Spectrum of Cholangiocarcinoma

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Abstract: We present here 8 cases of cholangiocarcinoma with spectrum of location extending from peripheral, intra hepatic and extra hepatic bile ducts. The tumor can arise from any part of bile duct epithelium i.e. from terminal ductules to the ampulla of vater and also involve peri biliary glands (intra and extra mural). Cholangiocarcinoma is usually classified as intra hepatic or extra hepatic cholangiocarcinoma. Intrahepatic cholangiocarcinoma is further classified as peri hilar or hilar [1, 2]. Cholangiocarcinoma is usually classified as intra hepatic or extra hepatic cholangiocarcinoma. Intrahepatic cholangiocarcinoma is further classified as peri hilar or hilar [1, 3]. There is association of cholangiocarcinoma with intra hepatic stone disease, cholelithiasis, Caroli disease, clonorchiasis, primary sclerosing cholangitis and inflammatory bowel disease [5]. The tumor is classified as extra hepatic (87-92 %) or intrahepatic (8-13 %) [6]. Depending on the site of origin it presents with wide spectrum of radiologic appearances. The intra hepatic subtype, perihilar is believed to arise peripheral to secondary bifurcation of the left or right hepatic duct and the hilar subtype arises from one of the hepatic ducts or the bifurcation of the common hepatic duct [4].

Peripheral cholangiocarcinoma classically manifests as a large, well-defined hepatic mass with lobulated margins and peripheral rim enhancement at both contrast material–enhanced spiral computed tomography (CT) and dynamic magnetic resonance (MR) imaging [7].

Hilar cholangiocarcinoma typically takes one of three shapes: infiltrative, exophytic, or polypoid. Most extra hepatic cholangiocarcinomas are infiltrative, causing a focal stricture of the bile duct.

CASE REPORTS

CASE 1
A 60 years female patient came with complaints of jaundice and weight loss. CECT of abdomen showed dilated IHBR and proximal CBD due to ill defined mixed density mass in distal CBD. Few metastatic deposits were seen in right lobe of liver.

CASE 2
A 55 years male presented with jaundice. CECT of abdomen showed dilated IHBR & hepatic ducts. Hypodense mass was seen in common hepatic duct.

CASE 3
A 70 years male came with complaints of jaundice and loss of appetite. CECT of abdomen showed dilated IHBR and proximal CBD due to hypodense mass in distal CBD.

CASE 4
A 79 years female came with complaints of jaundice and weight loss. CECT of abdomen showed dilated IHBR due to mass in proximal CBD.
CASE 6

A 64 years male came with complaints of jaundice. CECT of abdomen showed infiltrating mass in peripheral intrahepatic biliary tree with retraction of margins of right lobe of liver.

CASE 7

A 56 years male came with complaints of jaundice & loss of appetite CECT of abdomen showed dilated IHBR and proximal CBD due to mass in distal CBD.

CASE 8

A 59 years male patient presented with jaundice. MRCP revealed dilated IHBR and CBD due to abrupt narrowing in midsegment of CBD.

Fig. 1: CASE 1: CECT abdomen image (A) & (B) shows dilated IHBR and proximal CBD (black arrows). Image (C) shows mixed density mass lesion in distal CBD (green arrow).

Fig. 2: CASE 2: CECT abdomen image (A) shows dilated IHBR (black arrow). Image (B) shows hypodense mass in common hepatic duct (Green arrow).

Fig. 3: CASE 3: CECT abdomen image (A) & (B) shows dilated IHBR and proximal CBD (black arrows). Image (C) shows hypodense mass lesion in distal CBD (green arrow).
Fig. 4: CASE 4: CECT abdomen image (A), (B) & (C) shows mass in distal CBD (green arrow) near ampulla of vater. Black arrow demonstrates second part of duodenum.

Fig. 5: CASE 5: CECT abdomen image shows mass in proximal CBD (green arrow).

Fig. 6: CASE 6: CECT abdomen image shows infiltrating mass in peripheral intrahepatic biliary tree with retraction of margins of right lobe of liver (green arrow).

Fig. 7: CASE 7: CECT abdomen image shows hypodense mass in distal CBD (green arrow).
DISCUSSION
Classification of cholangiocarcinoma is done for understanding of imaging features and giving differential diagnosis. It also helps to understand the progression of disease and planning of surgery. Liver cancer study group of Japan [8] proposed morphological classification and classified cholangiocarcinoma as mass forming, periductal infiltrating and intraductal growth.

Mass forming type presents morphologically as a homogenous mass with irregular margins. The lesion shows irregular peripheral followed by gradual centripetal enhancement. Retraction of liver capsule adjoining the mass is generally seen. Few satellite nodules may be noted. The lesion is associated with dilatation of biliary tree in vicinity of the tumor. Surrounding normal liver parenchyma is compressed and may sometimes give appearance of a peripheral ring [9]. Branches of portal vein are often engulfed, but intramural thrombus within the vessel is a rare finding [10-13].

Periductal infiltrating type presents with growth along bile duct without mass formation. It manifests as speculated or branchlike abnormality. Diffuse periductal thickening with enhancing walls is seen as a result of tumor infiltration. Abnormally dilated or narrowed duct and peripheral ductal dilatation is noted as well. This presentation is commonly seen in hilar type of cholangiocarcinoma.

The periductal infiltrating type needs to be differentiated from benign stricture in early stage and from periportal lymphangitic metastasis in late stage. Malignant stricture on CT is long-segment having an irregular margin, asymmetric narrowing of the duct with enhancement. Periductal soft tissue is seen associated with the stricture along with surrounding enlarged lymph nodes [16, 17].

Intraductal cholangiocarcinoma has an atypical presentation. The variety of CT appearances are - Diffuse duct dilatation with or without a papillary mass, intraductal polypoid mass within focally dilated duct, intraductal cast like lesion within mildly dilated biliary duct or focal stricture with resultant proximal duct dilatation [14, 15].

CONCLUSION
The incidence of cholangiocarcinoma is increasing and it has diverse presentations depending on its type. Hence it is important to characterize the cholangiocarcinoma on basis of imaging findings so as to help in surgical management and predict its extension and prognosis.

ACKNOWLEDGEMENT
Authors acknowledge the great help received from the scholars whose articles cited and included in references of this manuscript. The authors are also grateful to authors / editors / publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed. Authors are grateful to editorial board members and team of reviewers who have helped to bring quality to this manuscript.

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