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Case Report

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A Rare Case of Primary Cystic Duct Adenocarcinoma

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1. Summary

Primary cystic duct adenocarcinomas is rare amongst the extrahepatic bile duct malignancy. Moreso it has a very difficult preoperative diagnosis and is diagnosed intraoperatively or postoperatively. It has a better prognosis compared to other extrahepatic biliary tumours. We describe a case which was preoperatively diagnosed as carcinoma neck of gall bladder, intraoperatively showing growth at cystic duct-common hepatic duct junction and postoperatively showing primary cystic duct carcinoma.

2. Background

Primary carcinoma of cystic duct carcinoma has incidence 2-3.6% amongst all the extrahepatic biliary malignancies [1]. Less than 70 cases have been reported worldwide [1,2]. Most of these cases were reported from East Asia. Surgery with en bloc resection of gallbladder, cystic duct, common bile duct, and regional lymphadenectomy is the mainstay of treatment. Due to the stringent criteria of Farrar and its initial phase of this disease being like gallbladder disease, early diagnosis is not only difficult but dilemmatic. This paper describes a rare case report of primary cystic duct tumour adhering to the new criteria and the presentation as well as management.

3. Case Report

A 72-year-old gentleman presented to surgical gastroenterology outpatient with recurrent abdominal pain in the right upper quadrant since last 4 to 5 months. He also complained of yellowish discolouration of eyes 2 months back which was gradually progressing associated with fatigue, pruritus and darkening of urine. He was icteric on general examination and gallbladder was palpable on abdominal examination.

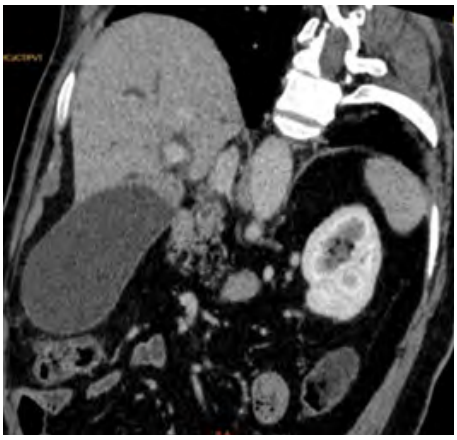

Figure 1: CECT abdomen Coronal view showing hydrops gall bladder with epicentre of growth in the cystic duct region

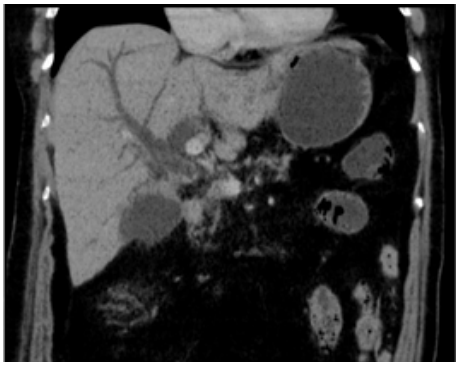

Figure 2: CECT abdomen coronal view showing the trifurcation of the hepatic ducts along with dilatation and growth in the cystic duct

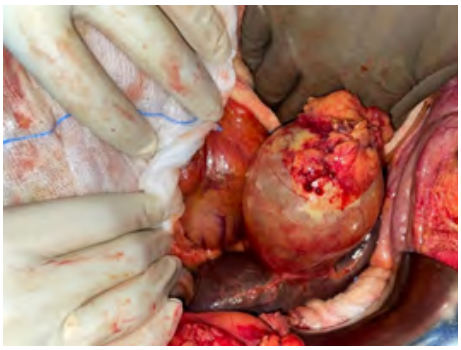

Figure 3: Shows the intraoperative photo of hydrops gall bladder commonly seen in cystic duct tumours

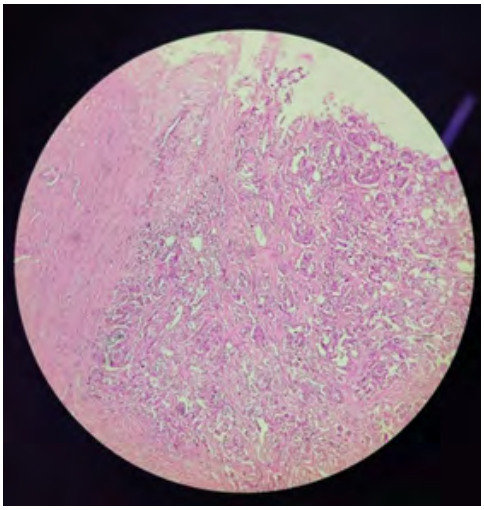

Figure 4: Microscopic view, moderately differentiated adenocarcinoma with perineural invasion

4. Investigations

USG abdomen was suggestive of distended gall bladder with mass obstruction at the neck and cystic duct region of the gall bladder with intrahepatic biliary radicles dilatation and no ascites or liver lesions. Contrast enhanced tomography showed malignant lesion involving the neck of the gall bladder extending to cystic duct- common hepatic duct junction with upstream dilatation of intrahepatic biliary radicles with periportal lymph nodes and absence of ascites and liver lesions. Blood investigations showed normal haemoglobin with marginally elevated total leucocyte counts- 12200 cells/cumm. Liver function tests showed total bilirubin- 30.19mg/dl with SGOT/SGPT- 343u/l, 362u/l and alkaline phosphatase 278u/l. PET-CT was done to rule out distant metastasis. After prehabilitation patient was planned for open procedure in view of malignancy, Radical cholecystectomy with bilio-enteric bypass (Hepaticojejunostomy). Intraoperatively gall bladder was grossly distended with epicenter of the tumour was at the cystic duct- common hepatic duct junction and no secondaries to liver. Patient underwent radical cholecystectomy with common bile duct excision and Roux-en-Y hepaticojejunostomy with regional lymphadenectomy achieving a macroscopically negative margin.

The histopathology of the specimen showed a growth of 2.5x1.5cm with its epicentre at the cystic duct extending to the cystic duct- common bile duct junction with perimuscular and adjacent tissue serosal involvement, with extensive perineural and no lymphovascular invasion and gall bladder showing features of chronic cholecystitis, moderately differentiated adenocarcinoma, pT2bN0.

5. Outcome

Post operative period was uneventful with gradual waning of the jaundice. Post op chemotherapy has been advised.

6. Discussion

Primary carcinoma of cystic duct is extremely rare [5, 6]. Incidence of primary cystic duct carcinoma in autopsy studies was found to be 0.03-0.0.5% [7] hence very little research material is available. Males are affected more than females and average age is 65 years (8, 9,10). It usually has a vague presentation and main symptom is abdominal pain and jaundice [11, 12]. Diagnosis is made either intraoperatively or post operatively [11-13]. Resectability of cystic duct carcinoma is decided using CDC (cystic duct carcinoma) classification systems.

The oldest was that of Farrar's which was as follows; (i) Growth restricted to the cystic duct (ii) Absence of neoplastic process in the GB, hepatic, or CBD (iii) Histological evidence of carcinoma (1). However cystic duct being a short muscular structure, as it primary tumour advances it becomes a part of surrounding structure that is common bile duct or common hepatic duct (1,2). Since the definition was too restrictive, several other classifications like ozden et al, kim et al were also described to overcome its shortcomings [2, 3].

Kim's classification is as follows (i) Type I-carcinoma confined within the cystic duct(ii) Type II-carcinoma extended to the GB neck and infundibulum or bile duct of cystic duct side without obstructive jaundice(iii) Type III-carcinoma extended up to the GB body or bile duct on the contralateral side of cystic duct opening which then causes obstructive jaundice (centre located in the cystic duct) [3].

Yokoyama says A gallbladder tumor with centre of which is located in the cystic duct:(i) hepatic hilum type (HH)-tumor mainly invades thehepatic hilum (ii) cystic confluence type (CC)-tumor mainly involvesthe confluence of the cystic duct [4].

Nakata describes the cystic duct tumours based on its extent of spread; Type I-the tumor was located wholly within the cystic duct Type II-the tumor extended to the gallbladder. Type III-the tumor extended to the common hepaticduct or the common bile duct (including extension into the lumen and external invasion to the bile duct wall). Type IV-the tumor extended to both the gallbladder and the bile duct [5]. Currently, classifications of kim, Nakata and Yokoyama are widely used [4-6].

Our tumour according to the above classifications is type 3 kim, type 2 yokoyama and type 3 nakata [3-5]. Most common tumour associated with cystic duct is adenocarcinoma, however, small cell carcinomas, carcinoid tumour and mucin producing tumours have also been reported [3,9,13]. Perineural invasion is the main prognostic

factor and lymphatic invasion is less compared to cholangiocarcinoma or gall baldder cancer [2,4,15]. The recommended treatment is radical surgery which consists of cholecystectomy with non-anatomical gall-bladder fossa resection and excision of extrahepatic bileduct with regional lymphadenectomy and bilio-enteric bypass. [3,5,9,10,11] The average survival was 27.2 months while that of gall-bladder carcinoma was only 5.8 months and of other extrahepatic biliary ducts 3.2-11.4 months [3, 15].

7. Conclusion

Primary cystic duct carcinoma is a rare entity which is usually misdiagnosed as gall bladder cancer. Its diagnosis is usually made intraoperatively or post operatively. We describe here one such case of a 72-year-old gentleman, initially diagnosed as carcinoma neck of gall bladder. Intraoperatively the growth was at the epicentre of cystic duct extending to cystic duct- common hepatic duct junction. He underwent cholecystectomy with bile duct excision and Roux-en-Y hepaticojejunostomy with regional lymphadenectomy. Histopathology showed primary cystic duct tumour; moderately differentiated adenocarcinoma with perineural invasion and no lymphovascular invasion. According to the above classifications the tumour described here is type 3 kim, type 2 Yokoyama and type 3 Nakata [3-5].

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