

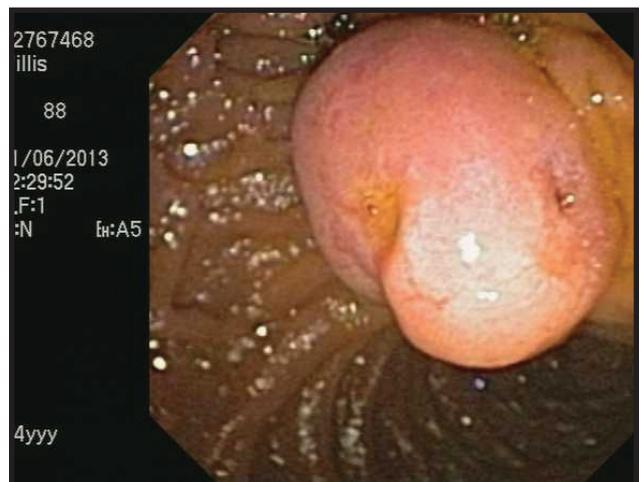
Ampullary Carcinoid as a Rare Cause of Acute Relapsing Pancreatitis

by John M. Petersen, David H. Holloman

Fewer than 150 cases of ampulla of Vater carcinoid have been reported. These lesions commonly present with abdominal pain, jaundice or gastrointestinal (GI) bleed. A significant percentage of tumors may be metastatic at the time of diagnosis. The patient in whom this carcinoid was found presented with numerous bouts of acute, relapsing pancreatitis. Endoscopic ultrasound (EUS), computed tomography (CT), endoscopic retrograde cholangiopancreatography (ERCP) and ampullectomy were key in making the diagnosis. This case emphasizes the need for clinicians and endoscopists to consider structural or neoplastic lesions as a cause of relapsing pancreatitis not related to alcohol, autoimmune disease or biliary tract pathology. Endoscopic mucosal resection and ampullectomy may be an appropriate definitive treatment.

CASE

A 50-year-old African-American woman with Von willebrand disease had numerous bouts of periumbilical pain, nausea and vomiting with documented biochemical pancreatitis over a five month period. There was no alcohol abuse, and her gallbladder was normal on ultrasound and nuclear (HIDA) scan. The triglycerides were normal and her anti-nuclear antibody (ANA) was negative. There was no pertinent family history. Physical exam was normal with a soft abdomen that was non-tender and without mass, ascites or organomegaly. Laboratory data revealed a normal complete blood count (CBC) and normal liver enzymes with the exception of a gamma-glutamyl transpeptidase (GGT) of 68 mg/dl (normal 20-45). Her amylase levels, captured during the attacks of pain,



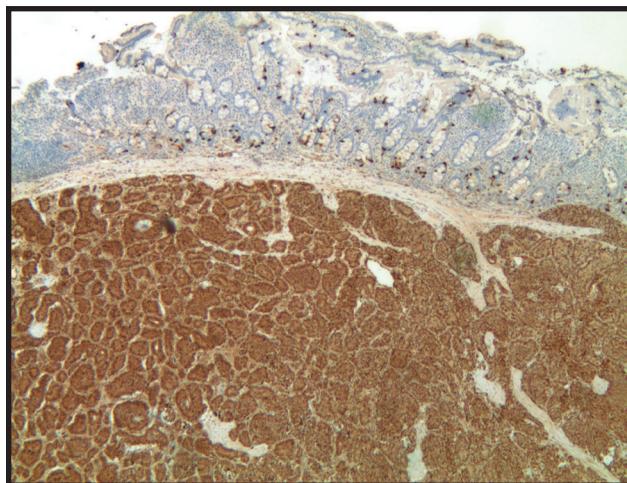
1. Ampullary tumor at duodenoscopy

(continued on page 64)

John M. Petersen, DO, FACP, FACP¹ David H. Holloman, MD² Baptist Medical Center^{1,2}
Departments of Gastroenterology,¹ Pathology¹ Borland-Groover Clinic¹ Jacksonville, FL

A CASE REPORT

(continued from page 62)



2. Post-ampullectomy site

ranged from 280 to 960 IU/L (normal 30-110 IU/L) and his lipase was 350 to 650 U/L (normal 25-250). Abdominal ultrasound and CT were negative. EUS revealed a 3.5 cm homogeneous, hypoechoic ampullary mass. The pancreatic duct was not dilated, the common bile duct (CBD) was 7 mm in the head of the pancreas and the pancreatic parenchyma was normal. The mass did not invade the CBD, pancreatic tissue or muscularis propria of the duodenum. (Figure 1). No adenopathy was noted; the liver had fatty infiltrate. After a saline lift, endoscopic ampullectomy was performed. (Figure 2) Post-removal ERCP revealed pancreas divisum with a normal accessory papilla. The CBD and pancreatic duct were patent and thus a stent was not placed. Pathologic evaluation revealed a neuroendocrine tumor consistent with a carcinoid tumor. There was no nuclear atypia, the margins were negative, and stains were positive for synaptophysin and chromogranin. (Figure 3) Histologically, the mitotic rate was 1/10 HPF. This was determined to be a low grade, well-differentiated tumor with +CD 56 and +Ki67 in only 2% of the cells.

The patient returned a month later and repeat endoscopy showed that there was no residual neoplastic tissue. Serum chromogranin-A was normal as was urine 5-HIAA and serum gastrin. An octreotide scan was negative for any foci of metastatic activity. Follow-up at 12 months revealed the patient asymptomatic, with a negative positron emission test (PET)/CT and octreoscan.

3. Carcinoid: benign, low-grade tumor with Chromogranin staining tumor cells

DISCUSSION

Carcinoid tumors of the ampulla of Vater are extremely rare, with less than 150 cases reported worldwide. These numbers suggest that ampullary lesions account for 0.05% of all carcinoid tumors. In contrast to carcinoid lesions of the mid-gut (jejunum, ileum and appendix), ampullary lesions very rarely cause the carcinoid syndrome, unless they are metastatic. Patients with these neoplasms often present with lingering abdominal pain and jaundice, if the CBD becomes encased. GI bleeds and weight loss or relapsing bouts of pancreatitis may occur.¹ In the 105 patients tabulated by Hartel, 47% presented with metastatic disease, and 58% underwent Whipple resection.² Metastases to regional nodes or the liver may be seen at the time of presentation. Carcinoid tumors of the ampulla may range in size from 0.5 cm to 6 cm. The time from first symptoms to actual diagnosis is typically greater than three months. Men and women are equally affected and the mean age at diagnosis is 48 years.³

Reports of relapsing pancreatitis have also been seen in carcinoid of the minor papilla.⁴ Cases of ampullary carcinoid prior to 2006 have typically been treated with duodenectomy, but as our case reveals, endoscopic ultrasound and endoscopic mucosal resection (EMR) can provide a sufficient non-surgical approach. EMR can be performed if the lesion invades into the submucosa, duodenal wall, CBD or pancreatic duct/parenchyma. Large tumors or those with nodal

involvement are best treated with Whipple resection.⁵ Carcinoid tumors of the ampulla are typically benign despite their large size, and have an overall five-year survival of 90%. Non-functioning carcinoid tumors constitute 15-20% of all pancreatic neuroendocrine tumors. These lesions seem to have a higher malignancy rate than their hormonally functioning counterparts. They tend to be slow growing, and metastatic disease does not preclude extended survival.⁶

This case accentuates the need to consider structural and mass lesions in pancreatitis not related to alcohol, autoimmune, or biliary tract disease. Endoscopic ampullectomy may be curative. ■

References

1. Simpson, WF, Adams, DB, Metcalf, JS, Anderson, MC, Non-functioning neuroendocrine tumors presenting as pancreatitis, *Pancreas* 1988; 3:223-231.
2. Hartel, M, Wente, MN, Sido, B, et al., Carcinoid of the ampulla of Vater, *Jnl Gastro and Hepatology* 2005; 20:676-681.
3. Ricci, JL, Carcinoid of the ampulla of Vater.local resection or pancreaticoduodenectomy, *Cancer* 1993; 71: 686-690.
4. Singh, VV, Bhutani, MS, Draganov, P, Carcinoid of the minor papilla in incomplete pancreas divisum presenting as acute relapsing pancreatitis, *Pancreas* 2003; 27: 96-97.
5. Hatzitheoklitos, E, Buchler, MW, Friess,H, et al., Carcinoid of the ampulla ofVater. Clinical characteristics and morphologic features, *Cancer* 1994; 73:1580-88.
6. Metz, DC, Jensen, RT, Gastrointestinal neuroendocrine tumors: Pancreatic endocrine tumors, *Gastro.* 2008; 135: 1469-92.

PRACTICAL GASTROENTEROLOGY

*Celebrating
40 Years
of Service*

practicalgastro.com

Answers to this month's crossword puzzle:

1	S	T	E	A	T	O	H	E	P	A	T	I	T	I	S				
	A		U	O	I	R		R		B					E				
9	L	A	P	A	R	O	S	C	O	P	I	C		10	C	C			
	I		E	R		T	B			I			11	P		R			
12	N	A	P	E		13	S	O	R	E			14	O	B	E	S	E	
	E		S		16	O	I	L					D		17	P	I	T	
		18	S	Y	M	P	T	O	M	A	T	I	C		19		20	P	I
21	I	N												22	D	P	T		O
	R		23	F	A	T	T	Y	L	24	I	V	E	R		25	P	N	
	O		I		Z											26	E		
27	N	A	S	H						28	F	I	B	R	O	S	I	S	30
			S		31	N		32	T	A	D		U			T			I
33	S	P	I	R	O	C	H	A	E	T	E			35	R	A	T		
	A		O		G		A				E				37	I	C	E	
38	P	E	N	T	O	X	I	F	Y	L	L	I	N	E					

Interactive Crossword and Answers
can also be found on our website:
practicalgastro.com