

Right extended hepatectomy for hilar cholangiocarcinoma: A case report

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Abstract

Radical resection in a case of hilar cholangiocarcinoma is the only curative option. However resection in a hilar cholangiocarcinoma is a challenging procedure because of the low resectability rate. Only a few cases of hilar cholangiocarcinoma are operable because of the advanced nature of disease at presentation. Furthermore, the extent of surgery makes it a complicated process to attempt. We recently had a patient who underwent an open extended right hepatectomy and hepaticojejunostomy for a type IIIa hilar cholangiocarcinoma.

The tumor was 20 mm in diameter and was located between the right hepatic duct and common hepatic duct. Radiological examination showed that the hepatic artery was not involved but the right portal vein was invaded by the tumor. CT volumetry was done and the future liver remnant was only 20% in the jaundiced patient. Preoperative drainage was done with percutaneous transhepatic biliary drainage from the left side. Portal vein embolization was done to augment future liver remnant to 30%.

The patient underwent an extended right hepatectomy (right trisectionectomy combined with caudate lobectomy). The operation time was nearly 300 min, and the intraoperative blood loss was about 500 ml. However, in the postoperative period, the patient developed post hepatic liver failure which was managed successfully with conservative treatment. The postoperative hospital stay was 23 days.

The final diagnosis was hilar cholangiocarcinoma with no nodal metastasis (pT2bN0M0) stage II (American Joint Committee on Cancer, AJCC).

Key words: Extended hepatectomy; Hilar cholangiocarcinoma; Portal vein embolization; Surgical technique.

INTRODUCTION

Cholangiocarcinoma is a rare disease. Of these, the majority are hilar cholangiocarcinoma (hCCA) or

peri-hilar cholangiocarcinoma (pCCA) ranging from 60–70%¹. Complete surgical resection (R0 resection) is the only curative treatment, similar to most other malignant tumors. However, only a few cases of hilar cholangiocarcinoma are operable because of the advanced nature of the disease at presentation. Among the hilar cholangiocarcinoma, 20-50% of the cases are unresectable. Only a few (30-70%) among the diagnosed cases of hilar cholangiocarcinoma can be resected, with improving resection rates and with more experience^{2,3}. These tumors are difficult to treat because of their tumor location. The close proximity to the major hepatic vessels makes it cumbersome and at times very difficult to achieve free distal margin with R0 resections. The proximity to the liver parenchyma in addition to the involvement of hepatic vessels often warrants the complex hepatic resections along with bile duct excision. The cholangiocarcinoma is also associated with high desmoplastic reactions and has a rich tumor microenvironment, all these characteristics contribute to its therapeutic resistance⁴. Preoperative optimization

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is required and liver volume augmentation is necessary to achieve curative resection in those cases where the future liver remnant (FLR) volume is insufficient.

We present a case of hCCA Type IIIa in which the tumor involves the biliary confluence and extends towards the right hepatic duct. The patient had to undergo Portal Vein Embolization (PVE) due to insufficient FLR. After 3 weeks of PVE, curative (R0) resection was finally possible.

CASE REPORT

The patient is a 67-year-old male who presented with complaints of anorexia for nine months and yellowish discoloration of his body for one month. He was evaluated for the symptoms which turned out to be Type IIIa hilar cholangiocarcinoma. The diagnosis was made with contrast-enhanced CT scan of abdomen and pelvis. The tumor was 20 mm in diameter and located between the right hepatic duct and common hepatic duct. Radiological examination showed that the hepatic artery was not involved but the right portal vein was invaded by the tumor. Volumetric analysis was done and the FLR was calculated to be 20%. Preoperative drainage was done with PTBD from the left side by the Interventional Radiology (IR) team. The initial CT scan showed a total liver volume (TLV) to be 1546cc making FLR with caudate lobe: $331/1546 = 21.41\%$ and FLR without caudate lobe: $310/1546 = 20.05\%$.

The patient was planned for PVE as his FLR was inadequate. After PVE, repeat CT scan, was done after 3 weeks which showed a TLV of 1267cc, segment II and III of 383 cc with a FLR of 30%. The kinetic growth rate was calculated using the formula for kinetic growth calculation to be $10\%/3\text{weeks}$, i.e. 3.3% per week ($>2\%$ per week) which was satisfactory⁵.

In this case the issue of inadequate FLR (20%) was tackled with PVE, which improved FLR to 30% in 3 weeks.

Staging laparoscopy was done initially. Intraoperatively, there was no ascites, no peritoneal or other deposits suggestive of metastasis. With these findings, we proceeded with the surgery and were able to remove the tumor. During the procedure, modified Makuuchi incision was given, liver was mobilized, right hepatic vein was mobilized, right portal vein was clamped and parenchymal transection was done. There was an intraoperative blood loss of 500ml and the duration of surgery was 300 minutes.

In the postoperative period, the patient developed Grade B Post Hepatic Liver Failure (PHLF) which was

managed conservatively with albumin and Fresh Frozen Plasma (FFP). His bilirubin level elevated more than 18 times the normal range, with reduction in his platelets count upto 42000 per microliter of blood. He started to improve only after the 10th postoperative day indicating the slow recovery from PHLF.

The right subhepatic drain was draining more than 500 ml of ascitic fluid till 14th postoperative day, which was also managed conservatively with IV fluids, oral fluids (with ORS) and enteral nutrition. The amount of drain then decreased slowly over a period of time.

Histopathological examination showed the tumor to be a well differentiated cholangiocarcinoma, biliary type in the perihilar duct. There was no lymphovascular invasion. The resection margins were free of tumor. Hilar vessels, liver, and lymph nodes were also free of tumor (PT2aN0). However, perineural invasion was seen.

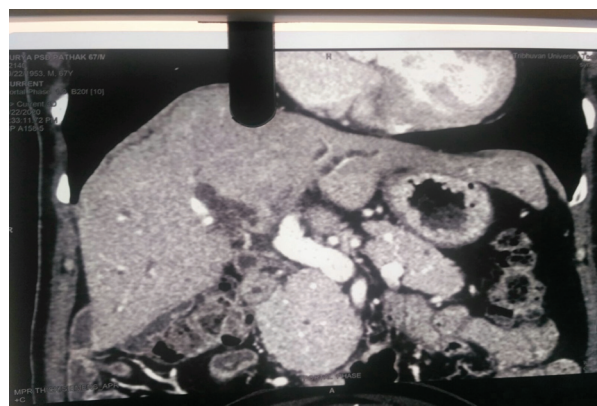


Figure 1: Preoperative CECT showing occlusion of right portal vein



Figure 2: Cholangiogram via the left duct showing the non-visualization of right duct

He was on regular follow up and on his 50th day of follow-up at the OPD, he was able to ambulate with support and take a normal diet. His wound had healed and the drain showed 1300ml of serous fluid which was gradually decreasing with time. His bilirubin level and platelets count were also in the normal range.



Figure 3: Final resected specimen

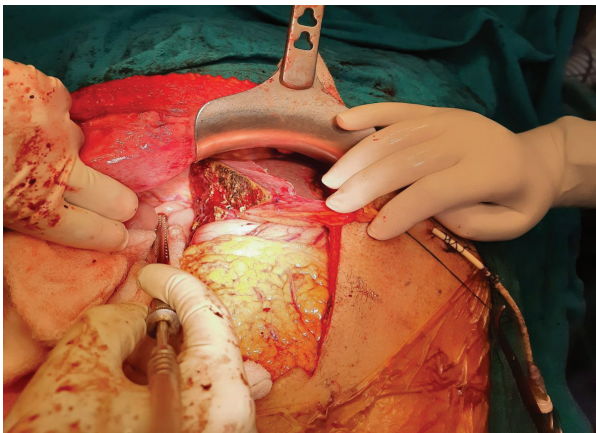


Figure 4: Resected margin of liver

DISCUSSION

Cholangiocarcinoma (CCA) is a group of rare aggressive intra and extra hepatic malignancy of the bile duct. They often present with locally advanced or metastatic disease⁶. The 7th and the 8th edition of the American Joint Committee on Cancer (AJCC) staging system, have now separated extrahepatic cholangiocarcinoma into perihilar (proximal) and distal cholangiocarcinoma based on the differences in their biological behavior, management and the prognosis.⁷. Therefore, there are three anatomical types of CCA: intrahepatic cholangiocarcinoma (iCCA), perihilar cholangiocarcinoma (pCCA) and distal

cholangiocarcinoma (dCCA)⁶.

Peri-hilar cholangiocarcinoma (pCCA) or hilar cholangiocarcinoma (hCCA) is the most common among the CCA, ranging from 60–70%¹. pCCA are those cholangiocarcinoma that involve or are near the biliary confluence of the right and left hepatic duct and are located proximally to the area between the secondary branch of bile ducts and distal to the insertion of the cystic duct into the common bile duct⁴. Proximal and distal are explained according to the direction of bile flow. The patient had a tumor in the hilum that was extending to the right hepatic duct. So this was a type IIIa hilar cholangiocarcinoma.

These patients usually present with symptoms of jaundice, weight loss and abdominal discomfort. Radiological diagnosis is done mainly by contrast-enhanced computed tomography (CE-CT) and contrast-enhanced magnetic resonance imaging (CE-MRI). We diagnosed the case with the help of clinical symptoms and CE-CT. We did a cholangiogram via PTBD catheter to delineate the biliary anatomy and intrabiliary tumor extent.

Radical surgery provides the only curative option for hCCA, with a 5-year survival rate ranging from 9 to 41%⁸. Surgical treatment is thus the preferred option, with appropriate consideration for the involvement of the vascular structures and lymph nodes⁹.

However, the resectability rate of these tumor is low as they usually have a silent evolution and the patients present late. The low resectability of hilar cholangiocarcinoma is thus due to the advance nature of the disease at presentation. The deep tumor location, close proximity to the major hepatic vessels makes it cumbersome and at times very difficult to achieve free distal margin with R0 resections. Cholangiocarcinoma is associated with its high desmoplastic reactions and has a rich tumor microenvironment, all these tumor characteristics contribute to its therapeutic resistance⁴. The proximity to the liver parenchyma in addition to the involvement of hepatic vessels often warrants the complex hepatic resections along with bile duct excision. Therefore, extended resections, including right trisegmentectomies, result in the highest rate of R0 resection¹⁰. Right trisectionectomy was done in our case to achieve the R0 resection. Mobilization could be done without much difficulty. Liver transection was done with the help of harmonics and crush-clamp technique. Due to desmoplastic reaction, hilar dissection was difficult.

However we could achieve the left proximal margin and anastomose the left duct with a roux-en-y limb of jejunum.

In cases of liver resections, apart from R0 resections, we must make sure that the future liver remnant (FLR) volume is adequate. Due to inadequate FLR, many patients require liver augmentation technique like PVE by means of which resectability can be improved. At the same time, major hepatectomies in these cases are still associated with high rates of morbidity and mortality, nearly being 40% and 10% respectively despite improvement in the surgical technique and postoperative care⁹. Due to all these reasons, excision of hilar cholangiocarcinomas requires a HepatoPancreatoBiliary (HPB) surgeon trained in the respective field to carry out the surgery. Preoperative accurate staging which is crucial in clinical decision-making helped us in choosing the surgery required to achieve the R0 resection. After liver volumes calculation, PTBD and PVE was done with the help of IR team available at our centre and resection of the hCCA could finally be done in our patient with good results.

The patient developed PHLF in the postoperative period. PHLF is a possible complication of major hepatic resections which can occur in up to 10 % of the patients who undergo major liver resections¹¹. As the patient could be managed with fresh frozen plasma and albumin without any invasive treatment, it was a Grade B PHLF.

Although complex, extended hepatectomies can be done safely in hilar cholangiocarcinoma with good outcome by trained surgeons with the help of a multidisciplinary team.

CONCLUSION

Resection rate for hilar cholangiocarcinoma is low because of the advanced nature of the disease at presentation. Resectability may be improved by volume augmentation as with PVE. The resection of hilar cholangiocarcinoma is a complex surgery but holistic care by trained surgeons and team can produce good results.

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