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A Case of Type IV Hilar Cholangiocarcinoma

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Abstract: ***Cholangiocarcinoma is an aggressive malignancy of biliary epithelium that may arise anywhere in the biliary tract, from the intrahepatic biliary canaliculi to the terminus where the common bile duct enters the duodenum at the duodenal ampulla. Cholangiocarcinoma's comprise about 3% of gastrointestinal malignancies, are the second most common primary liver tumours, and account for approximately 10% to 15% of all hepatobiliary malignancies. The incidence of intrahepatic cholangiocarcinoma has been rising, possibly due to improved diagnostic and classification techniques, while the incidence of extrahepatic lesions has been falling in recent years Cholangiocarcinoma is classified by anatomical origin as intrahepatic cholangiocarcinoma or extrahepatic cholangiocarcinoma, which is subdivided into perihilar cholangiocarcinoma and distal cholangiocarcinoma. More than 95% of cholangiocarcinoma's are adenocarcinomas. The radiologic evaluation includes a right upper quadrant ultrasound examination, which may show intrahepatic biliary ductal dilation but does not usually identify the actual site of obstruction. With hilar cholangiocarcinoma's, the gallbladder and visualized extrahepatic biliary tree are usually decompressed, whereas distal lesions will have extrahepatic biliary ductal dilation and gallbladder distention. Typically, computed tomography alone is insufficient for the assessment of appropriateness of resection. Other imaging methods include percutaneous transhepatic cholangiography, ERCP and MRCP. While surgery is the only cure, radiation, chemotherapy, and targeted therapy can also be used in conjunction with surgery. All patients with suspected or confirmed cholangiocarcinoma should be evaluated for distant metastasis; almost 75% of patients have nonresectable or metastatic disease at presentation.***

Keywords: Cholangiocarcinoma, malignancy, ultrasound, computed tomography, percutaneous transhepatic cholangiography, metastasis

1. Introduction

Cholangiocarcinoma is a rare disease entity that carries a relatively poor prognosis. The incidence of cholangiocarcinoma is rising worldwide and is now the second most common primary cancer of the liver behind hepatocellular carcinoma. Tumours involving perihilar and intrahepatic lesions are known as proximal lesions, while those involving the periampullary region are known as distal disease. More than two thirds of all cholangiocarcinoma's involve the proximal biliary tree near the bifurcation, known as a Klatskin tumour. These tumours typically harbour mutations of *K - RAS*, *BRAF*, *TP53, HER2, PTEN, BAP1* and more. Precursor lesions for cholangiocarcinoma include high - grade dysplasia and intraductal papillary mucinous tumour of the bile duct (IPNB) - which encompasses intraductal papillary cholangiocarcinoma and precursor lesions such as biliary papillomatosis and biliary intraductal papillary mucinous neoplasm.

Risk factors for development of cholangiocarcinoma include congenital lesions, such as choledochal cysts, infection with the liver flukes Clonorchis sinensis and Opisthorchis viverrini (common in Southeast Asia), recurrent pyogenic cholangitis associated with bile duct stone formation and primary sclerosing cholangitis (most common risk factor in the West).

The three distinct pathologic subtypes include sclerosing, nodular, and papillary cholangiocarcinoma. Sclerosing cholangiocarcinoma tends to occur in the proximal bile, while distal cholangiocarcinoma's are associated with nodular and papillary subtypes. The two pathologic factors most influencing prognosis after resection are complete (R0) resection to negative margins and absence of lymph node metastases.

Obstructive jaundice is the most common symptom, associated with early satiety and loss of weight. However, constant pain upon presentation suggests more advanced disease. Manifestations of direct hyperbilirubinemia are evident, such as pruritus, dark urine, and steatorrhea. Cholangiocarcinoma tends to extend in a submucosal route, with associated perineural invasion. A palpable gall bladder is present if the obstruction is in the distal common bile duct (Courvoisier's sign).

Biochemical investigations will confirm the presence of obstructive jaundice (elevated bilirubin, alkaline phosphatase and gamma - glutamyl transferase). The tumour markers CEA and CA19 - 9 may also be elevated, and may be useful postoperatively in the surveillance of recurrence.

Cross - sectional imaging by triphasic CT allows not only assessment of metastatic disease but also evaluation of resectability. The location of the tumour can be identified, and its relationship to vascular structures also can be assessed. Cholangiography by MRCP, PTC, or ERCP helps determine the proximal extent of resection. Endoscopic cholangiography carries the additional risk of cholangitis by the introduction of enteric bacteria into an undrained portion of the biliary tree

The only curative treatment option for cholangiocarcinoma is surgical extirpation, and 5 - year survival rates of 63% have been reported after R0 resection. locoregional therapies represent an alternative treatment option, as does Trans - arterial chemoembolization (TACE). Options for restoration of biliary drainage include endoscopic, percutaneous, and surgical techniques. Endoscopic and percutaneous methods are based on placement of biliary stents, whereas surgical approaches create a bypass via a cholangiojejunostomy.

External beam radiation and intraoperative or intraductal brachytherapy have been suggested for palliative treatment.

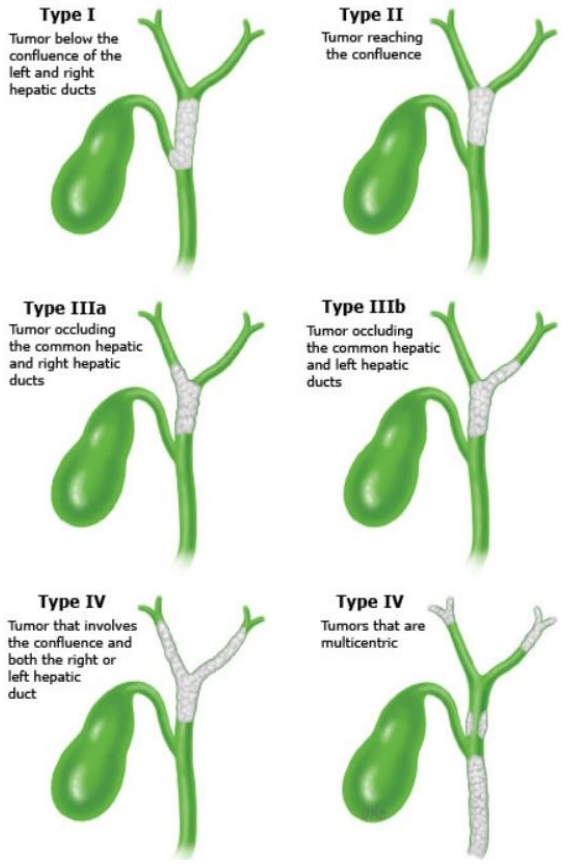

Figure 1: Bismuth - Corlette Classification of Cholangiocarcinoma

2. Case Details

Chief Complaints and History of Presenting Illness:
This case report pertains to a 67 - year - old female, a known case of bronchial asthma on regular medication, who was admitted with chief complaints of yellowish discolouration of the eyes for the past two months, associated with dark urine, pale - coloured stools, pruritus and intermittent abdominal pain in the right upper quadrant, with no specific aggravating and relieving factors. There were no other significant complaints. The patient had undergone postpartum sterilisation 25 years ago and laparoscopic appendicectomy 10 years ago. She had normal bowel, bladder and sleep habits, and had negative history of consumption of alcohol, tobacco or other banned substances. She had 2 healthy children born through elective caesarean section.

General and Systemic Examination:
The patient was conscious, oriented, afebrile and adequately hydrated. She was moderately built and well nourished. Her vital signs were normal, with blood pressure coming under the pre - hypertension range (130/70 mm Hg). General examination revealed presence of icterus. Examination of the abdomen revealed presence of palpable non - tender gallbladder. There were visible scars of incisions from previous surgeries. No abnormal findings were found in the examination of the other systems.

Investigations Done:
Routine investigations were carried out, including complete blood count, liver function test, renal function test and coagulogram, which revealed mild anaemia (Hb = 10.5g/dl) and decreased RBC count (3.28 million/ml), conjugated hyperbilirubinemia (12.7mg/dl), elevated AST (50 IU/L), elevated ALT (27 IU/L) and increased prothrombin time (18.8 seconds).

Tumour markers were tested due to suspicion of malignancy, which were found to be elevated. The serum level of CA 19 - 9 was 39 U/ml, while that of CEA (carcinoembryonic antigen) was found to be 5.5 ng/ml. Oesophago - gastro - duodenoscopy (OGD) was performed, upon which no abnormalities were found.

Radiological Findings:

CT Abdomen:
Evidence of relatively well - defined circumferentially enhancing soft tissue thickening noted involving common hepatic duct for a length of 2.8cm was present, extending superiorly up to hilum and into both hepatic ducts (right hepatic duct = 1.3 cm, left hepatic duct = 1.2 cm), causing bilateral intrahepatic biliary radical dilatation, and extending inferiorly down to the insertion site of cystic duct.

Gall bladder, cystic duct and common bile duct appeared to be collapsed. Subcentimetric periportal lymph node was noted. No evidence of peripancreatic and paraaortic lymph nodes enlargement was noted.

CT Abdominal Angiography:
Abdominal angiography revealed focal ectasia of suprarenal abdominal aorta for a length of 4.3cm with maximum luminal diameter of 2.7cm. The descending thoracic aorta measured 2.2cm, and the infrarenal abdominal aorta measured 1.7cm. Severe narrowing of short segment of proximal celiac trunk, 3mm from its origin, for a length of 8mm causing complete occlusion with distal reformation through collaterals was noted. Evidence of narrowing of short segment in ostium of right renal artery causing 20 - 30% luminal narrowing with normal flow in right renal artery was observed. Superior and inferior pancreaticoduodenal arteries and gastroduodenal artery appeared prominent. Diffuse circumferential mixed plaque in supra - renal and infra - renal abdominal aorta causing 10 - 20% luminal narrowing was present.

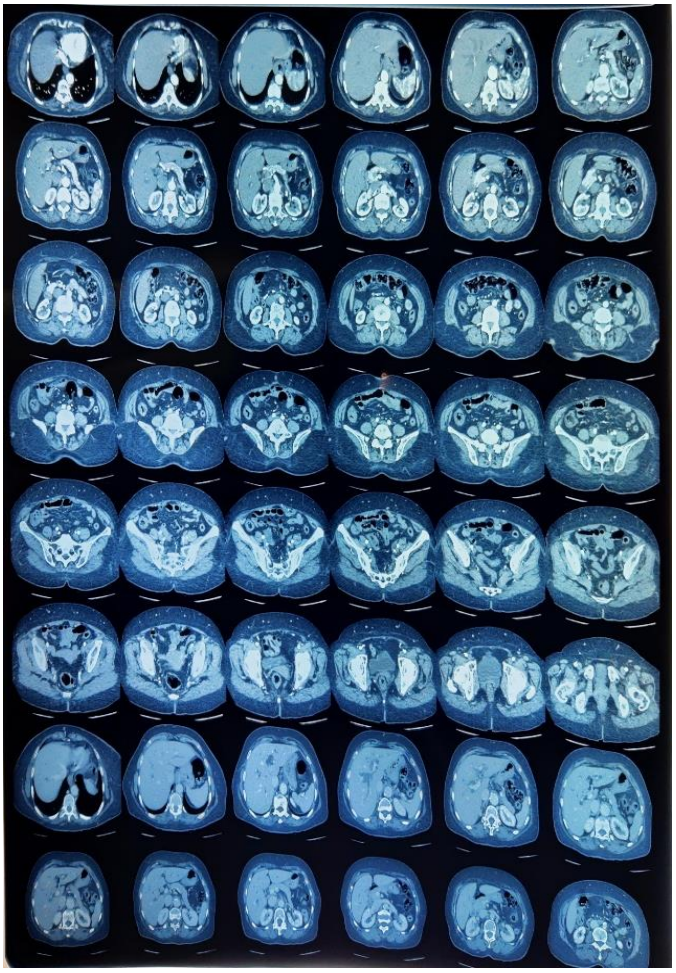

Figure 2: CT Abdomen

MRCP (Magnetic Resonance Cholangiopancreatography):
An ill - defined T2 hypointense lesion measuring 2.3 x 1.9 cm was noted at the confluence of right and left hepatic ducts, which showed patchy areas of diffusion restriction with low ADC (apparent diffusion coefficient) value, causing bilateral asymmetric dilatation of the intra - hepatic biliary radicles. No free fluid was present. No obvious abdominal lymph node enlargement was seen. The gallbladder was normal in size, with normal wall thickness. No peri - cholecystic fluid was present. The cystic duct appeared to be normal. The liver, pancreas and spleen appeared to be normal.

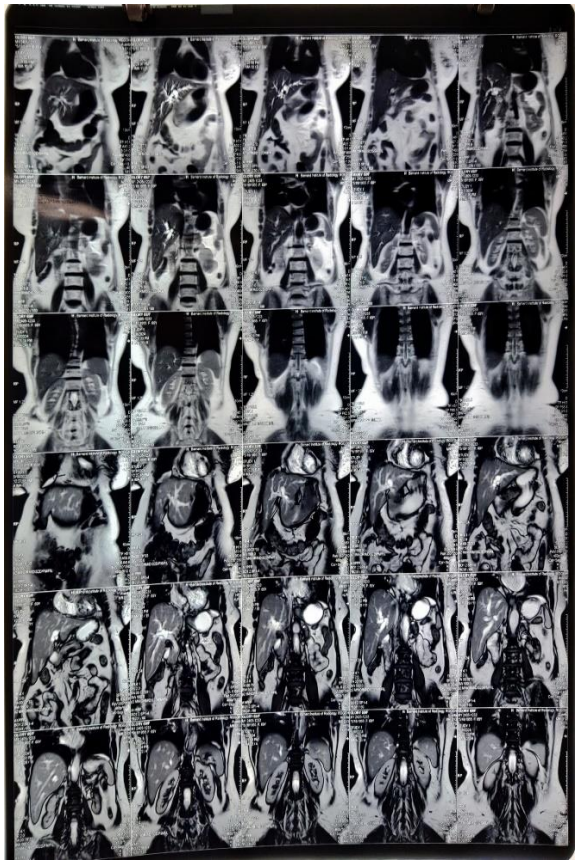

Figure 3: MRI Abdomen

Surgical Removal of the Tumour:
The patient underwent parenchymal preserving hepatectomy (caudate lobe and segment IVb), under d - laparoscopic setting, along with extra - hepatic bile duct excision and Roux - en - Y cholangiojejunostomy (RCJS). The cholangiojejunostomy (biliary enteric anastomosis) was performed under general anaesthesia and SAP prophylaxis, with the patient in supine position. It was performed under the end - to - side technique, where a cholecystectomy with cystic duct ligation was performed, followed by the ligation of the extra - hepatic bile duct at the level of the planned resection. The common bile duct was opened at the level of the planned anastomosis, remainder of the duct was resected, with the distal stump oversewn using 3 - 0 absorbable sutures. Roux - en - Y limb of jejunum was passed retro - colic and to the right of the middle colic vessels and was positioned to lie adjacent to the proximal healthy common bile duct in a tension - free manner. The posterior wall of the duct was sutured to the jejunum using running 3 - 0 absorbable sutures. The tail of the suture and needle were left intact. A jejunotomy was formed along the duct, and single interrupted 3 - 0 absorbable sutures were used to approximate the jejunal mucosa to the duct mucosa, with the knots tied on the inside of the lumen. The anterior portion of the anastomosis was then completed using the running sutures used to construct the posterior part. Finally, sponge was placed circumferentially around the newly created anastomosis to ensure the absence of a significant leak, and the wound was closed in layers after complete haemostasis.

Intra - Operative Findings:
A hard growth was noted at the hilar confluence of the right and left hepatic ducts. There was no arterial and venous

involvement. There was no ascites nor presence of liver metastasis.

Pathologic TNM Stage: Stage II (T2aN0M0)

Impression: Type IV Cholangiocarcinoma of the Intraductal - Growing type

3. Conclusion

Surgical treatment is considered the only truly effective treatment, based on the principle of margin - negative hepatectomy (R0 resection), with preservation of a postoperative future remnant liver of adequate size and function and regional lymph node dissection with the need for resection and reconstruction of the portal vein and/or hepatic artery, and with biliary reconstruction usually performed by Roux - en - Y choledojejunostomy. Three - segment resection, vascular resection and hepatopancreatoduodenectomy may also be needed. The 5 - year overall survival rate after surgery is around 63%. It is recommended that patients with lymph node metastasis undergo standard hepatoduodenal lymph node dissection in the surgical treatment of intraductal cholangiocarcinoma, but it is associated with a significant increase in postoperative complications. Liver transplantation is also an alternative surgical treatment. Suitable candidates for liver transplantation include: the presence of unresectable tumours with a radial diameter of <3 cm, and the absence of intrahepatic or extrahepatic metastatic disease. Pre - operatively, any pre - existing co - morbidities should undergo medical evaluation in anticipation of major surgery. Also, advanced discussion with the anaesthesiologist is often helpful to anticipate potential intraoperative needs (e. g., blood products) and the appropriate level of venous access and hemodynamic monitoring.

References

[1] Rizvi S, Gores GJ. Pathogenesis, diagnosis, and management of cholangiocarcinoma. Gastroenterology.2013; 145: 1215–29. doi: 10.1053/j. gastro.2013.10.013.
[2] Amin MB, Greene FL, Edge SB, Compton CC, Gershenwald JE, Brookland RK. et al. The Eighth Edition AJCC Cancer Staging Manual: Continuing to build a bridge from a population - based to a more "personalized" approach to cancer staging. CA Cancer J Clin.2017; 67: 93–9. doi: 10.3322/caac.21388.
[3] Razumilava N, Gores GJ. Combination of gemcitabine and cisplatin for biliary tract cancer: a platform to build on. J Hepatol.2011; 54: 577–8. doi: 10.1016/j. jhep.2010.10.010.
[4] Saha SK, Zhu AX, Fuchs CS, Brooks GA. Forty - Year Trends in Cholangiocarcinoma Incidence in the U. S.: Intrahepatic Disease on the Rise. Oncologist.2016; 21: 594–9. doi: 10.1634/theoncologist.2015 - 0446.
[5] Razumilava N, Gores GJ. Cholangiocarcinoma. Lancet.2014; 383: 2168–79. doi: 10.1016/S0140 - 6736 (13) 61903 - 0.
[6] Menon G, Garikipati SC, Roy P. Cholangiocarcinoma. [Updated 2024 May 6]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan