Endoscopic resection with pancreatic duct stenting for periampullary gangliocytic paraganglioma: successful treatment of rare tumor.

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Learning objectives

Gangliocytic paraganglioma (GP) – rare correctly diagnosed tumor with unclear malignant potential, commonly located in duodenum.

GP should be differentiated with neuroendocrine tumor and gastrointestinal stromal tumor.

Given the malignant potential, tumor excision are recommended

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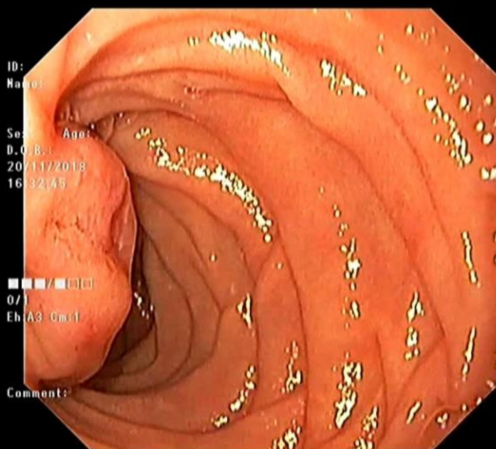
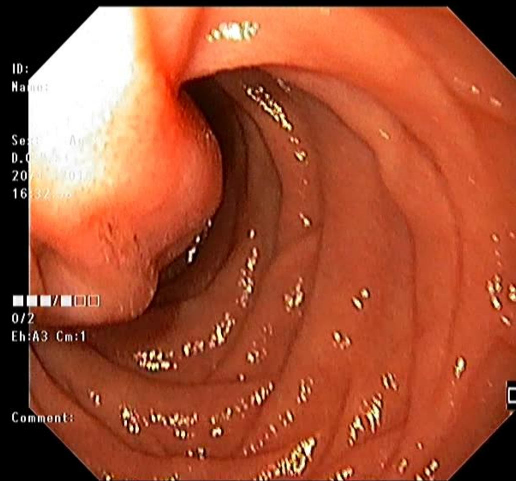
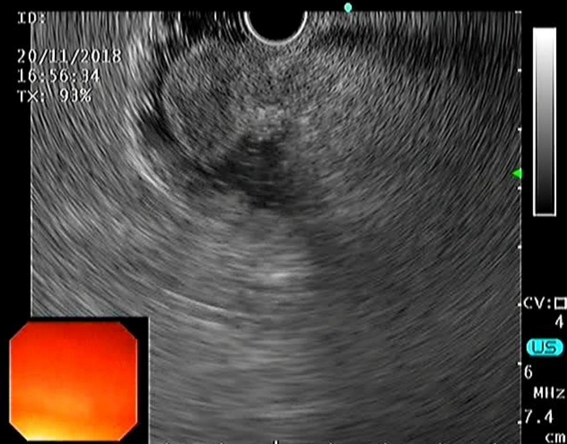
CLINICAL CASE

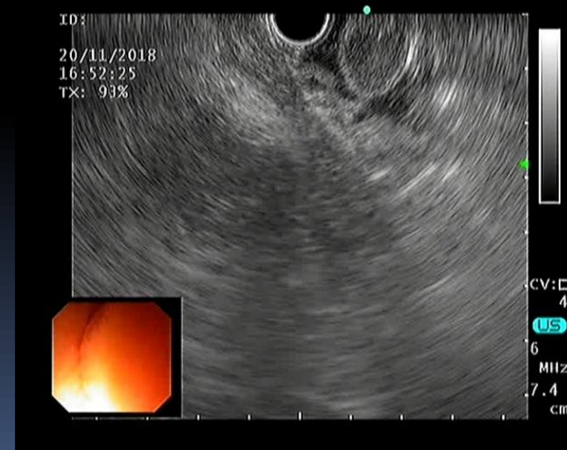
A 57 year-old woman with abdominal pain visited two hospitals and underwent upper GI endoscopy twice. Endoscopic finding in the duodenum was interpreted as a «polyp» or «adenoma of the ampulla of Vater».





Forward view endoscopy





Endosonography

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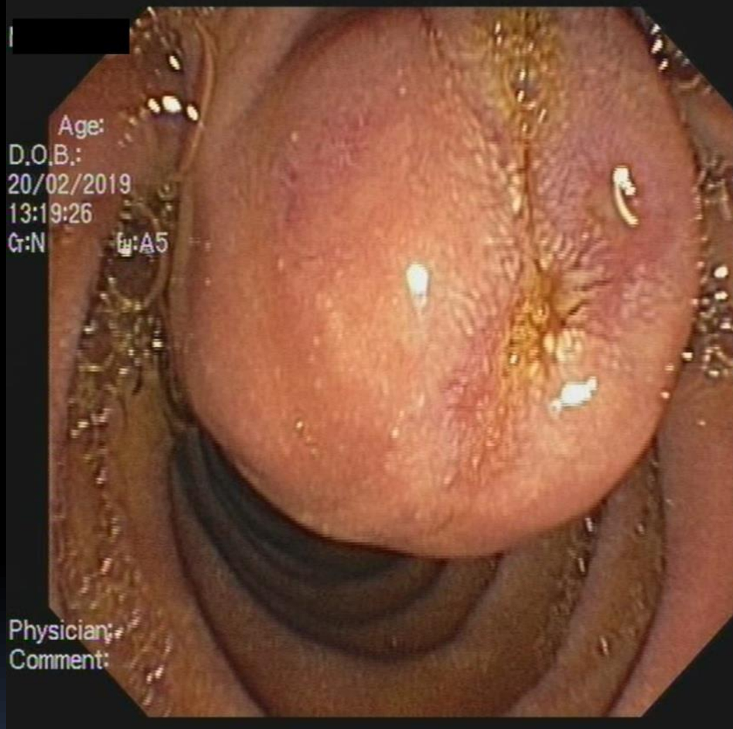
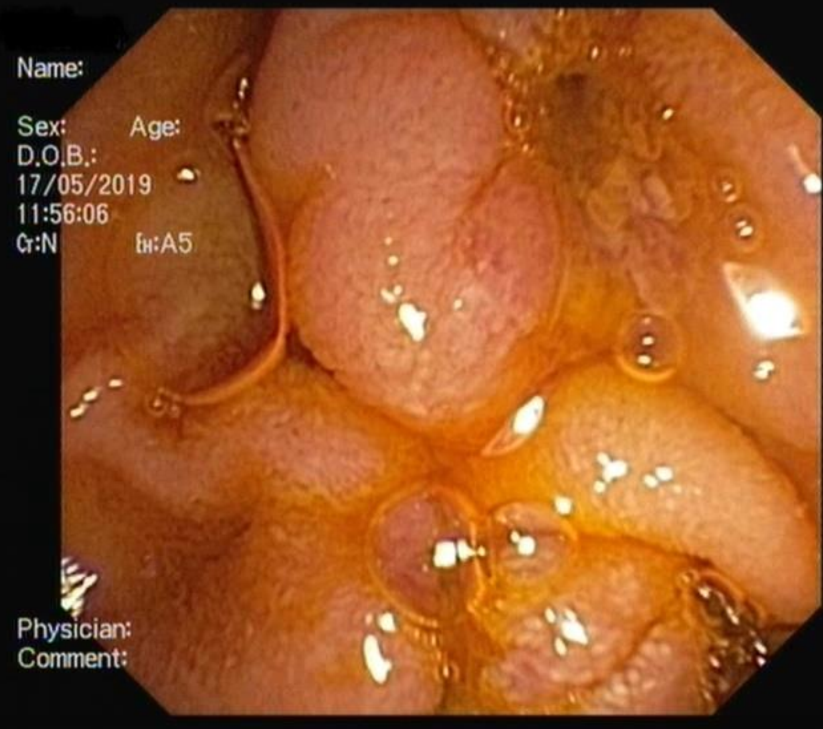
CLINICAL CASE





Side view endoscopy before operation

Side view endoscopy 3 month after operation

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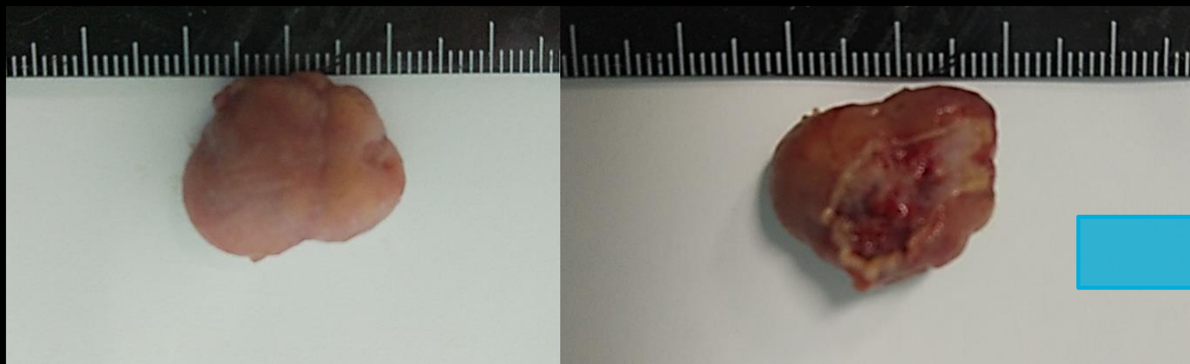
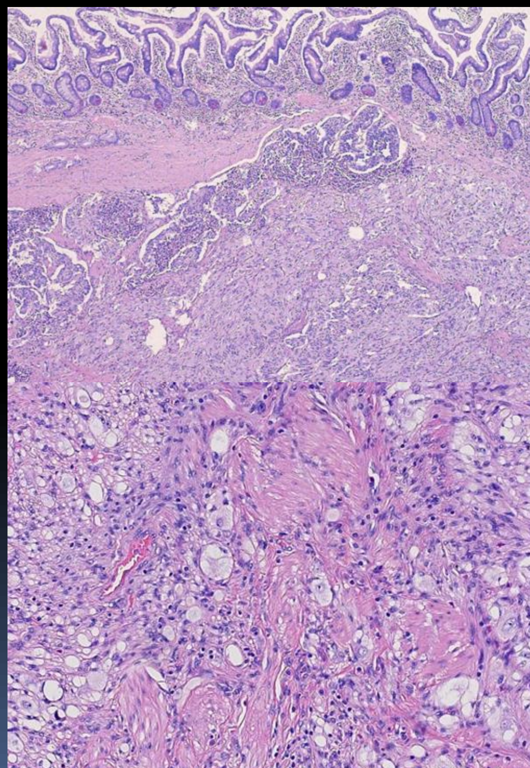
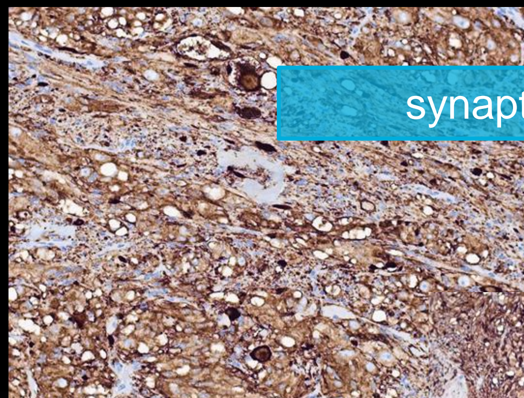
TUMOR PATHOMORPHOLOGY



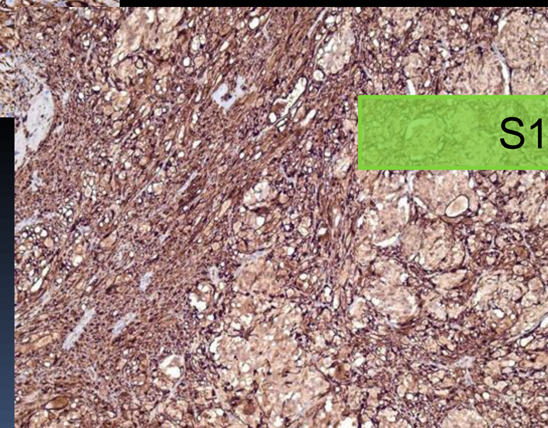
Tumor size – 2,5*1,5 cm



H&E stain



synaptophysin



S100

Immunohistochemistry

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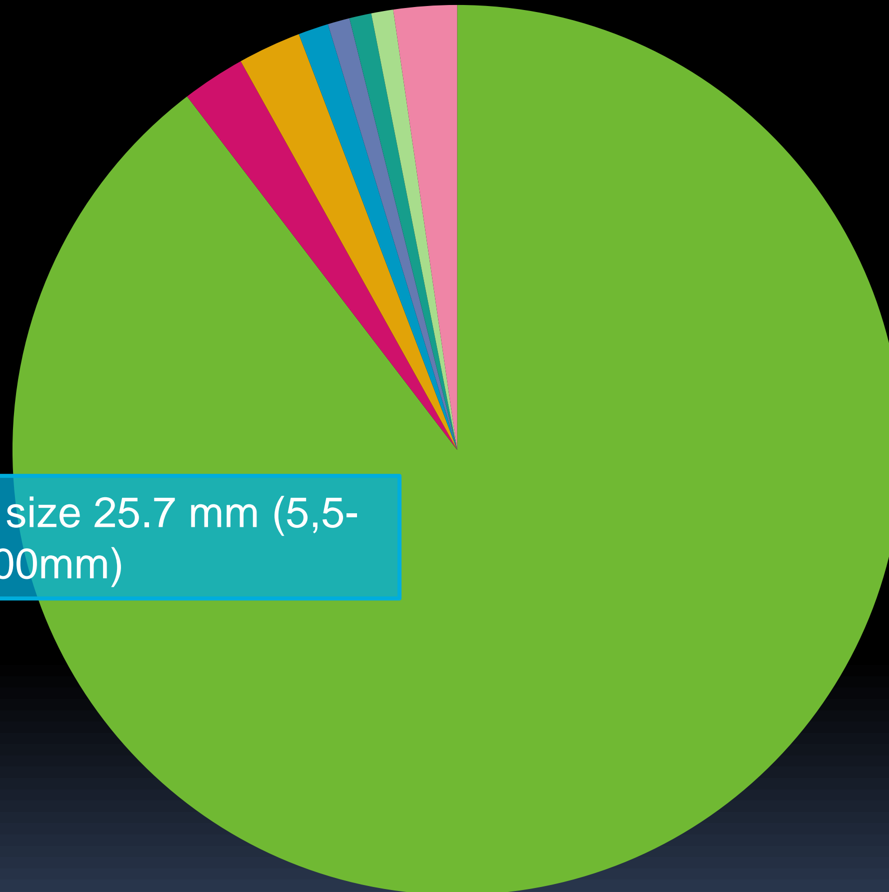
МНИОИ им. П.А.Герцена

LITERATURE DATA

(n = 263)



- duodenum 236
- bronchus 6
- meninges 6
- pancreas 3
- small intestine 2
- esophagus 2
- appendix 2
- other localizations 6

Median tumor size 25.7 mm (5,5-100mm)

Okubo Y Frontiers in oncology Vol 8 2018 doi: 10.3389/fonc.2018.00291

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DUODENAL GANGLIOCYTIC PARAGANGLIOMA WITH METASTASES

Ref.	Year of publication	Age at diagnosis (yr)	Sex	Presenting symptoms	Primary location	Largest diameter, primary (mm)	Site(s) of metastasis	LNs sampled	LNs positive	Therapy	Follow-up (mo)
Büchler et al	1985	50	M	GI bleeding	D2, ampulla	30	Peripancreatic LN	NR	1	Local resection	20, NED
Korbi et al	1987	73	F	GI bleeding, weight loss, cardiac decompensation	D2, ampulla	90	Peripancreatic LN	NR	1	PD	0, died POD 7
Inai et al	1989	17	M	GI bleeding	D2, ampulla	20	Peripancreatic LN	NR	1	Local resection, followed by PD with LND	32, NED
Hashimoto et al	1992	47	M	Asymptomatic, incidental	D2, ampulla	65	Peripancreatic LN	NR	1	PD with LND	14, NED
Dookhan et al	1993	41	M	Abdominal pain, partial duodenal obstruction	D2	25	Mesentery, mesenteric LNs	NR	2-3	Local resection (1981); resection D4, proximal jejunum, mesenteric mass (1992)	131, recurrence and metastasis
Takabayashi et al	1993	63	F	Abdominal pain	D3	32	Regional LN	NR	1	PPPD	24, NED
Tomic et al	1996	74	M	Anemia, steatorrhea, abdominal pain, weight loss	Pancreas, head	40	Peripancreatic LN	NR	1	PD	19, NED
Henry et al	2003	50	M	Jaundice	Pancreas, head	30	Manubrium	NR	0	FNA[1], followed by PD, followed by manubrium resection	21, NED
Sundararajan et al	2003	67	F	Asymptomatic, incidental	D2	50	Regional LNs	NR	2	PD with LND	9, NED
Wong et al	2005	49	F	GI bleeding, abdominal pain	D2, periampullary	14	Periduodenal and Peripancreatic LNs	7	6	PPPD with LND, radiotherapy	12, NED
Witkiewicz et al	2007	38	F	Abdominal pain	D2, periampullary	15	Regional LNs	7	2	Local resection, followed by PPPD	NR
Mann et al	2009	17	F	Duodenal obstruction, weight loss, abdominal pain	D2/D3 junction		Regional LNs	11	4	PD	7, NED
Okubo et al	2010	61	M	GI bleeding, abdominal pain	D2, ampulla	30	Regional LNs	NR	1	PPPD with LND	6, NED
Saito et al	2010	28	M	GI bleeding	D2, ampulla	17	Regional LNs	N/A	2	Local resection, followed by PD	N/A
Uchida et al	2010	67	F	Anemia	D2	N/A	LN	N/A	N/A	PD	N/A
Ogata et al	2011	16	M	GI bleeding	D2, ampulla	35	Peripancreatic LNs	NR	4	PPPD with LND	36, NED
Barret et al	2012	51	F	GI bleeding	D2, ampulla	25	Peripancreatic LNs	NR	2	FNA, followed by PD	96, NED
Rowsell et al	2011	52	F	Asymptomatic, incidental	D2, periampullary	10	Regional LNs, liver nodule	23	2	PD, post-op octreotide injections	27, No change in residual liver metastases
Dustin et al	2011	56	F	Abdominal pain, weight loss	D2, periampullary	18	Retroperitoneal LN, later resection Peripancreatic LNs	10	3	Local resection of retroperitoneal mass, followed by duodenal mass FNA, followed by PPPD with LND and cholecystectomy	NR
Fiscaletti et al	2011	61	M	Abdominal pain, weight loss	D2, minor papilla (discovered incidentally)	15	Peripancreatic LN	7	1	FNA[2], followed by PPPD	12, NED
Amin et al	2013	57	M	Abdominal pain, vomiting	D2, ampulla	30	Portal hepatic LNs, Liver	NR	NR	Resection of duodenal mass, retropancreatic mass, part of hepatic lesion, enlarged portal lymph nodes	8, Residual liver lesion slowly enlarging
Choi et al (poster)	2014	41	M	GI bleeding	D2	25	Periduodenal LN	11	1	PD	NR
Li et al	2014	47	M	Abdominal pain	D2, periampullary	30	Regional LNs, pelvic cavity, liver	16	7	PD, radiotherapy, chemotherapy	13, died secondary to liver and pelvic metastases
Micev et al (poster)	2014	57	M	Abdominal pain, back pain, intermittent jaundice	D2, ampulla	35	Regional LNs	NR	2	NR	NR
Shi et al	2014	47	M	Abdominal pain, weight loss	D2, ampulla	40	Regional LNs	20	8	PD with LND	24, NED
Dowden et al	2015	59	F	Abdominal pain, weight loss	D2, ampulla	28	Regional LNs	22	2	FNA, followed by PPPD	5, NED
Lei et al	2015	45	M	GI bleeding, weight loss, vomiting and diarrhea, abdominal cramps (functional tumor)	D2	15	Periduodenal LN	NR	1	FNA, followed by ampullectomy with periduodenal and retropancreatic LND	3, functional symptoms and CT showing lymphadenopathy, lost to follow-up
Sun et al (poster)	2015	40	F	Abdominal pain	D2, ampulla	20	Peripancreatic LN	NR	1[3]	FNA, followed by PD	12, NED
Wang et al	2015	49	M	Abdominal pain	D2	33	Regional LNs	9	3	PD with LND, chemotherapy	36, NED
Hu et al	2016	65	M	GI bleeding	D2	30	LN	NR	1	Local resection	2, NED
Current case	2016	62	M	Asymptomatic, incidental	D2, periampullary	20	Regional LNs	8	3	FNA, PD	30, NED

[1]Suggestive of ductal adenocarcinoma; [2]Fibroinflammatory changes consistent with pancreatitis; [3]Involved by direct extension and replacement. GI: Gastrointestinal; D2: Duodenum 2nd portion; D3: Duodenum 3rd portion; LN: Lymph node; LND: Lymph node dissection; PD: Pancreaticoduodenectomy; PPPD: Pylorus-preserving pancreaticoduodenectomy; NED: No evidence of disease; N/A: Not available; NR: Not reported; LtFU: Lost to follow-up.

N1 – 11,01%

M1 – 2,1%

- 31 cases GP with lymph node metastasis and 5 cases GP with liver metastasis are described in literature

Cathcart S J World J Clin Cases 2017 June 16; 5(6): 222-233

TREATMENT OPTIONS OF GP

Transduodenal excision (40.6%) Endoscopic resection (17.7%)

- Tumor size 2 cm without lymph node metastasis

Pancreaticoduodenectomy (30.1%)

- Large size of tumor
- suspicious lymph nodes
- noncurative local excision/resection

Barret M. European Journal of Gastroenterology & Hepatology 2012, 24:90–94

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TAKE HOME MESSAGE

- Gangliocytic paraganglioma (GP) – rare correctly diagnosed tumor with unclear malignant potential

- GP should be differentiated with neuroendocrine tumor and gastrointestinal stromal tumor.
- Given the malignant potential, we recommended tumor excision.
- Endoscopic resection (in cases of absence of muscular layer invasion and suspicious lymph nodes) is preferred operation.

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