

A case report: endoscopic enucleation of gastrointestinal stromal tumor of the ampulla of Vater

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Abstract

Objective To report a case of endoscopic enucleation of a gastrointestinal stromal tumor of the ampulla of Vater.

Design The tumor resection was performed by needle knife endoscopic submucosal dissection.

Results The tumor was resected completely without serious complications and the patient recovered rapidly.

Conclusion Endoscopic enucleation may be an alternative treatment for gastrointestinal stromal tumors of the ampulla of Vater for patients who fulfill certain requirements.

Keywords GIST · ESD · Ampulla of vater

Introduction

Gastrointestinal stromal tumors (GISTs) of the duodenum ampulla are extremely rare, and the main treatment is surgical resection. We think this is the first report of endoscopic enucleation of a GIST.

Case history

A 56-year-old man with a 2-month history of bloating, but without jaundice liver dysfunction, anemia or gastrointestinal bleeding, underwent a duodenoscopy which revealed a tumor in the upper right of the duodenal papilla. The patient had undergone endoscopic retrograde

cholangiopancreatography (ERCP) preoperatively which showed the bile duct and pancreatic duct to be normal. Endoscopic ultrasound (EUS) showed a hypoechoic, homogeneous mass (15 × 20 mm) in the ampulla of Vater which originated in the fourth layer with a regular border (Fig. 1). Abdominal computed tomography (CT) scans showed a 2-cm hypervascular lesion with a regular border lying between the head of the pancreas and the second part of the duodenum. Ultrasonography, CT, ERCP and magnetic resonance imaging indicated no dilatation of the intrahepatic and extrahepatic bile duct, and no metastasis and enlargement of the lymph nodes.

With the results from the EUS, duodenoscopy and CT we suspected it was probably a GIST, either leiomyoma or schwannoma. Considering EUS fine needle aspiration (FNA) may cause bleeding, tumor metastasis, damage the pancreatic duct, we have not performed it before we decided to perform endoscopic submucosal dissection (ESD). Tumor resection was performed with a needle knife (KD-10Q-1; Olympus, Japan) ESD following a submucosal injection of 20 ml saline containing epinephrine at a concentration of 1:10,000. During the procedure it is necessary to avoid damage to the bile duct and pancreatic duct, and to find the space between the tumor capsule and surrounding normal tissues, and completely remove the tumor along the capsule (Fig. 2). No massive bleeding and perforation occurred during or after the operation.

The tumor which measured 1.6 cm × 1.2 cm × 0.8 cm was resected completely and sent for pathological analysis. The mitotic index was <3 per 50 high-power fields (Fig. 3). By immunohistochemical staining, the spindle cells were positive for DOG1 but negative for CD117, CD34, S-100, SMA. Ki-67 antigen was expressed in two percent of the tumor cells. The resected margin was negative, and there was no lymphatic invasion or venous invasion. A diagnosis

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Fig. 1 GIST of the ampulla of Vater before ESD. **a, b** GIST was found by a forward-viewing endoscope and a side-viewing duodenoscopy. **c** Bile duct cannulation. **d** EUS evaluation of the tumor

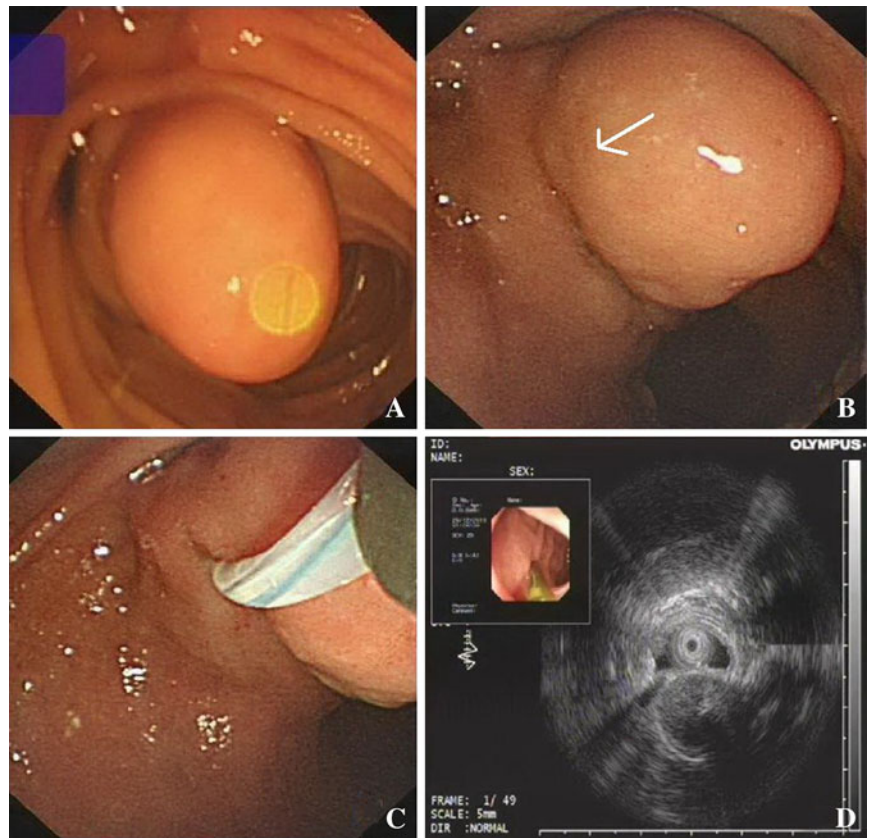


Fig. 2 Endoscopic enucleation of the tumor. **a, b** Detached from the mass, the overlying mucosa was incised by a needle-knife. **c** Wounds left by endoscopic enucleation. **d** The mass was resected completely

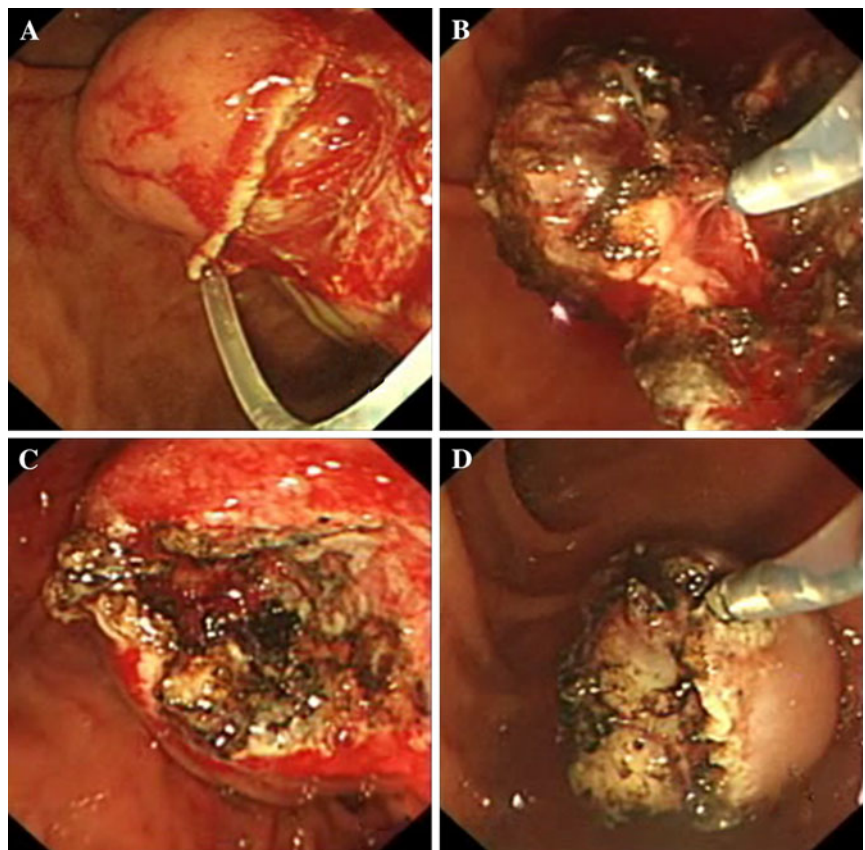
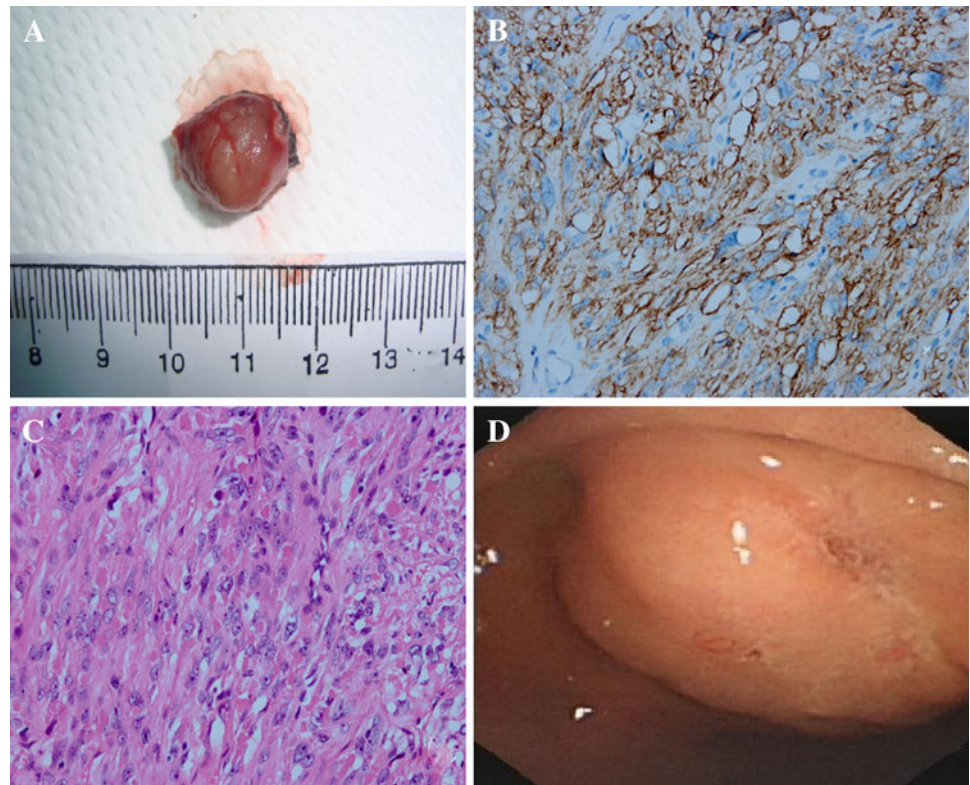


Fig. 3 **a** Pathology revealed a spindle-cell neoplasm. **b** Immunohistochemistry showed spindle cells to be DOG-1 positive. **c** Enucleated tumors. **d** Scarring of the mucosa. Duodenoscopy follow-up of the ampulla of Vater 3 months later



of GIST with low potential risk for malignancy was made. The patient recovered rapidly and was discharged 4 days after resection. Three months later, the duodenoscopy showed a scar of the mucosa without any signs of recurrence (Fig. 3).

Discussion

GISTs are the most common mesenchymal tumors in the gastrointestinal tract; 50–70 % of them occur in the stomach, 20–30 % in the small intestine, 10 % in colon-rectum, and only 3–5 % in the duodenum [1]. Most of them express CD117 and CD 34, but DOG1 antibodies also show high sensitivity and specificity for GIST [7]. GIST of the duodenum ampulla is extremely rare [2] with GI bleeding, obstructive jaundice and abdomen pain as the most common clinical presentation.

The main treatment for primary GISTs is surgical resection, when possible. However, GISTs rarely metastasize to lymph nodes [3], and wide margins may not be necessary with extensive lymphadenectomy. Some authors reported local excision of the tumors by surgery in recent years [4, 5], thus avoiding a Whipple's procedure. Because of trauma and long recovery, it is difficult to select surgery for patients with GISTs. Recently, some articles have

reported on endoscopic resection for small GISTs [6]; however, there are no reports about ESD of GISTs of the ampulla of Vater. In our case, we ensured the patient's safety as a precondition, and the tumor was removed successfully by ESD instead of surgery, thereby avoiding some of the complications. Not all duodenal GISTs are suitable for endoscopic enucleation. In such cases, we emphasize the importance of undergoing EUS and abdominal CT before the procedure to make sure the cancer has not metastasized and the tumors are located in the duodenum wall.

Endoscopic removal of duodenum GISTs, especially of the ampulla of Vater, is not an established clinical practice because of the high risk of severe complications, such as perforation and massive bleeding and, as such, it should be performed by a skilled endoscopist with ERCP and ESD performed weekly to ensure lower complication rates. Another important issue is to ensure complete removal of the tumor to avoid implantation metastasis, although there is insufficient literature about this.

In conclusion, endoscopic enucleation may be an alternative for the treatment of GISTs of the ampulla of Vater for patients who fulfill certain requirements including no metastasis, no infiltration to the serosa and no ulcer in the tumor. The success rate and safety of the technique depends upon the experience of the endoscopist.

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Frampton AE, Bong JJ, Kyriakides C, et al. En bloc resection of the pancreatic head and second part of duodenum for a duodenal gastrointestinal stromal tumor: a multi-media report. *J Pancreas*. 2010;11(4):396–400.
2. Kim SH, Kim JH, Baik GH, Baek I, Hahn T, Oh SO, et al. Malignant gastrointestinal stromal tumor of the ampulla of Vater: a case report. *Korean J Gastroenterol*. 2004;43:66–70.
3. Laurini JA, Carter JE. Gastrointestinal stromal tumors: a review of the literature. *Arch Pathol Lab Med*. 2010;134:134–41.
4. Liyanage CA, Abeygunawardhana S, Kumarage S et al. Duodenum-preserving local excision of a gastrointestinal stromal tumor. *Hepatobiliary Pancreat Dis Int*. 2008; 7: 214–6.
5. He QS, Jiang JB, Liu FJ, et al. Partial duodenectomy and translocation of the distal common bile duct in repairing duodenal defect near the papilla of Vater for a gastrointestinal stromal tumor. *Chin Med J*. 2007;120(16):1462–4.
6. Bai J, Wang Y, Guo H, et al. Endoscopic resection of small gastrointestinal stromal tumors. *Dig Dis Sci*. 2010;55(7):1950–4.
7. Espinosa I, Lee CH, Kim MK, et al. A novel monoclonal antibody against DOG1 is a sensitive and specific marker for gastrointestinal stromal tumors. *Am J Surg Pathol*. 2008;32(2):210–8.