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Long-term follow-up of duodenal papillary somatostatinoma treated by endoscopic papillectomy

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Somatostatinomas are rare neuroendocrine tumors usually associated with Von Recklinghausen's disease, multiple endocrine neoplasia, or Von Hippel-Lindau syndrome. Nevertheless, it may be associated with sporadic occurrence in healthy patients[1]. Approximately 55% of somatostatinomas are pancreatic tumors and the remainder arises in ampullary and periampullary region or, rarely, in jejunum[2]. A patient with pancreatic somatostatinoma usually presents a triad of steatorrhea, weight loss and diabetes mellitus, due to inhibition of gastrointestinal peptides[1,3]. In duodenal papillary somatostatinomas (DPS), these symptoms occur in less than 10%. Endoscopic retrograde cholangio-pancreatography (ERCP), magnetic resonance or computed tomography can aid in diagnosis, which is only confirmed by psammoma bodies on histologic studies, a rare finding in pancreatic tumors[4]. Surgical resection (duodenopancreatectomy) is the treatment of choice. The authors present a case of DPS with cholestasis, endoscopic treated with excellent results and follow-up for 7 years.

44-year-old woman with biliary colic for 1 month and evolving with 1-week jaundice. Bilirubin =14 mg/dL (conjugated =12 mg/dL), AST, ALT and GGT increased and chromogranin A =6 U/l. ERCP revealed choledochal dilatation, presence of spontaneous choledocoduodenal fistula and a papillary tumor. A 10F plastic prosthesis was inserted through the fistulous orifice and biopsy was performed. The pathology report revealed neuroendocrine tumor. Magnetic Resonance Imaging unveiled a tumor on duodenal papilla (2.0 cm) and three peri duodenal lymph nodes (LN). PET/CT (FDG) revealed hypermetabolic area on duodenal papilla without LN involvement (FIGURE 1). Endosonography revealed a lesion confined to duodenal papilla without invasion of adjacent structures (uT1) and 3 LN (FIGURE 2). EUS-FNA of the LN and endoscopic papillectomy were performed for whitish gallstones extraction (FIGURE 3, E-VIDEO* and FIGURE 4). The LN pathology was negative for metastasis and papillectomy specimen revealed neuroendocrine tumor of 2.1x1.7x1.3 cm, free

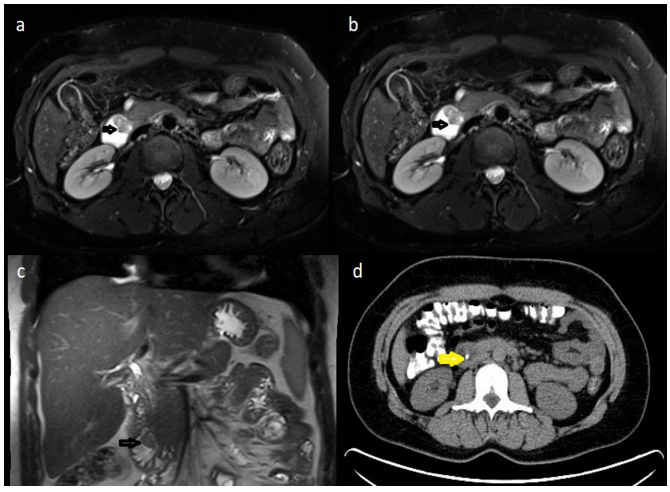

FIGURE 1. Abdominal magnetic resonance imaging shows a mass (black arrows) inside the duodenal lumen on topography of duodenal papilla (a, b and c). PET/CT (FDG) shows hypermetabolic area (yellow arrow) on topography of duodenal papilla (descending part of duodenum) (d).

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***E-VIDEO: https://youtu.be/7_Y3LyhPBpQ**

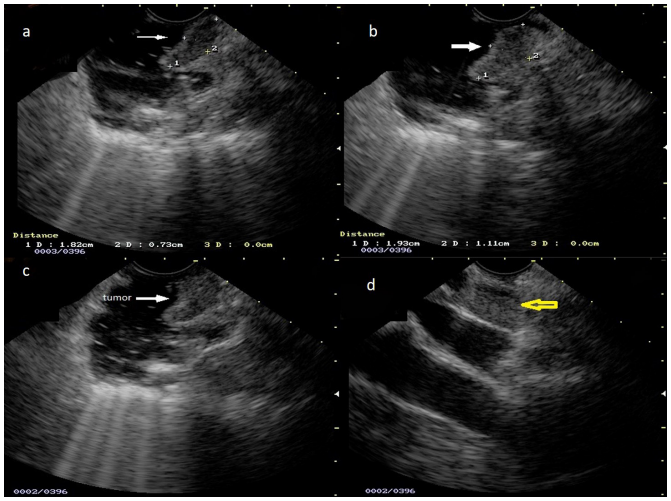

FIGURE 2. Endoscopic ultrasound. Hypoechoic and homogeneous area with precise limits (white arrows) in duodenal papilla measuring 1.82x0.73 (major axes) (a, b and c). In the image d the yellow arrow points to a perilesional lymph node that was biopsied with a 22G needle, just before the endoscopic papillectomy.

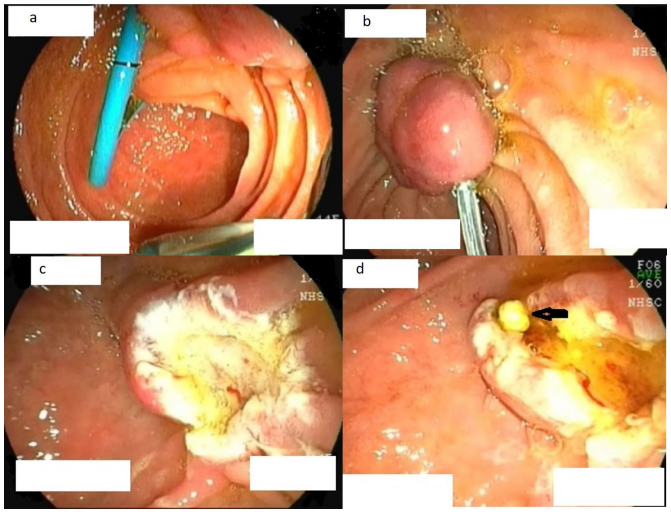

FIGURE 3. Endoscopic images. (a) plastic prosthesis inserted by spontaneous choledocoduodenal fistula. (b) Apprehension moment of the papilla tumor with polypectomy loop. (c) Image immediately after remove the entire tumor in monoblock. (d) Multiple stones extraction after enlarging the choledocean orifice with the papillotome.

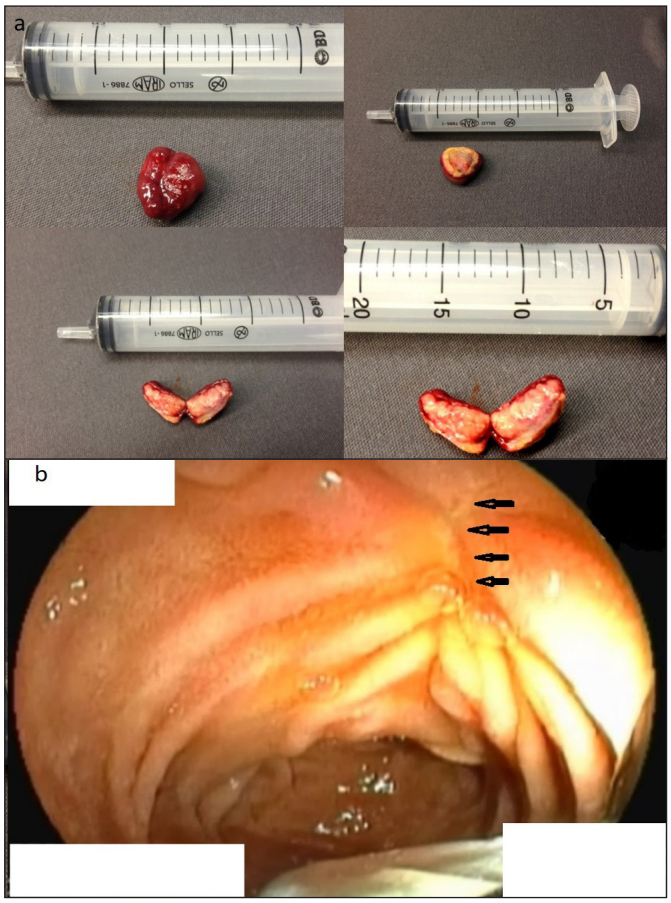

FIGURE 4. Final images of endoscopic papillectomy (a). (b) Endoscopic control image (1 year) of the endoscopic papillectomy site. It is possible to identify a well-defined scar (black arrows).

margins with vascular invasion. Immunohistochemistry showed a G1 neuroendocrine neoplasia with CD56 (+), synaptophysin (+), chromogranin A (+), Ki-67 of 0.75 and anti-somatostatin antibodies (+). After 7 years of follow-up the patient is in good condition without recurrence.

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