

Intrahepatic intraductal papillary mucinous neoplasm (IPMN): Case report

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Abstract

Background: IPMN is a rare condition in which the bile ducts are dilated and filled with papillary or villous neoplastic epithelium. IPMNs usually are of pancreatic or extrahepatic origins. Intrahepatic variation of IPMNs is increasingly being reported [4], and a common feature in reported cases is bile duct dilatation.

Methods: 65-year-old man was being investigated for iron deficiency anaemia (IDA) and there was an incidental finding of a liver lesion on Computed Tomography (CT) scan. He was then referred to the Hepatobiliary (HPB) team for management. Following management, his case files including clinic letters, test results, radiological imaging, operative notes, records of admissions and histopathology reports were reviewed for this retrospective case report.

Results: At first presentation in the HPB clinic, he was asymptomatic with normal liver function tests and the plan was referral for Multidisciplinary team (MDT) discussion. The case was discussed, and decision was to manage as suspected early cholangiocarcinoma with liver resection. He was electively admitted and had a left lateral sectionectomy. He had an uneventful peri-operative period and discharged home on recovery. Histology reported the specimen as a cystic variant of IPMN arising in intrahepatic bile duct with no evidence of invasive cancer. After this, there was a further discussion at MDT and the plan was for discharge. This happened in follow up clinic, at which time patient remained asymptomatic and well overall.

Conclusion: Intrahepatic IPMNs are rare premalignant lesions. There are no specific symptoms, so diagnosis is often post liver resection.

Keywords: liver; IPMN; intrahepatic; bile duct

Introduction

Intraductal papillary mucinous neoplasm (IPMN) of bile ducts is a rare condition in which the bile ducts are dilated and filled with papillary or villous neoplastic epithelium. IPMNs were not recognised as a separate entity to ductal adenocarcinoma of the pancreas until a couple of decades ago [1]. These lesions are different from mucinous cystic neoplasms (MCNs) and their incidence is estimated at 2.04/100,000 population [2]. IPMNs usually are of pancreatic or extrahepatic origins and present with pancreatic duct obstruction symptoms like abdominal pain (most common presentation), obstructive jaundice and weight loss [3].

Over the last three decades, IPMN is increasingly being reported in radiological imaging and pathological specimens [4]. Reported cases of intrahepatic IPMNs have shown variable imaging results from duct dilatation, multilobular cysts to mass lesions. Cellular atypia was also variable in reported cases – mild to moderate atypia, high-grade atypia, and carcinomas. A reported case of synchronous adenocarcinoma and intrahepatic cholangiocarcinoma showed histologic evidence of underlying precursor lesions including IPMN. A common feature of intrahepatic IPMN in most reported cases remain bile duct dilatation.

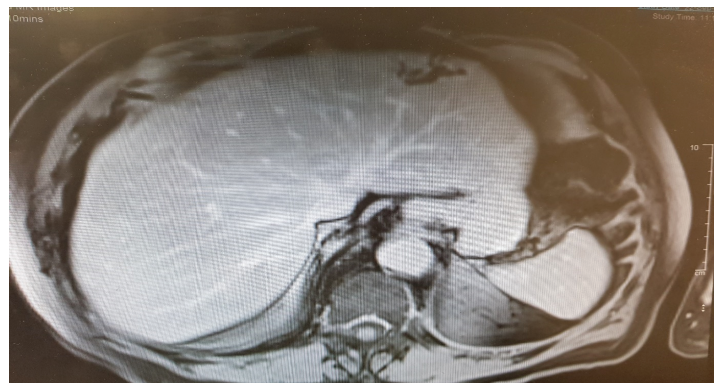


Figure 1: Radiological Image - MRI showing the liver with ductal dilatation in Segment 2

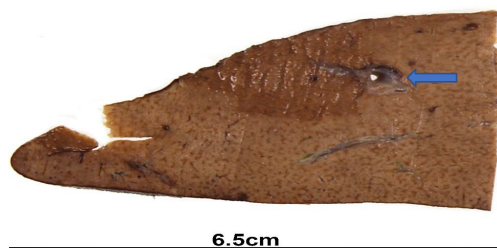


Figure 2: MACROSCOPIC IMAGE - Resected section of the gross liver specimen demonstrating focal cystic dilatation

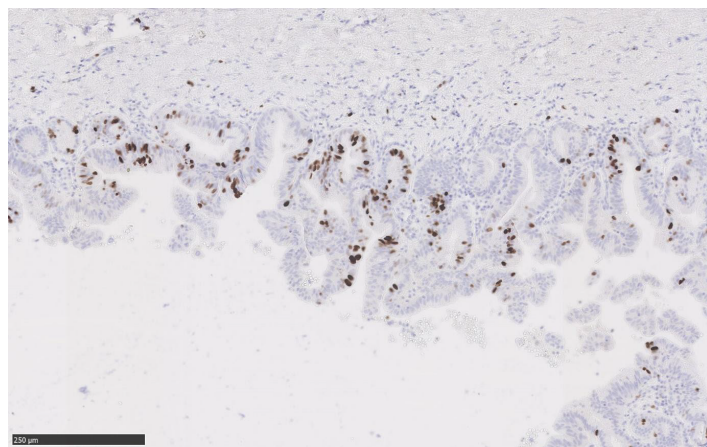
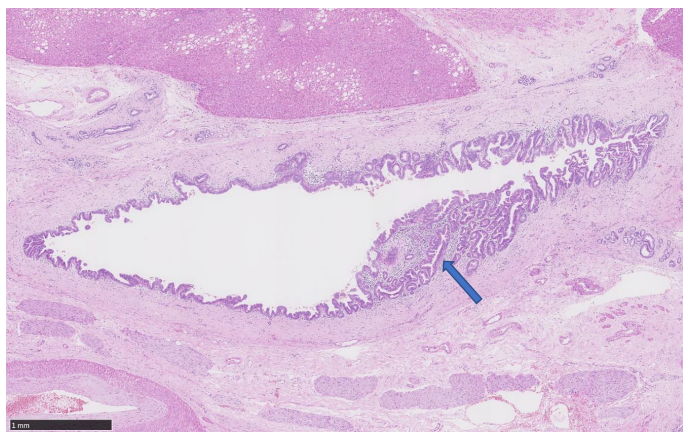


Figure 4: Immunohistochemistry – KI67 proliferative index 5%

3.1: Microscopy of the resected lesion showing papillary proliferation within the cyst



3.2: Papillary structures composed of complex architecture of atypical epithelial cells with eosinophilic cytoplasm, and hyperchromic nucleoli

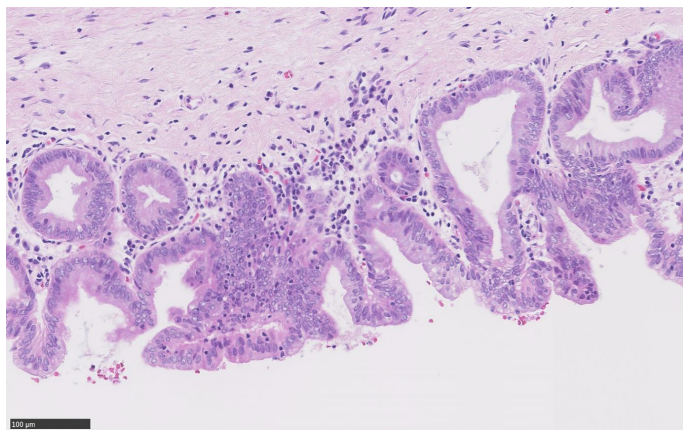


Figure 3: Histopathological Images

Case report

Presentation

A 67-year-old man with Type 2 Diabetes, COPD, Hypertension, and abdominal aortic aneurysm repair, was being investigated for iron deficiency anaemia (IDA) by the Gastroenterology team. As part of his workup, he had a CT scan which showed a liver lesion for which referral to Hepato-biliary (HPB) team was made. On referral to HPB, he was discussed in the MDT and he was diagnosed as a suspected early cholangiocarcinoma, and the decision for liver resection was made.

At the HPB outpatient clinic, an assessment was made, and investigations were reviewed. Options including surveillance and surgical resection were discussed. The risks and benefits of both options were discussed, with risk of mortality explained.

Management

He was electively admitted and had a left lateral sectionectomy. The operation and recovery were uneventful. Intra-operative findings include adhesions around previous midline incision and a fatty-looking liver, but no mass lesion palpated. Liver was mobilised and an on-table ultrasound of the liver showed no mass. Left liver parenchyma was transected, haemostasis achieved, and no bile seen.

He stayed in the High Dependency Unit (HDU) for 3 days and suffered no post-operative complications. He spent a week in total in hospital and was discharged with a plan for further discussion at MDT.

Histology

Macroscopically, tissue specimen measured 190 x 120 x 50mm, with noted dilated ducts on sectioning with no obvious solid lesion seen. On microscopy, multiple sections taken from the hepatic resection showed cystic lesion with intrahepatic papillary proliferation and mild nuclear atypia. There was marked cystic dilatation in many sections but surrounding soft tissue showed no evidence of ovarian-like stroma. There was mild focal chronic inflammation in some of the sections. These appearances were compatible with IPMN of intrahepatic duct, and the features mimic that of IPMN of pancreatic origin. The surrounding liver tissue showed mild macro and microvesicular steatosis but there is no evidence of inflammation or malignancy..

Discharge

A further MDT discussion took place and the plan was for no further treatment. At his follow-up clinic, he remained well and symptoms-free, and he was reassured and discharged.

Discussion

Intrahepatic IPMN is a rare condition and only a few cases have been reported in literature [5]. They present as dilated bile ducts filled with papillary or villous epithelium [6]. Intrahepatic IPMN arise from the stem cells of the bile ductules, lining biliary epithelium or epithelium of peribiliary glands [7]. Most of the intrahepatic IPMN tend to cause obliteration or stricture of the biliary ducts as they form nodules full of mucinous material, however sometimes they grow along the lining of the ducts [8]. They are usually found in association with chronic biliary disease like clonorchiasis or hepatolithiasis.

There are no specific symptoms can indicate intrahepatic IPMN, so the diagnosis is always after liver resection which usually performed to treat intrahepatic biliary dilatation of unknown cause [9]. However, most of the patients have reported having abdominal discomfort [10]. The main diagnostic tools remain CT and MRI scans. The main findings can include the presence of a solid mass, duct dilatation and increase in size on serial CT/MRIs. New modalities have shown better detection of these tumours and can differentiate better between benign and malignant IPMNs, those modalities are F-labelled fluorodeoxyglucose CT and PET scans [11].

On the premise that IPMN is considered a premalignant condition, surgical intervention should be offered to the patients. According to the guidelines, all the IPMNs should be resected unless the comorbidities and fitness of the patient is in question, then medical treatment of the symptoms is the second line of management [12]. Survival after resection of invasive carcinoma in association with intrahepatic IPMN is like that of the pancreatic cancer in association with IPMN, the median was 2.8 years and 5-year survival rate were 38% [10]. The indicator for survival is the depth of the invasion of the cancers associated with intrahepatic IPMN [13-15].

Conclusion

Intrahepatic IPMN are rare conditions that has tendency to turn into malignant lesions such as cholangiocarcinoma. The lesion will show on scans as intrahepatic dilated bile ducts with no other features. The definitive treatment of intrahepatic IPMN remains liver resection.

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