
Case of A Rare Ampullary Mass*

Hannah Celene O. Hernandez, MD

ABSTRACT

The ampulla of Vater, a common channel for biliary and pancreatic secretions, is where periampullary cancers develop. Identifying their anatomical source, the transitory nature of this area, its proximity to other structures and anatomical variances make up the diagnostic struggle with these tumors.

The rarest of them all, primary ampullary adenocarcinoma, has the best chance of survival even in advanced illness and has the highest rate of potentially curative resection. Four to six cases per million, or 0.5% of all gastrointestinal and 7% of all periampullary cancers, are reported annually.

Locating the level, extent and origin of obstruction and differentiating benign from malignant causes are the challenges in the diagnosis of obstructive jaundice. In order to identify the causes of blockage, imaging is crucial.

The gold standard in exploring the biliopancreatic tract is ERCP. Due to high complication rates linked to it, a less invasive, safe, and highly sensitive diagnostic procedure is urgently needed for patients with suspected bile duct or pancreatic duct anomalies.

Magnetic resonance cholangiopancreatography (MRCP) is a recently described, completely non-invasive technique that produces projectional images similar to those obtained with ERCP without the use of contrast agents. It became the standard technique of noninvasive investigation of the biliary tree to visualize intraluminal filling defects, ductal dilatation and strictures.

In this report, the author aimed to present the case of an elderly male patient who presented with blood streaked stool and jaundice. During his workup, which included contrast enhanced CT scan of the whole abdomen and MRCP, he was found to have dilated intrahepatic and extrahepatic ducts, and intraluminal thickening in the ampullary region. ERCP and tumor biopsy confirmed the diagnosis of ampullary adenocarcinoma.

Key words: Ampulla of Vater, ampullary adenocarcinoma

Periampullary malignancies may arise in the vicinity of the ampulla of Vater, an anatomical structure within the major duodenal papilla. It is a dilated channel that connects the common bile duct and major pancreatic duct, draining each organ's

secretions into the descending duodenum through the papillary aperture.

Primary ampullary malignancies are rare with an incidence of four to six per million representing 0.5% of all gastrointestinal and 7% of all periampullary malignancies despite being the most common site of neoplastic transformation in the small intestine.¹⁻³

According to histology, they may develop in the ampullary complex, pancreatic tissue, the distal bile duct wall, the duodenal wall, or the pancreatic tissue.

*From the Department of Radiology

The transition between these structures is one of the potential causes of the development of neoplasms, and such areas of transition are inherently unstable. As observed by Cattell and Pyrttek in 1949, the ampullary region is “an area of epithelium transition which is constantly being irritated chemically and mechanically”.

In comparison to tumors of pancreatobiliary origin, the biology of these tumors is more similar to that of small intestinal cancers. They have greater adenomatous characteristics histologically, and individuals with familial adenomatous polyposis and hereditary non-polyposis colorectal cancer have a higher prevalence of ampullary carcinoma, which suggests a shared pathway with colorectal carcinogenesis. These could be diagnosed earlier than the usual age for sporadic instances, which is in the sixth or seventh decade of life.¹⁻²

Most of the clinical presentation includes abdominal pain, nausea, dyspepsia, weight loss, malabsorptive diarrhea, and fatigue. Liver enzymes are elevated and uncommon symptoms may be present such as gastrointestinal bleeding or duodenal obstruction secondary to tumor ulceration. In up to 72%–90% of cases, the tumor’s strategic placement in the bile outflow system causes obstructive jaundice, which enables the early diagnosis of tiny tumors that are malignant.⁴

The diagnosis of an early stage tumor, identifying its anatomical origin and finding the level of obstruction present diagnostic challenges. However, neither specific markers for this disease nor conclusive methods for early diagnosis are currently available. Various imaging and procedures may be used in evaluating ampullary or periampullary tumor: ultrasound, CT, MRCP and ERCP. Each method has an advantages and disadvantages over the others and all of them are constantly undergoing development.

In order to establish the best course of treatment for each patient, radiologists evaluate biliary obstruction by evaluating its degree, scope, understanding its etiology and likely distinguish between benign and malignant.

The best method for examining the biliopancreatic area is still endoscopic retrograde cholangiopancreatography (ERCP). Given that ERCP is associated with high complication rates, there is a significant need for a less invasive, safe, and highly sensitive diagnostic technique for patients with suspected bile duct or pancreatic duct anomalies.

A growing interest in using magnetic resonance imaging to evaluate biliopancreatic disorders has been brought on by the emergence of fast imaging sequences and significant improvements in abdominal image quality.⁵ Several authors have evaluated the accuracy of MRCP in the diagnosis of common bile duct illnesses.^{6,7} It became the standard technique to visualize ductal dilatation, strictures as well as intraluminal filling defects. Without the addition of contrast agents, it projects images that are comparable to those obtained with ERCP.

The objective of this report was to present a rare case of ampullary adenocarcinoma in an elderly male patient.

THE CASE

This is a case of a 77-year-old male patient who was brought in the emergency room due to loss of consciousness. History of present illness started four days prior to admission, patient experienced one bout of blood streaked stool. There was no associated abdominal pain, fever, nausea, weight loss, appetite changes or vomiting.

One day prior to admission, patient had another episode of blood streaked stool, now associated with jaundice. Patient suddenly complained of blurring of vision, dizziness and then loss of consciousness.

Few hours prior to admission, patient once again experienced blurring of vision, dizziness and loss of consciousness, hence was brought in the emergency room where he was seen and examined and was subsequently admitted.

His past medical history was significant for hypertension and coronary heart disease. He was diagnosed with Transient Ischemic Attack last 2021. He underwent laminectomy in 2016.

Upon physical examination, patient was conscious, coherent and oriented to time place and person. He had jaundiced skin, pale nails and palpebral conjunctiva and icteric sclerae. The chest and abdomen were unremarkable. There were no palpable mass, abdominal tenderness or guarding. Dark brown colored stool was noted on the examination finger on rectal exam. Neurologic examination was also unremarkable.

Initial impression was upper gastrointestinal bleeding secondary to bleeding peptic ulcer disease, anemia secondary.

Initial medications were ordered and given. Patient was hydrated with intravenous fluids and transfused with packed RBC.

Laboratory examinations were requested. Pertinent findings of complete blood count revealed decreased RBC, hemoglobin and hematocrit suggestive of anemia, and increased WBC count suggestive of bacterial infection. Tumor markers like AFP, CEA and CA 19-9 were also done and showed normal AFP, elevated levels of CEA 5.6 ng/mL (0 – 3 ng/mL) and CA 19-9 of 1743.88 U/mL (0 – 37 U/mL).

Initial chest radiograph upon admission was requested and revealed hazy and reticular densities in the right paracardiac area, suggestive of pneumonia.

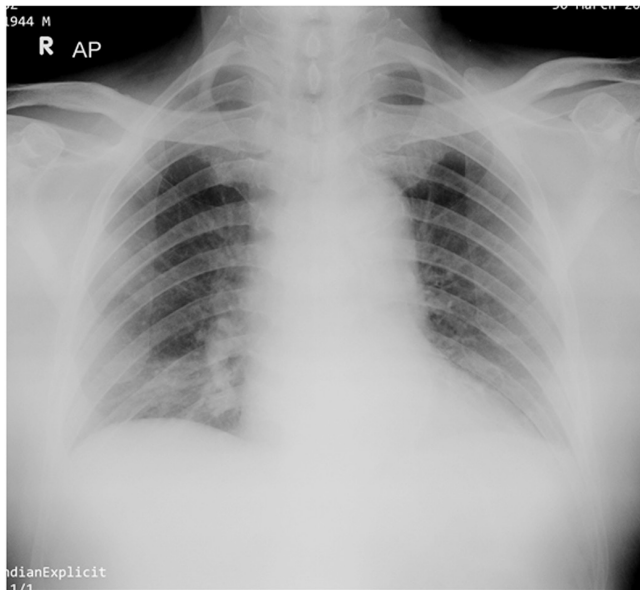


Figure 1. Chest radiograph of a 77 year old male. hazy and reticular densities in the right paracardiac area, suggestive of pneumonia.

Subsequently, contrast enhanced CT Scan of the whole abdomen was requested, facilitated and revealed normal liver with dilatation of both intrahepatic and common bile ducts measuring 2.1cm.

There were no obstructing lithiasis within the biliary tract or mass lesion seen in the CT scan. During the hospitalization, patient had persistence of jaundiced skin, and had another episode of melena and now with fever. No associated abdominal pain noted.

Magnetic resonance cholangiopancreatography (MRCP) was then requested for further evaluation, and revealed the intraluminal thickening with luminal filling defect in the ampullary region measuring approximately 1.5cm x 0.7cm x 1.0cm, suggestive of a neoplastic process. Histopathological correlation was recommended at that time.

Histopathologic findings of the ampullary mass revealed a fibrocollagenous tissues with well - differentiated neoplastic glands exhibiting pleomorphism and hyperchromatism and invading the underlying stroma. The overall diagnosis was well- differentiated ampullary adenocarcinoma.

The patient was then advised to have a surgical operation (Whipple's surgery), but he and his family decided to decline. Currently, his symptoms improved after the stent placement. He is well, alive and asymptomatic. There was no recurrence of jaundice, blood streaked stools or melena. His heart and hypertension medications were kept up to date, along with follow-up examinations.

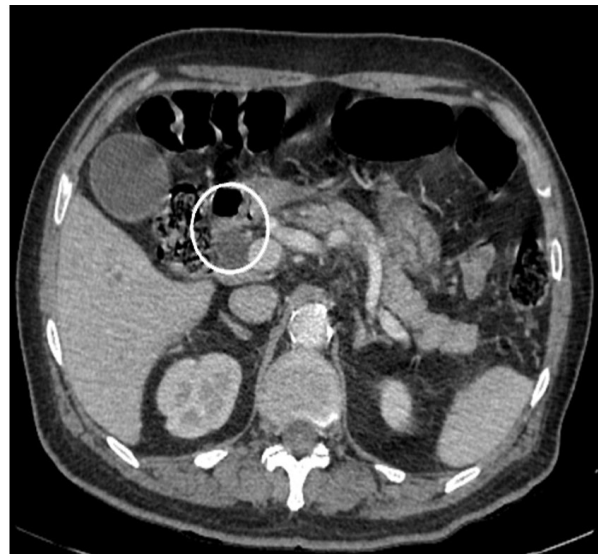


Figure 2. Contrast enhanced CT Scan of the abdomen. (A) Axial plain CT scan image showing dilatation of intrahepatic ducts (B) Axial contrast enhanced. Dilated common bile duct (green circle) measuring 2.1cm

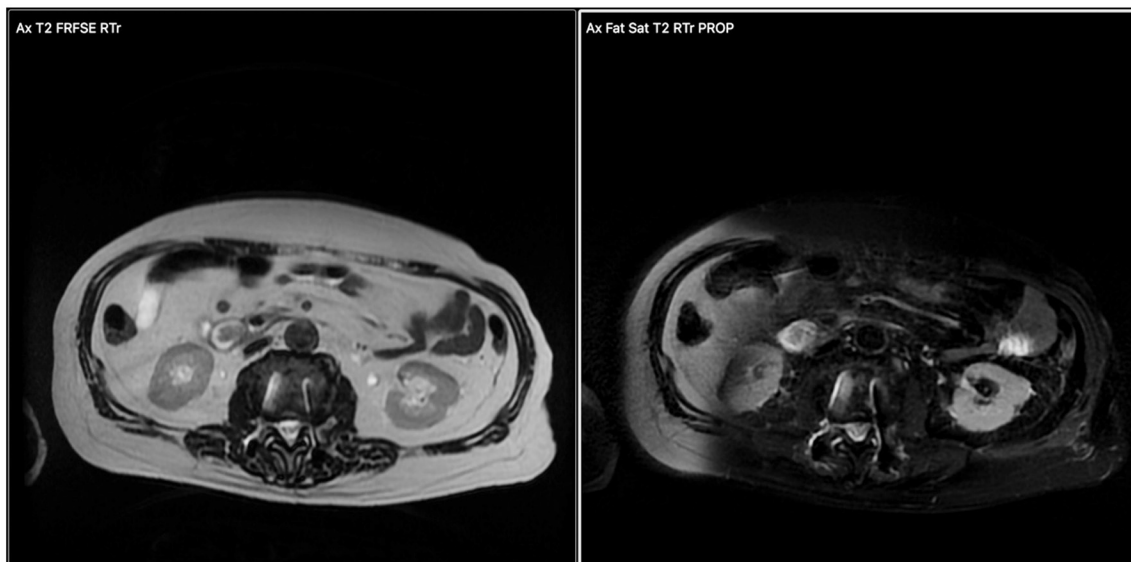
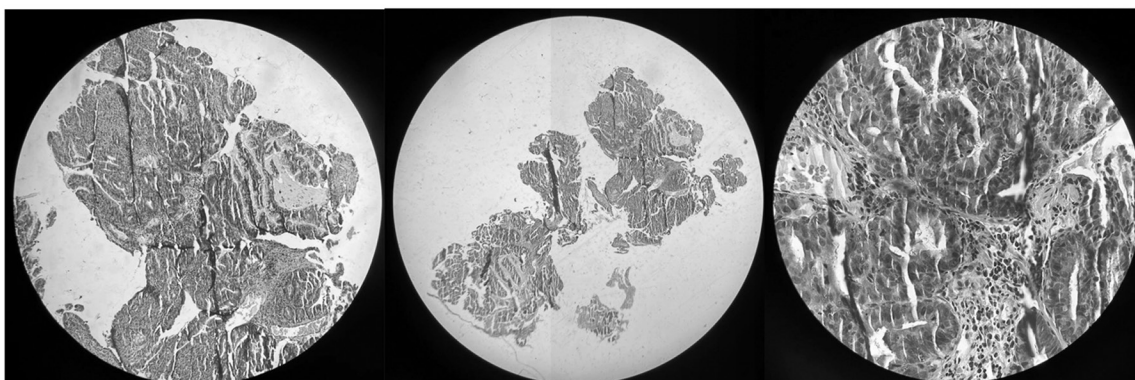


Figure 3. Magnetic resonance cholangiopancreatography (MRCP). Axial T2. There is an intraluminal thickening with luminal filling defect in the ampullary region (red circle) measuring approximately 1.5cm x 0.7cm x 1.0cm.



Figure 4. MRCP. Coronal view T2. Dilatation of the right and left hepatic ducts measure 0.8cm and 1.0cm, respectively. Likewise, there is dilatation of the common hepatic and common bile ducts, measuring 1.3cm and 1.8cm, respectively. The gallbladder is enlarged approximately measuring 10.4cm x 8.8cm x 3.5cm with no demonstrable filling defect intraluminally.



DISCUSSION

The glandular epithelium of the ampulla of Vater gives rise to ampullary carcinomas.¹³ Recent publications, however, have advocated that ampullary carcinoma should be categorized as a duodenal cancer since both have similar molecular developments and have better clinical outcomes than pancreatic and bile duct cancer. Additionally, the biliary and pancreatic ducts flow into the duodenum separately, and about 40% of people lack the common ampulla itself.

The most common clinical presentation of ampullary cancer is obstructive jaundice although other symptoms such as weight loss, general malaise, pruritis, abdominal pain, dyspepsia and malabsorptive diarrhea are also seen.² Because of the bile duct's erosion and intermittent permeability, the jaundice is often variable. Upper intestinal bleeding that results in anemia from ulceration is possible. However, in ampullary cancer initially presenting with gastrointestinal bleeding (blood streaked stool) as in the current case is uncommon.

One of the pertinent findings in this case is the elevated levels of CEA 5.6 ng/mL (0 – 3 ng/mL) and CA 19-9 of 1743.88 U/mL (0 – 37 U/mL).

A retrospective study by Kau Sy, et al. where 143 surgical patients with periampullary cancer from 1989 to 1997 had preoperative serum levels of CEA and CA 19-9 levels examined for prognostic significance. These researchers concluded that high CEA and CA 19-9 levels are significant diagnostic and prognostic value in periampullary tumors.

The presence of blood streaked stools and melena raised the initial suspicion of a bleeding peptic ulcer disease or perforated diverticulitis. Since jaundice is present, a stone-caused blockage of the hepatobiliary tract is also taken into consideration. An initial request was made for a contrast-enhanced CT scan of the whole abdomen. It revealed dilatation of the intrahepatic and extrahepatic ducts with no evidence of obstructing stone or mass lesion within the biliary tract. There were no bleeding ulcers or perforated diverticulitis found.

Magnetic resonance cholangiopancreatography (MRCP) was then recommended for further evaluation. It appeared as intraluminal thickening with luminal filling defect in the current study, which resulted to an upstream dilatation of the intrahepatic and extrahepatic ducts. The involved segment appeared as low signal intensity with narrowing and abrupt tapering of the distal CBD.

Ampullary carcinomas usually show at the time of diagnosis as tiny tumors due to the relatively early onset of their symptoms, and the mass itself is frequently not evident on imaging.¹³ They have a tendency toward intraluminal growth and frequently manifest with a polypoid or papillary form, and extraluminal extension is relatively infrequent.

According to the study "Differential diagnosis of Periapillary carcinomas at MR Imaging", by Joo Hee Kim, et.al, ampullary masses present as a hypointense lesion on T2-weighted images and typically had a diameter of 0.5cm to 1.8cm (mean: 1.0 cm). Eighteen (62%) of the 29 patients with ampullary cancer had a prominent nodular mass with an uneven filling defect at the distal border of the pancreaticobiliary junction (nodular type). Six people (21%) presented with aberrant periductal thickening around the pancreaticobiliary junction. Papillary bulging into the duodenum was seen in 19 people with nodular (n = 14, 48%) and periductal thickening (n = 5, 17%) tumor types. In 15 patients (52%), biliary and pancreatic ducts dilatation (double-duct sign) was appreciated, while the remaining 14 people only presented with bile duct dilatation.¹³

The evaluation of ampullary malignancy is still limited by MRCP, despite its development as an accurate diagnostic tool for pancreaticobiliary diseases. This is due to the region's tiny size and relatively complex structure, as well as the fact that there is little to no fluid in the tapering area of the distal biliary and pancreatic ducts. Evaluation of the ampullary region is also challenging due to the sphincter of Oddi's physiological contraction.

For optimal assessment of ampullary masses in terms of both detection and evaluation of the extent, the combination of conventional T1- and T2-weighted MR imaging with MRCP, including gadolinium-enhanced dynamic MR imaging, is important.

To distinguish between pathologic stenosis and physiologic constriction of the sphincteric segment, serial single-section MRCP images (kinematic MRCP) may be acquired. On T1- and T2-weighted MR images, the masses typically appear poorly to moderately enhanced and nearly isointense or hypointense. In light of this, variations in signal intensity characteristics cannot consistently indicate malignancy, and it is challenging to distinguish ampullary carcinomas from uncommon benign tumors using simply imaging findings.

For confirmed or suspected cases of ampullary adenocarcinoma, the standard surgical treatment is

pancreaticoduodenal resection (Whipple procedure). It involves removal a portion of small intestine (duodenum), pancreatic head, gallbladder and part of the common bile duct. Likewise, some cases may require a combination of chemotherapy and radiation.

SUMMARY AND CONCLUSION

It was proven in the current case that MRI-MRCP was more effective than contrast- enhanced CT scan in identifying the extent and cause of biliary blockage. The use of MRCP along with histopathological correlation is strongly advised when there is a high suspicion of ampullary or periampullary cancer.

For effective management and a favorable prognosis, the detection of small tumors, their tissue origin, and adequate staging are vital. As a result, radiologist must attempt to carefully examine the distal CBD in the setting of biliary obstruction and dilatation.

Additionally, it's important to know the patient's history, clinical presentation, laboratory and imaging results, as well as their personal and family history and any relevant risk factors. It significantly contributes in a successful examination and accurate diagnosis. If we investigators not aware of this information, their findings will be incidental, mistaken or they will only find obvious lesions. The objective is to understand what is being studied, and the possible procedures derived from the findings. By bearing the information in mind, they will know what it is they are looking for and the possibility of finding lesions, especially small ones, will increase.

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