Case Report

Intraductal Papillary Mucinous Tumor Of Common Bile Duct – A Rare Disease Mimicking A Rare Pancreatic Malignancy

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Abstract :

Intraductal papillary mucinous neoplasms are known malignant lesions of pancreas but recently a similar tumor has been reported and recognized by WHO in 2010 which involves the bile ducts. The involvement of common bile duct by this malignant tumor is extremely rare. Intraductal papillary mucinous tumors of bile ducts have been found to have much better prognosis than the other cholangiocarcinoma. We hereby present a case of an elderly lady who presented to the surgery department of our hospital with obstructive jaundice and was followed up on a six-monthly computed tomography study.

Keywords: Bile duct, Computed tomography and Intraductal papillary mucinous tumor

Introduction

Intraductal papillary mucinous neoplasm (IPMT) is a known entity-involving pancreas [1]. Its counter part in bile ducts is intraductal papillary neoplasm (IPNB) [2]. Recently incidence of papillary neoplasms involving bile ducts has been extensively reported. These mainly comprise of involvement of intrahepatic biliary system with very few literature on involvement of common bile duct. In a study published in 2012 author had reflected that until then only one case of IPN involving common bile duct had been reported in literature [3].

IPMT are uncommon ductal epithelial mucin producing tumors of pancreatic ductand comprise approximately 10-15% of cystic pancreatic neoplasms [4,5]. In recent years intraductal papillary neoplasm involving the bile ducts has come into prominence with WHO recognizing it as a distinct clinical and pathological entity in 2010 [6]. These first caught attention as mucin producing tumors of the bile ducts and were given various names such as mucin-producing cholangiocarcinoma, mucin-hypersecreting bile duct tumor, and intraductal papillary mucinous tumor of the bile duct [7]. They have been found out to be biliary counterparts of intraductal papillary mucinous tumors of pancreas because of their similar morphologic features [8]. Based on histological diagnosis intraductal papillary neoplasms of bile ducts have an incidence of approximately 9% of all bile duct tumors [9].The IPNB has a better outcome than the more common nodular-sclerosing type of cholangiocarcinomas [5].

We here present a case of an elderly (64-year-old) female who presented with signs and symptoms of obstructive jaundice and was

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diagnosed with moderately differentiated papillary carcinoma of extrahepatic bile duct extending upto ampullary and periampullay region.

Case report

A 64-year-old female presented to the gastrosurgery department of our hospital with clinical features of obstructive jaundice. The patient was advised to undergo routine hematological, liver function tests and ultrasound abdomen.

The serum bilirubin levels were 14.7 mg/dl (total), 8.3 mg/dl (direct) & 6.4 mg/dl (indirect). The SGOT, SGPT & serum alkaline phosphatase levels were also raised and were 162 IU/l, 77 IU/l & 365 IU/l respectively. The serum amylase level of the patient was severely raised and measured 3208 IU/l. Hematological investigations showed raised Erythrocyte Sedimentation Rate of 55mm. Rests of the routine hematological investigations were within normal limits. The patient was HIV non-reactive.

Ultrasound of the abdomen was done using Siemens Acuson X 300 machine. Mild hepatomegaly with moderately dilated intrahepatic biliary radicals (IHBRs) was seen. A large echogenic lesion (measuring approximately 5.6cm x 2.5cm) was seen in distal part of dilated common bile duct (CBD) (measuring 23mm in luminal diameter). The lesion showed no obvious vascularity. Sludge was noted in gall bladder lumen. Based on these findings the patient was advised a contrast enhanced computed tomography (CECT) scan, which was performed on 128-slice Dual Source Siemens Somatom Definition Flash scanner. CECT revealed a large relatively homogeneously enhancing tubular shaped isodense mass completely filing the entire extent of CBD. Biliary confluence was formed with dilated IHBRs seen in both lobes of liver. The mass was seen to be lying at distance of 2cm from the floor of the biliary confluence. The lesion/ CBD had well defined fat planes with the adjacent structures. No significant lymphadenopathy or ascites were noted. A diagnosis of cholangiocarnima was made in accordance with the CECT findings.

Whipple's operation was performed without any complications after due pre- anesthetic check up. Histopathology was suggestive of moderately differentiated papillary carcinoma of extrahepatic bile duct and extending upto the ampullary and periampullary region. Duodenal mucosa, pancreas, resected margins of the tumor (containing stomach) & mesentery were free of tumor deposits.

A review scan was advised on six months follow up.

The follow up CECT revealed no radiological evidence of residual/ recurrent lesion. The presence of air foci in IHBRs suggested patency of the new surgically formed biliary system. Patient also had no complaints on follow up.

Discussion

Intraductal papillary neoplasm of the bile duct (IPNB) is a uncommon variant of the bile duct tumors, which is characterized by papillary or villous growth within the bile duct lumen.

Previously they were known by various names such as mucin-producing cholangiocarcinoma, mucin-hypersecreting bile duct tumor and intraductal papillary mucinous tumor of the bile duct and were said to be biliary counterpart of the intraductal papillary mucinous neoplasm of the pancreas. Variants of biliary intraductal tumors with papillary growth pattern but no mucin production have

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also been identified. In 2010 WHO classification, it has now been identified as a distinct clinical and pathological entity.

IPNB is defined as a biliary epithelial tumor with exophytic nature exhibiting papillary mass within the bile duct lumen and with prominent intraductal growth pattern [7]. It can involve either or both of the intrahepatic and extrahepatic bile ducts. According to 2010 WHO classification, IPNB has been classified into IPN with low- or intermediate- grade intraepithelial neoplasia, IPN with high-grade intraepithelial neoplasia and IPN with an associated invasive carcinoma. These papillary tumors may or may not discharge mucin.

IPN with low- or intermediate- grade intraepithelial neoplasia, IPN with high- grade intraepithelial neoplasia are considered ' premalignant' lesions [10].

Histopathological studies inclusive of immunohistochemistry have proved that as we go down the biliary tree the malignant potential of intraductal papillary neoplasms of bile ducts tends to decrease [11]. Histopathologically more than one-third of IPNB has been found to be papillary adenocarcinomas in situ [1].

Most common clinical complaint of patients with cholangiocarcinoma is abdominal discomfort [8] as was present in our case.Dilatation of proximal intrahepatic biliary radicals and proximal CHD/CBD and deranged liver function tests were seen in our patient. IPNB lesions have a tendency to spread superficially along the bile duct mucosa and secrete large amounts of mucin which may disturbs bile flow and causes flow disturbances [12].

Presence of different immunohistochemical markers in IPNB like MUC2, CDX2 and CK20 as compared to MUC1 in non-papillary cholangiocarcinomas has been proposed as factor in better prognosis of IPNBs. IPNB have been histopathologically classified as pancreaticobiliary, intestinal and gastric types or adenocarcinoma, adenoma, dysplasia and mixed types [1,2].

On imaging, bile duct carcinomas including intraductal papillary neoplasms are associated with bile duct dilatation [13] which may be asymmetrical [2].

We found an echogenic lesion on USG in our case. Literature shows that IPNB are usually seen as echogenic lesions on ultrasound with no posterior acoustic shadow and are hypovascular in nature. However there are case reports about hypoechoic tumors seen on ultrasound. [2]

CT presents the intraluminal soft-tissue mucinous neoplasms with hypoattenuating or isoattenuating post contrast enhancement patterns. The enhancement pattern depends on weather the tumor is attached to the bile duct wall or not. Computed tomograpy can also show the maintance of integrity of the bile duct wall [13]. We also saw that the intraluminal soft tissue lesion was isoattenuating on pre and post contrast (portal phase) studies compared to surrounding structures. However bile duct dilatation is almost always seen, sometimes because of mucin secreation by the papillary tumor [2,14].

ERCP also provides information about presence and site of obstruction along with depicting the amount of mucin secreations which might be responsible for bile duct dilatation [15].

Histopathology is often required for definite diagnosis [2].We although were able to make the definitive diagnosis of cholangiocarcinoma

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also relied on histopathology for formation of definite diagnosis. However retrograde study of the case suggests presence of highly suggestive radiological features of the lesion in accordance with IPNB, especially on multi detector computed tomography.

Figure 1: Non-Contrast Axial computed tomography image showing Isodense lesion in CBD.

Figure 2: Early arterial phase computed tomography axial image showing relatively homogeneous enhancement of the lesion.

Figure 3: Portal phase computed tomography axial image showing relatively isodense post contrast enhancement of the lesion.

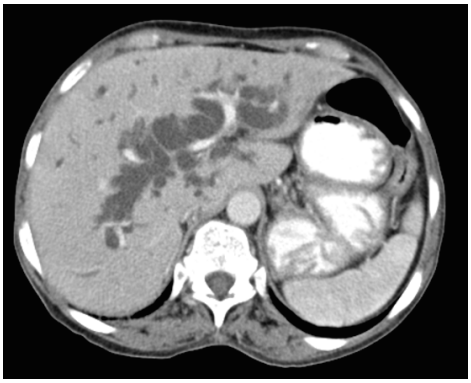

Figure 4: Axial post contrast computed tomography image showing grossly dilated intrahepatic biliary radicals.

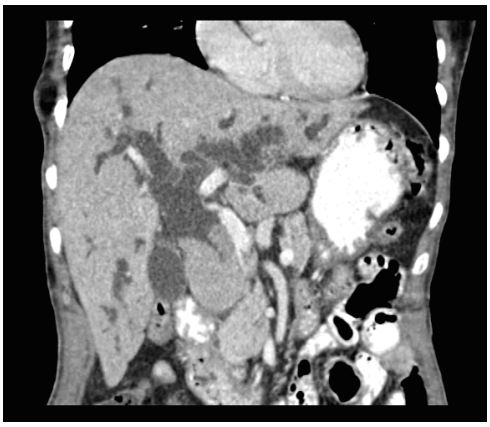

Figure 5: Coronal reformatted post contrast image showing large intraluminal mass in common bile duct with dilated intrahepatic biliary radicals.

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Figure 6: Follow up post contrast axial computed tomography image showing presence of air in intrahepatic biliary radicals suggesting competence of operative procedure on 6-month follow up study.

Figure 7: Follow up post contrast axial computed tomography image showing no recurrence or residual lesion on 6-month follow up study.

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