Radiological spectrum of malignant tumors of the bile ducts

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Aims and objectives

The purpose of our study was to review the different types of malignant tumors of the bile ducts and to become familiar with the typical imaging findings and the spectrum of differential diagnoses.

Methods and materials

We report a retrospective study that evaluates 61 cases with malignant bile duct tumors, seen from January 2010 to January 2013. Most of the patients are elderly, the average age was 59 years and jaundice was the usual presenting symptom. As diagnostic imaging techniques, some patients underwent abdominal ultrasonography (n=53), MR cholangiography imaging (n=15) and all of them undergo abdominal CT scanning (n=61).

Results

The malignant bile duct tumors identified were as follows:

The hilar cholangiocarcinoma (n = 15) including three cases of the periductal infiltrating type (n=3) and twelve cases of the mass forming type (n = 12); Gallbladder cancer was diagnosed in 25 patients with intraluminal mass (n=6) and tumor invading liver (n=19); Cholangiocarcinoma of the distal bile duct (n=13); Vater's Ampulloma (n=8). Three cases were identified with pancreatic involvement (n=3), lymph-node involvement (n=2) and visceral metastases(n=5).

1) The hilar cholangiocarcinoma

Hilar cholangiocarcinoma (HCCA) or Klatskin tumors or perihilar cholangiocarcinomas are rare malignant tumor arising from the epithelium of the bile ducts that account for up to 3% of all gastrointestinal cancers.

Clinical features:

Obstructive jaundice is the main clinical feature and can appear relatively early, even with small neoplasms. It may progress rapidly or fluctuate.
Other symptoms may include weight loss, pruritus, right-upper quadrant pain or fever and chills if cholangitis develops.

**Diagnostic features and Imaging findings:**

Diagnostic features of HCCA include intrahepatic segmental biliary dilatation, periductal thickening, endoluminal lesions and direct tumor spread to the liver or adjacent vessels.

On the basis of the Japanese Liver Cancer Group classification, cholangiocarcinomas are classified into three types:

* **mass-forming cholangiocarcinoma (n=12):**

  - Utrasonography commonly used to confirm the presence of bile duct obstruction, to identify the extent of obstruction, and to determine the cause of obstruction. Most of time, it shows:
    
    - dilatation of the intrahepatic bile ducts
    - polypoid intraluminal masse (Fig. 1)
    - associated with wall thickening.

  - On CT and MRI: mass-forming intrahepatic cholangiocarcinoma is usually seen as a bulky lesion with infiltrating features around the adjacent peripheral branches of the portal vein. Mass-forming HCCA are usually heterogeneous hypovascular masses with rim-like peripheral enhancement in the arterial and portal phase and delayed enhancement in the equilibrium phase (Figures 1,2,3,4). The bile ducts peripheral to the tumor are usually dilated because of obstruction by the tumor. Lobar or segmental hepatic atrophy is usually associated with vascular invasion.

* **intraductal growing cholangiocarcinoma:**

  - Utrasonography demonstrated asymmetrical dilated bile ducts nodular and discrete smooth mass associated with wall thickening.

  - On CT or MRI: intraductal growing cholangiocarcinoma appears as a nodular, well defined mass and shows intense enhancement after contrast injection. The mass is confined within the bile ducts and there is preservation of the bile duct wall.

* **and periductal infiltrating cholangiocarcinoma (n=3)**

The most prevalent and forms the majority of perihilar cholangiocarcinomas.
- On ultrasonography, most infiltrating cholangiocarcinomas appear as mural and periductal soft-tissue thickening or focal irregularities of the bile duct.

- On CT, the involved bile ducts are diffusely narrowed or obliterated. It shows tumoral involvement as an irregular periductal thickening completely obstructing or narrowing a short segment of the biliary tree around the biliary bifurcation or common hepatic duct. Periductal thickening is usually iso or hypo enhancing in arterial and portal phases and shows marked enhancement on delayed phase imaging (Figure 5).

- On Magnetic resonance cholangio-pancreaticography (MRCP), non union of the right and left hepatic ducts is a typical finding of infiltrating hilar cholangiocarcinoma.

**Differential Diagnosis:**

- Choledocholithiasis
- Primary sclerosing cholangitis
- AIDS cholangiopathy
- Autoimmune cholangitis
- Inflammatory liver pseudotumor
- Lymphoplasmacytic cholangiopathy
- Mirizzi Syndrome
- Benign strictures (after invasive procedures)
- Parasitic infections (Ascaris lumbricoides, liver flukes)

**Treatment and prognosis:**

- 5 years survival: 30%
- Recurrence after liver transplantation: very common
- Surgical resection (#20% of tumors are resectable)
- Radiation, laser therapy and biliary stenting as palliative treatment.

**2- The gallbladder (GB) carcinoma**

Carcinoma of the gallbladder is a malignant epithelial neoplasm arising from gallbladder mucosa. It is the most common malignant tumour of the biliary tract and also the fifth
most common among malignant neoplasms of the digestive tract, characterized by silent presentation, poor prognosis, and limited therapy. The etiology of this tumor is complex, but there is a strong association with gallstones.

**Clinical presentation:**

* Similar signs and symptoms as seen in cholelithiasis & cholecystitis: nonspecific right upper quadrant (RUQ) abdominal pain, nausea and tenderness;

* Late, when tumor has invaded surrounding structures or metastasized. Most common symptoms: RUQ pains, anorexia, weight loss, jaundice, pruritus, vomiting are indicative of advanced disease.

Clinical profile: Elevated bilirubin and elevated alkaline phosphatase with biliary obstruction.

**Imaging Studies:**

-Ultrasonography is often the first imaging technique

* Early carcinoma can be identified as a fixed intraluminal polypoidal mass without acoustic shadowing or a complex intraluminal mass with localised thickening of the gallbladder wall.

* If the tumour is advanced, there is a loss of the interface between the gallbladder and liver. This feature is indicative of tumour invasion, metastasis to the liver and lymph nodes, dilated bile ducts, and ascites.

* Ultrasonography can also identify polyps and confirm the presence of associated gallstones.

- Computed tomography (CT) can accurately detect gallbladder abnormalities, diagnose the extent of the tumor and establish whether there is direct infiltration into adjacent tissues, vessels, nodal or distant metastases.

* Non enhanced CT scan may show calcification of GB wall or calcified gallstones.

* Contrast-enhanced CT scan identified the presence of an hypovascular mass completely occupying or replacing the gallbladder lumen, focal or diffuse asymmetric thickening of the gallbladder wall, or an intraluminal polypoid lesion (Figure 6 and 7).

* Also the presence of a mass in the gallbladder fossa, with the gallbladder itself being indiscernible was described as another common imaging finding.
- Magnetic resonance imaging (MRI) is particularly useful for detecting invasion of the hepatoduodenal ligament, portal-vein encasement, and lymph-node. The morphological appearance of GB carcinoma is similar to that obtained with CT.

* On T1-weighted images: tumors appear iso or hypointense with increased signal compared to normal liver, and hypovascular after IV gadolinium.

* On T2-weighted images: the mass appear slightly increased in signal intensity compared to liver.

* Magnetic resonance cholangio-pancreaticography (MRCP) provides information of dilated bile ducts due to common hepatic duct obstruction.

- Nuclear medicine findings, using a hepatoiminodiacetic acid (HIDA) scan can show obstruction of the bile duct or gallbladder disease, but cannot pinpoint the cause of the obstruction.

**Differential Diagnosis:**

- Gallbladder polyp
- Complicated cholecystitis
- Adenomyomatosis
- Metastatic disease to gallbladder fossa (hepatocellular carcinoma, cholangiocarcinoma...)

**Treatment and prognosis:**

- Very poor prognosis: 5% 5 year survival rate and 75% of patients have metastases at time of diagnosis.

- Cholecystectomy for lesions confined to GB wall without liver invasion.

- Radical cholecystectomy and/or partial hepatectomy with regional node dissection for lesions infiltrating porta hepatis.

**3) The Cholangiocarcinoma of the distal bile duct**

* It’s an extrahepatic bile duct cancer, relatively a rare disease in which malignant cells form in the part of bile duct that is outside the liver.
Extrahepatic cholangiocarcinomas can be classified according to macroscopic growth pattern as:

- nodular
- sclerosing
- papillary

* In terms of the distribution of large duct (extrahepatic) tumours:

  # upper third of the duct including the hepatic hilum: 50-75%
  # middle third CBD: 10-25%
  # distal third CBD: 10-20%

* These tumours are most commonly infiltrating, although both exophytic (mass-forming) and polypoid (intraductal) types.

* They have similar clinical and radiological appearances to their intrahepatic counterparts (Figure 8).

* Involvement of the periductal lymphatics, nerves, and perineural tissue is frequent.

**Treatment and prognosis:**

- Very poor prognosis
- Pancreaticoduodenectomy remains the mainstay of treatment of distal bile duct malignancy.

**4) Vater's Ampulloma**

Ampullary carcinomas are neoplasms that arise from the glandular epithelium of the ampulla of Vater. These kind of cancers can be divided into three types according to gross morphologic features: protruded, ulcerative, and mixed.

**Clinical presentation:**

* Similar signs and symptoms as seen in other tumors of the bile ducts.

**Imaging Studies:**

* Transabdominal ultrasound:
- is the initial imaging method to evaluate the common bile duct (CBD) and pancreatic duct (PD), but it cannot identify the ampulla of Vater itself.

- "Double duct" sign with obstruction of both CBD and PD is essentially diagnostic for extrahepatic obstruction and is highly suggestive of ampullary or periampullary tumors.

- 10 to 15% of patients with normal common bile duct findings on ultrasonography demonstrate extrahepatic biliary obstruction on a computed tomography (CT) scan.

- Ultrasonography can also help to reveal metastatic disease in the liver or regional lymph nodes.

* Contrast-enhanced CT scans:

- may assess for the presence of biliary ductal dilation, examine the periampullary region, differentiate between causes of distal biliary obstruction, assess for the presence of malignant-appearing lymph nodes, evaluate the local region of interest and evaluate for possible metastases and establish the relationship of the tumor to nearby abdominal vascular structures.

- often demonstrates a mass but is not helpful in differentiating ampullary carcinoma from tumors of the head of the pancreas or periampullary region especially if the lesion is smaller than 2 cm (Figure 9).

- Such findings as pancreatic and/or bile duct dilation are highly suggestive of pancreatic and periampullary malignancy and require further evaluation, usually with endoscopic retrograde cholangiopancreatography (ERCP) (Figure 10).

* MRI with magnetic resonance cholangiopancreatography MRCP:

- can visualize the biliary tract and pancreatic ducts in great detail as well as identify pancreatic, biliary, or ampullary pathology.

- ampullary carcinomas appear as masses with periductal thickening and the tumor itself often appears as a filling defect protruding into the duodenal lumen, with a characteristic delayed enhancement (Figure 11).

- MRCP has a sensitivity of 94%, specificity of 82%, positive predictive value of 89%, and negative predictive value of 90% for the detection of malignant causes of bile duct obstruction.

* Endoscopic retrograde cholangiopancreaticography (ERCP) with or without an endoscopic biopsy:
- is the method for making a definitive diagnosis of disease in or around the ampulla of Vater.

- It allows to direct visualization of the ampulla and tissue acquisition via biopsy, also to provide therapeutic interventions such as pancreatic, biliary sphincterotomy and/or biliary stenting to treat jaundice.

* Positron emission tomography (PET) - CT scans can detect metastases that are too small to be reliably detected on a CT scan.

**Differential Diagnosis:**

- Neoplastic causes:
  - Cancer of the pancreas head
  - The lower bile duct Cholangiocarcinoma
  - Duodenal cancer

- Non-neoplastic causes:
  - Lithiasis of the CBD;
  - Parasitic barrier: cyst, roundworms or flukes.
  - Sclerosing cholangitis;
  - Pancreatitis
  - Inflammatory stenosis of the bile duct.
  - Sphincter of Oddi dysfunction
  - Diverticulum juxta-ampullary
  - Benign papilloma ‘forced’ migration after gallstone.
  - Duodenal adenoma
  - Ampullary adenoma

**Treatment and prognosis:**

- Survival depends on nodal and distal metastases at presentation

- Pancreatoduodenal resection in patients with good operative risk (whipple procedure).

**Images for this section:**
**Fig. 1:** Mass forming cholangiocarcinoma. Axial CT scan in early portal phase shows an hypovascular mass (arrow) with peripheral enhancement. The lesion is associated with ductal infiltration and biliary dilation.
Fig. 2: Mass forming cholangiocarcinoma. Axial CT scan in delayed phase shows an hypovascular mass (arrow) with peripheral enhancement. The lesion is associated with ductal infiltration and biliary dilation.

Fig. 3: Mass forming cholangiocarcinoma. Axial CT scan showing a Klatskin tumor, a central cholangiocarcinoma at the porta hepatis (arrow) with dilated and tortuous intrahepatic bile ducts.
Fig. 4: Coronal CT scan image of the liver reveals a tumoral mass (arrow) at the liver hilum causing thickening of the bile duct and intrahepatic biliary obstruction.
Fig. 5: Contrast enhanced axial CT image in the portal phase at the level of the hepatic hilus shows an irregular periductal thickening completely obstructing the common hepatic duct with intrahepatic biliary dilatation consistent with periductal cholangiocarcinoma.
**Fig. 6:** Contrast-enhanced axial CT scan during portal venous phase shows focal nodular thickening (arrow) and diffuse asymmetric gallbladder wall thickening.
Fig. 7: Contrast-enhanced coronal CT scan image during portal phase shows intraluminal polypoid enhancing gallbladder mass (arrow) with asymmetric irregular wall thickening.
**Fig. 8:** Contrast-enhanced axial CT images show an extrahepatic cholangiocarcinoma as a mass encircling choledocal lumen with dilatation of intrahepatic bile ducts and common bile duct.
**Fig. 9:** Contrast-enhanced axial CT scan shows mild bulging of the papilla with increased targetlike enhancement (arrow).
Fig. 10: Contrast-enhanced axial CT image shows mild dilatation of the upstream bile duct and cystic branch duct (yellow arrow), also a dilated main pancreatic duct (white arrow), secondary to Vater’s ampulloma.
Fig. 11: On the T2-W SSFSE sequence (A) and 2D Bili-MRI (B) show the presence of an expanding intrahepatic bile ducts (IBD) and common bile duct (CBD) due to carcinoma of Vater's ampulloma.
Conclusion

Most of the bile duct tumors are malignant. The diagnosis is based on a combination of clinical and imaging findings of the endoscopic ultrasonography, MR Cholangiopancreatography and abdominal CT scan, which provide in-depth information in support of etiological diagnosis, treatment, and other clinical decision-making.

Personal information

Dr Omar ADDOU is a resident in radiology, participated in many conferences and radiology workshops.

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