrasound; FICE, Fuji Intelligent Chromoendoscopy.

and mortality rates are 50% and 2%, respectively [5]. As a result treatment by endoscopic papillectomy has become more attractive owing to its lower morbidity and mortality rates. We present a rare case of somatostatinoma in the MiDP, associated with pancreas divisum, which was treated by endoscopic papillectomy without the insertion of a pancreatic stent even though there was pancreas divisum.

A 60-year-old woman presented with epigastric pain, and esophagogastroduodenoscopy (EGD) showed bulging of the MiDP. A biopsy was taken; histology of this specimen revealed a NET (grade I according to the World Health Organization [WHO] classification). Her amylase level was 92 IU/dL; the results of chromogranin A and all other standard tests were normal. Computed tomography (CT) scanning showed the prominent MiDP and that the DPD was predominant, but that there was no evidence of metastases (▶ **Fig. 1**) and this was confirmed by magnetic resonance imaging (▶ **Fig. 2**). Endoscopic ultrasound (EUS), performed in planning for the endoscopic papillectomy,

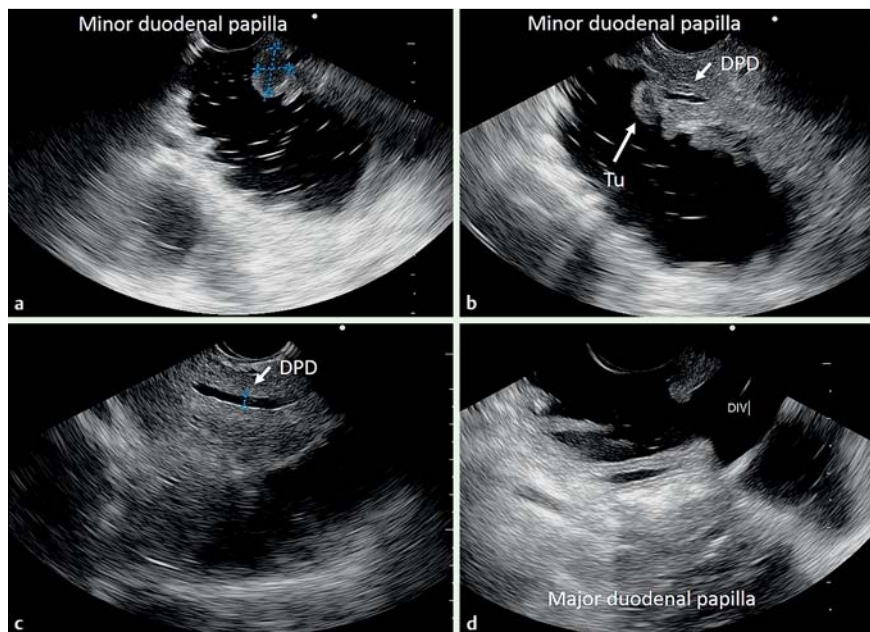


Fig. 3 Endoscopic ultrasound (EUS) images showing: **a** a hypoechoic homogeneous nodule with clear limits (1.3×0.8 cm) without invasion of the dorsal pancreatic duct (DPD), which had a diameter of 35 mm; **b** the dominance of the DPD in the head of the pancreas; **c** the dominance of the DPD in the body of the pancreas; **d** the peridiverticular major duodenal papilla (DIV).



Fig. 5 Endoscopic view showing bleeding from the ulcer, which was treated by hemoclips and adrenaline injection.

Histology showed a grade I somatostatinoma (WHO classification) with angio-lymphatic infiltration and the following immunohistochemistry results: somatostatin (+), neuron-specific enolase (+), synaptophysin (+), chromogranin (+), and a Ki-67/mitotic proliferation index of <2%.

Endoscopy_UCTN_Code_TTT_1AO_2AZ

Competing interests: None

Daniela I. Jara Letelier¹, Michele Lemos Bonotto^{2,3}, José Celso Ardengh⁴

¹ Gastroenterology Section and Endoscopy Unit, Hospital del Salvador and Faculty of Medicine, Universidad de Chile, Santiago, Chile

² Rio Grande do Sul Foundation of Gastroenterology (FUGAST), Porto Alegre, Brazil

³ Department of Gastroenterology and Hepatology, Santa Casa Hospital/Federal University of Health Sciences of Porto Alegre (UFCSPA), Porto Alegre, Brazil

⁴ Departments of Surgery and Anatomy, Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo, Ribeirão Preto, São Paulo, Brazil

References

- 1 Makhlof HR, Burke AP, Sobin LH. Carcinoid tumors of the ampulla of Vater: a comparison with duodenal carcinoid tumors. *Cancer* 1999; 85: 1241–1249
- 2 Waisberg J, de Matos LL, Waisberg DR et al. Carcinoid of the minor duodenal papilla associated with pancreas divisum: Case report and review of the literature. *Clinics* 2006; 61: 365–368
- 3 Noda Y, Watanabe H, Iwafuchi M et al. Carcinoids and endocrine cell micronests of the minor and major duodenal papillae. Their incidence and characteristics. *Cancer* 1992; 70: 1825–1833
- 4 Seyama Y, Kubota K, Sano K et al. Intraoperative direct pancreatography using ultrasound-guided puncture for accessory duct

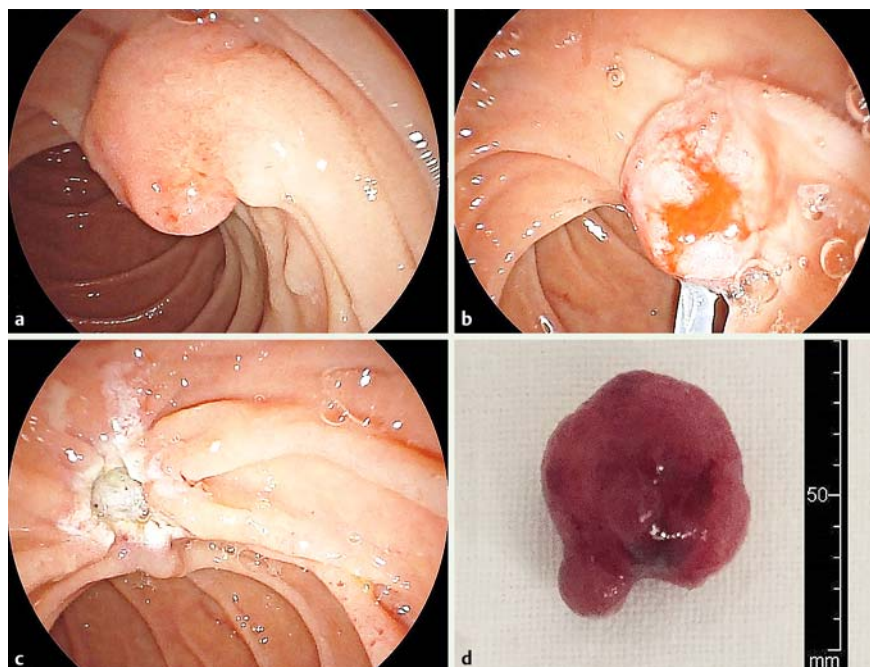


Fig. 4 Endoscopic views showing: **a** the minor duodenal papilla (MiDP) with a tumor causing a slightly reddened and rough surface; **b** craniocaudal capture with a polypectomy snare; **c** the ulcer left after en bloc tumor resection. **d** Photograph of the surgical specimen obtained by en bloc resection.

showed a rounded hypoechoic nodule of more than 2.0 cm in size, which was restricted to the MiDP with no invasion of the DPD, which was 35 mm in diameter, and without communication with the VPD (Fig. 3).

Endoscopic papillectomy removed the tumor en bloc and insertion of a pancreat-

ic stent was not required because of the dilatation of the DPD (Fig. 4). Hematochezia occurred 28 hours after the resection and was treated with clips and injection of 1:10 adrenaline and glucose (25%) (Fig. 5). The patient progressed well and was discharged 6 days after the endoscopic papillectomy.

sphincteroplasty in a patient with pancreas divisum. *Pancreas* 2003; 27: 98–100

- 5 *Ardengh JC, Kemp R, Lima-Filho ER et al.* Endoscopic papillectomy: The limits of the indication, technique and results. *World J Gastrointest Endosc* 2015; 7: 987–994

Bibliography

DOI <http://dx.doi.org/10.1055/s-0042-104928>
Endoscopy 2016; 48: E135–E137
© Georg Thieme Verlag KG
Stuttgart · New York
ISSN 0013-726X

Corresponding author

Daniela I. Jara Letelier, MD
Managua 2222-77
Ñuñoa
Santiago 7770443
Chile
[danijaralet@gmail.com](mailto:danjaralet@gmail.com)