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Original Research Paper

General surgery

A RARE CASE OF MUCINOUS CYSTADENOCARCINOMA OF LIVER PRESENTING AS HYDATID CYST

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ABSTRACT Hepatobiliary cystadenocarcinoma is a very rare cystic tumor that arises in the liver or, less frequently, in the extrahepatic biliary system mostly seen in elderly patients. It has been shown to arise in congenital liver cysts, bile ducts, biliary cystadenoma, in the context of fibro- polycystic disease and in the hepatoduodenal ligament. In the present case report, we describe a case of rare entity of hepatobiliary cystadenocarcinoma presented in a 36 year old male patient presented to opd with complaints of pain abdomen in right side of upper abdomen since 2 months. Main concern of this case report is preoperative imaging is imprecise and frequently misdiagnosed as benign cystic lesions and delays in treatment.

KEYWORDS : Hepatobiliary cystadenocarcinoma.

INTRODUCTION - :

Biliary cystadenocarcinoma is a very rare cystic tumour and constitutes less than 5% of intrahepatic cysts of biliary origin(1). Most commonly presented in elderly patients.Seen in 5th-6th decades. Commonly presents With symptoms includes an abdominal mass, abdominal pain, nausea, jaundice,fever or occasional ascites. Some patients are asymptomatic, the lesion being an incidental finding. Diagnosis of suspicious liver cysts with preoperative imaging modalities is imprecise and inconclusive and frequently misdiagnosed. So patients presenting with similar symptoms considered as a benign cystic lesions and less invasive methods like drainage, marsupilisation and interval follow up are ineffective and may delay the treatment of malignant conditions.

Case Report –

A 36 year old male patient came to opd with complaints of pain in rt side of upper abdomen since 2 months, which is colicky in nature, non radiating, no aggravating factors. No h/o fever, jaundice, vomiting . Patient had previous history of Hydatid cyst of liver in segment 4 previously operated. Done deroofing of cyst and omentopexy with intracystic drain 6 months ago. No significant family history noted.

General Examination –

Patient is conscious coherent and oriented. Palor present, no icterus, clubbing, cyanosis, pedal oedema and generalised lymphadenopathy. On examination vitals are stable, patient was afebrile and no signs of dehydration present.

Per Abdomen Examination –

Soft, no local rise of temperature and mild tenderness present at right hypochondriac region and epigastrium. Lump with well defined margins and smooth surface appears present in the epigastric region which extends into the rt hypochondrium.

Investigations –

Usg Report –

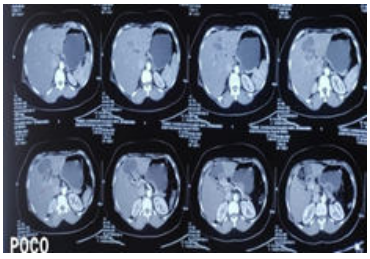
1. Cysts with biliary communication in the segment 4 of the liver likely recurrence.
2. Cyst in the distal body of pancreas ?IPMN? PSEUDO CYST.
3. Bilobar central IHBRD with wall thickening & few membranes with in.
4. Choledocholithiasis with cholelithiasis.

BIPHASIC CECT ABDOMEN REPORT–

1. Multilobular cystic lesion in segment 4b and 5 of liver with apparantly biliary communication ?cyst forming intraductal papillary neoplasm of biliay duct.
2. Unilocular pancreatic cystic lesion with MPD communication ? Main duct IPMN ? Other cystic pancreatic tumour.

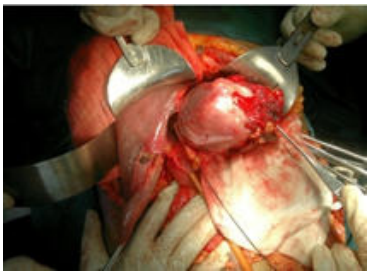
Mri Abdomen & Mrcp Report –

1. Cystic lesion in segment 4 of liver & at porta causing mass effect on biliary ducts ? Residual hydatid cysts.
2. Cystic pancreatic mass with communication or compression of MPD ? Cystic pancreatic neoplasm.



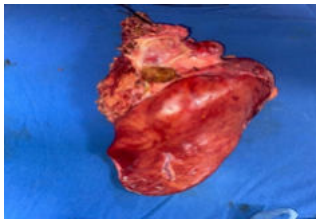
MANAGEMENT –

After preoperative evaluation, patient under general anaesthesia, left hepatectomy with distal pancreatectomy with gall bladder and hepatic duct resection with Roux-en-Y hepatico jejunostomy with jejuno jejunostomy was performed. Specimens were sent to histopathological examination, intra op bile for culture and sensitivity and pancreatic cystic fluid for CEA levels and amylase levels.



OPERATIVE FINDINGS –

1. Dense adhesions between liver surface and peritoneum and parietal layer of diaphragm.
2. Right lobe of liver hypertrophied.
3. Right hepatic artery was found densly adherent to CHD, running posterior to CHD.
4. 5*5 cm cystic mass involving segment 4 and 5.
5. 2*1 cm firm mass at proximal CHD and confluence.
6. 3*2 cm cystic mass at distal body of pancreas.

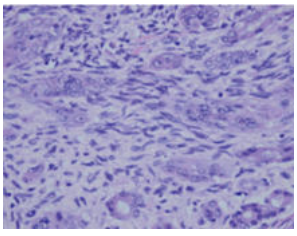


CUT SECTION OF SPECIMEN –

1. A 2*2 cm polypoidal mass with stalk arising from the wall of CHD.
2. Cystic dilatation of secondary biliary radicles draining in to the left hepatic duct & CHD.
3. Clear serous fluid aspirated from pancreatic body cystic mass.
4. Dilated MPD along with the parenchyma forming distal mass.

On Microscopic Examination :

Demonstrated a well differentiated adenocarcinoma arising with in a biliary cystadenoma with multifocal severe dysplasia. The malignant polyps contained a typical glands infiltrating into underlying mesenchymal stroma. A focus of metastatic disease was identified with in the pancreatic specimen as well.



After 1 year followup patient remained disease free with normal CT and ca19-9 level

CONCLUSIONS –

Biliary cystadenocarcinomas are tumors thought to arise from malignant transformation of biliary cystadenomas, but little is known about the risk or timing of malignant transformation. Cystadenocarcinomas arise from the intrahepatic bile duct and are composed of multiloculated mucin producing epithelial cells. The tumors should be carefully distinguished from distinct entities such as primary bile duct cancer (cholangiocarcinoma) with dilated intrahepatic ducts and carcinomas arising from simple hepatic cyst [1]. The criteria utilized to differentiate an atypical liver cyst from an adenoma/adenocarcinomas are nonspecific and include Multiloculated cyst with internal septations, thickened or irregular cyst walls, mural and papillary projections, calcifications, or enhancement. The combined sensitivity of CT, U/S, and FNA was 30% (true positives/true and false positives based upon imaging suspicion) [2]. Recent efforts have been made using modern computer tomography and contrast-enhanced ultrasonography to more accurately diagnose biliary cystic neoplasms [3,4]. The recommended management ofsuspicious liver cysts is complete surgical resection. Less invasive methods to include interval followup, drainage, or marsupialization are ineffective and may delay treatment of a malignant condition. There is imbalance between the incidence of benign liver cysts and incidence of cystadenoma/ carcinoma would make even the most sensitive radiographic test imprecise. At present, surgical resection should be recommended for all suspicious "biliary cystadenomas" but the clinician must understand the radiologic limitations.

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