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Isolated Extrahepatic Intraductal Papillary Neoplasm of the Bile Duct: A Rare Type of Bile Duct Tumour

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Introduction- Intraductal papillary neoplasm of the bile duct (IPNB) is a relatively rare type of bile duct tumor. The neoplasm features an intraductal papillary growth of neoplastic biliary epithelia with a fibrovascular stalk.¹ Its nomenclature was given regarding the tumor characters, for example, biliary papillomatosis², mucin-secreting bile duct adenoma³, mucin-producing bile duct tumor⁴, or mucin-producing cholangiocarcinoma.^{5,6} Term “intraductal papillary neoplasm of the bile duct” is a recent terminology.⁷ According to the latest classification of bile duct tumors, benign IPNB is classified as a pre-malignant lesion of intraductal - growth and papillary type of cholangiocarcinoma.⁸ The development of IPNB follows an adenoma-carcinoma sequence that correlates with the stepwise activation of common oncogenic pathways.⁹ IPNB usually progresses slowly bearing a better prognosis compared to the conventional cholangiocarcinoma.¹⁰

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Isolated Extrahepatic Intraductal Papillary Neoplasm of the Bile Duct: A Rare Type of Bile Duct Tumour

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I. INTRODUCTION

Intraductal papillary neoplasm of the bile duct (IPNB) is a relatively rare type of bile duct tumor. The neoplasm features an intraductal papillary growth of neoplastic biliary epithelia with a fibrovascular stalk.¹ Its nomenclature was given regarding the tumor characters, for example, biliary papillomatosis², mucin-secreting bile duct adenoma³, mucin-producing bile duct tumor⁴, or mucin-producing cholangiocarcinoma.^{5,6} Term "intraductal papillary neoplasm of the bile duct" is a recent terminology.⁷ According to the latest classification of bile duct tumors, benign IPNB is classified as a pre-malignant lesion of intraductal-growth and papillary type of cholangiocarcinoma.⁸ The development of IPNB follows an adenoma-carcinoma sequence that correlates with the stepwise activation of common oncogenic pathways.⁹ IPNB usually progresses slowly bearing a better prognosis compared to the conventional cholangiocarcinoma.¹⁰

The principle of intraductal papillary neoplasm of the bile duct treatment is similar to that of cholangiocarcinoma. The primary treatment of choice for an operable case is surgical resection. The systemic adjuvant chemotherapy and radiation in IPNB remain controversial.

II. CASE PRESENTATION

A 60-year-old Thai male had been experiencing right upper quadrant abdominal discomfort, progressive jaundice with generalized pruritis and loss of appetite for two weeks. He did not notice urine and feces discoloration. He had been diagnosed with pulmonary tuberculosis undergoing regular anti-TB medication 2 months earlier. Otherwise, he had been healthy with no history of parasitic infestation or hospitalization. Physical examination revealed obvious icteric sclera. Initial laboratory tests included total bilirubin 19.47 mg/dL, direct bilirubin 12.77 mg/dL, aspartate aminotransferase (AST) / alanine aminotransferase (ALT) 74/52 IU/L (<35 IU/L), and alkaline phosphatase (ALP) 371 IU/L (46-116 IU/L).

He was scheduled for endoscopic retrograde cholangiopancreatography (ERCP) which demonstrated

dilatation of the common bile duct with the mucinous plug at the ampulla of Vater (Figure 1, 2). The contrast did not fill intrahepatic duct. The imaging includes magnetic resonance imaging with magnetic resonance cholangiopancreatography (MRI with MRCP) demonstrated a severe narrowing with surrounding soft tissue mass at distal common bile duct. Neither lymph node nor distant organ metastasis was detected.



Fig. 1: ERCP Demonstrated a Dilatation of Common Bile Duct with Mucinous Plug in Common Bile Duct



Fig. 2: ERCP Demonstrated a Mucinous Plug at Ampulla of Vater

The patient underwent the endoscopic ultrasound (EUS) one day after his imaging. Diffusely dilated intrahepatic and common bile duct with an irregular intraductal polypoid lesion at the distal

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common bile duct causing the obstruction were found. The ultrasound revealed an enlarged lymph node size 1.8 cm at the pancreatoduodenal groove. Elastography showed a heterogeneous pattern. The pancreas and its duct appeared normal.

The imaging concluded a distal bile duct obstruction suggestive of intraductal papillary neoplasm of the bile duct (IPNB). The patient was in good performance status and the limitation of disease only in the bile duct; we planned to perform surgery.

Three weeks before the surgical appointment, he underwent right percutaneous transhepatic biliary drainage (PTBD) using 8 Fr drainage catheter with a tip at the hilar region. Cholangiogram showed dilated intrahepatic duct without demonstrable mass nor stones.

The operation was carried out as scheduled; an upper midline incision was performed. Regarding intraoperative findings, apart from the lesions depicted on preoperative imaging, there were several enlarged lymph nodes at station 7, 8, and 12. Common bile duct size was 2 cm in diameter. We performed Pancreaticoduodenectomy (PD). Choledochoscope allowed clear visualization of the mucinous plug at distal common bile duct and no other lesion at proximal bile duct nor intrahepatic duct. Intraoperative frozen section confirmed uninvolved margin. Total operative time was 7 hours and estimated blood loss was 400 ml. The intraoperative and immediate postoperative course was uneventful.

The surgical specimens consisted of distal part of stomach attached to a segment of the duodenum, head of the pancreas, 2 cm in length of the distal common bile duct and gallbladder. Examination of specimens disclosed an ill-defined firm gray-white mass measuring 2.2*1.5*0.5 cm at the distal common bile duct. Multiple enlarged lymph nodes were seen in mesenteric fat of duodenum, lymph node station 7, 8 and 12. (Figure 3).



Fig. 3: Papillary Growth Lesion at the Distal Common Bile Duct

The histopathology report revealed well-differentiated adenocarcinoma of the distal common bile duct. Sections showed marked dysplasia of bile duct mucosa with significant mural fibrosis (Figure 4, 5, 6).

However, the definite malignant gland is not identified at common bile duct. There was a metastasis of well-differentiated adenocarcinoma in 4 regional lymph nodes (surrounding distal common bile duct mass) (Figure 7). Duodenal wall and pancreatic duct were uninvolved. All resected margins were uninvolved with no lymphovascular invasion.

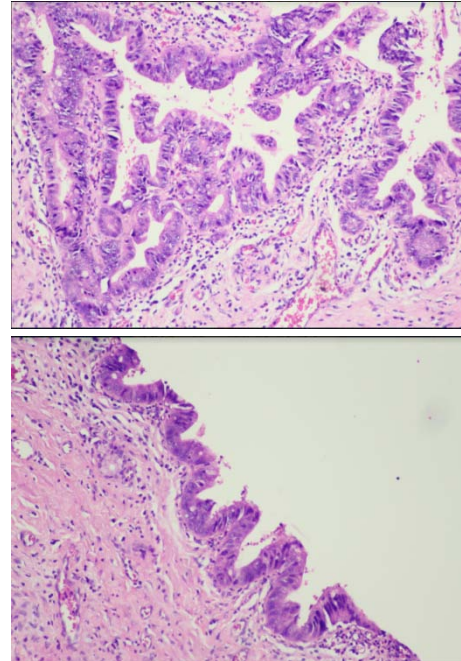


Fig. 4, 5: Marked Dysplasia of Common Bile Duct at X100

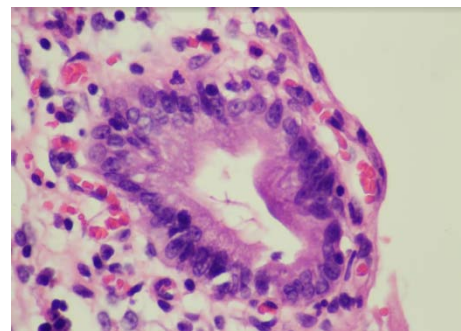


Fig. 6: Marked Dysplasia of Common Bile Duct at X400

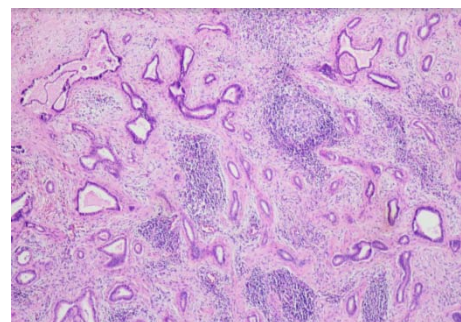


Fig. 7: Resected Lymph Node shows Component of Dysplastic Glands suggesting a Metastasis of Adenocarcinoma

His pathology report showed lymph node involvement, therefore, he was given Gemcitabine plus Cisplatin for postoperative systemic chemotherapy. Whole abdomen CT scan showed no evidence of tumor recurrence at 3 months post-operation. However, the follow up CT scan found a small liver nodule at segment 5, suggesting a metastasis.

III. DISCUSSION

We have described a case of isolated extrahepatic, comparatively rare, intraductal papillary neoplasm of the bile duct (IPNB). The patient underwent radical surgery and received adjuvant chemotherapy.

Benign IPNB is a precancerous condition of intraductal-growth and papillary type of the cholangiocarcinoma.⁸ It usually progresses slower and have better prognosis compared to the conventional cholangiocarcinoma.¹⁰ According to latest classification of IPNB based on preoperative imaging and the pathological findings of surgical specimens, there are 5 classification: Class I – classical intrahepatic IPNB; Class II - extrahepatic IPNB; Class III - cystic variant; Class IV - micro-papillary lesion; and, Class V - macroinvasion.¹¹

Previous studies report a more aggressive behavior in the extrahepatic type compared to the intrahepatic type.^{8,12,13} From Luvira et al with 103 IPNB case study, all the class II tumor (extrahepatic IPNB) are malignant IPNB and have the significant shorter survival period.¹¹

The patient in our case report was diagnosed with a class II IPNB. Although he was treated with radical surgery and adjuvant therapy, the metastasis and worsen condition could be expected.

IV. CONCLUSION

This case report showed a recurrence in intraductal papillary neoplasm of bile duct (IPNB) patient despite he had performed radical surgery and received postoperative chemotherapy. It demonstrated the poor prognosis of the more aggressive type of the bile duct neoplasm, a purely extrahepatic type.

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