

Original Research Article

Extrahepatic Cholangiocarcinoma Diagnosis and Therapeutic Management

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Abstract: **Introduction:** Extrahepatic cholangiocarcinoma is a rare and severe cancer, representing less than 3% of malignant digestive neoplasms and is the second most common primary liver cancer after hepatocellular carcinoma (HCC). Its frequency is often underestimated. The resectability rate depends on the tumor's location (5-30% in hilar forms). **Objective:** Evaluate the surgical treatment outcomes of extrahepatic cholangiocarcinoma. **Patients and Methods:** This retrospective, single-center, and analytical study spanned from January 1, 2018, to September 30, 2023, involving 20 patients (excluding Klatskin tumors). The mean patient age was 65 years (range: 55-74), with a male-to-female ratio of 2.3. Most patients were classified as ASA I and II (American Society of Anesthesiologists). **Results:** Clinically, jaundice was observed in 100% of cases. Nine patients exhibited general health deterioration. The diagnostic delay averaged 9 months (range: 9-18 months). Histologically, adenocarcinoma was identified in 100% of surgical specimens. Surgery was performed in 40% of patients, while palliative care was applied in 45%. Post-resection morbidity was 35%, and mortality was 37.5%. **Conclusion:** Surgical resection remains the only treatment offering prolonged survival, though five-year survival rates are low, even after curative procedures.

Keywords: Cholangiocarcinoma, resection, histopathology, outcomes.

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INTRODUCTION

Extrahepatic cholangiocarcinoma (ECH) is a malignant tumor with a poor prognosis, for which surgical resection remains the mainstay of treatment. It accounts for 3% of all digestive cancers and is the second most frequent primary liver tumor after hepatocellular carcinoma. The mean age is 70 years, with a male predominance. The five-year survival rate across all stages is less than 5%. Chronic inflammation of the bile ducts is a common risk factor. Clinical symptoms are nonspecific, leading to delayed diagnosis. Symptoms include general health deterioration, jaundice, abdominal pain, and hepatomegaly. Endoscopic biliary drainage (EBD) and radiological biliary drainage (RBD) are the two available techniques, with the choice depending on the operator's expertise. Imaging examinations should be performed before biliary stent placement.

Multiphase thoraco-abdominopelvic CT is the standard imaging method for assessing local, nodal, and metastatic extensions. MR cholangiopancreatography (MRCP), performed before biliary stent placement, provides better precision than CT for biliary extension

assessment and detecting hepatic metastases. Endoscopic ultrasound with fine-needle aspiration is preferred for distal and proximal bilio-pancreatic diseases.

General characteristics of patients:

This retrospective, single-center, and analytical study was conducted from January 1, 2018, to September 30, 2023, and included 20 patients. The mean age was 65 years (range: 55-74), with 14 men and 6 women, yielding a male-to-female ratio of 2.3. Most patients were ASA I or II. Parameters studied included age, gender, preoperative biological parameters, intraoperative findings, postoperative outcomes, and postoperative morbidity and mortality rates.

Clinical, biological, morphological and operative characteristics:

Functional symptoms were dominated by jaundice (100% of cases), weight loss, and general health deterioration. The diagnostic delay was nine months (range: 9-18 months). The mean preoperative hemoglobin was 11.56 g/L, and the mean preoperative albumin was 37.35 g/L.

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Table 1: General, clinical and morphological characteristics of patients

Features	Values
ASA	
ASA I	10 (50%)
ASA II	07 (35%)
ASA III	03 (15%)
Clinics	
Jaundice	20 (100%)
Weight loss	09 (45%)
Alteration of general condition	08 (40%)
Morphological	
Abdominal ultrasound	20 (100%)
Computed tomography	14 (70%)
Liver MRI	02 (10%)
Bili-MRI	03(15%)
CPRE	01 (5%)
Echo-endoscopy	00
Mean preoperative Hg	11.56
Mean preoperative albuminemia (g/L)	37.35
Midline laparotomy	19 (95%)
Tumor location	
Common bile duct	03 (37.5%)
Lower bile duct	05 (62.5%)
Type of resection	
DPC	05 (26.3%)
Resection of the CBD + Pedicle curettage	03 (15.7%)

Table 2: Operating parameters of patients

Post-operative care	values
Morbidity after resection	4 (50%)
Pancreatic fistula	1 (12.5%)
Biliary fistula	5 (62.5%)
Postoperative hemorrhage	1 (12.5%)
Mortality after resection	3 (37.5%)
Mortality without resection	9 (45%)
Survival after resection	
Patient alive without recurrence at 3 years	01
Patients living without recurrence at 1 year	02

Patient Operative Parameters:

All patients were operated during the inclusion period from January 1, 2018 to September 30, 2023 by median laparotomy. A cephalic duodeno-pancreatotomy was performed in 26.3% of patients and a resection of the common bile duct (CBD) with pedicle curettage in 15.7%. The operative parameters are summarized in (Table 1).

Postoperative parameters of patients: the overall postoperative mortality of the series was 26.3%. We observed postoperative complications after resection in 50% of patients. These were minor complications of grade I and II of clavien and Dindo in 05 (26.3%) patients and major complications of grade III and IV in 01 patient (5.26%). Survival after resection the operative parameters of the patients are summarized in (Table 2).

DISCUSSION

Extrahepatic cholangiocarcinomas (EHC) are categorized into two distinct types: proximal (perihilar) and distal (lower bile duct) EHC. It is important to note that hilar cholangiocarcinoma, also known as Klatskin tumor, is excluded from this classification. A thorough understanding of hilar anatomy, including its variations, is paramount for successful surgical resection of EHC. The anatomical complexity of biliary convergence, which is partially extrahepatic and partially integrated into the hepatic parenchyma, presents unique challenges. Specifically, during surgical resection, lowering the hilar plate is often necessary to access the upper edge of the biliary convergence.

This technical requirement underscores the importance of meticulous preoperative planning and an experienced surgical approach [6-8]. The clinical presentation of EHC is highly variable, with jaundice

being the most common initial symptom. In our study, late diagnosis, advanced patient age, and underlying comorbidities significantly complicated management. Diagnosis predominantly relies on imaging modalities, with thoraco-abdomino-pelvic computed tomography (CT) being the cornerstone of assessment. CT imaging, conducted in four phases (non-contrast, arterial, venous, and delayed), enables precise localization of the tumor, evaluation of hepatic condition, and detection of both locoregional and systemic involvement. CT is particularly valuable as the reference modality for assessing metastatic disease (Figure 1). Magnetic resonance imaging (MRI) complements CT by offering superior visualization of hepatic anatomy and the biliary tree, particularly in cases of high biliary strictures [3, 4, 7,8]. Histological confirmation is most commonly achieved through bile duct brushing during endoscopic retrograde cholangiopancreatography (ERCP) or percutaneous techniques. While obtaining histological proof is strongly recommended prior to initiating palliative or neoadjuvant therapy, it is not mandatory before pursuing curative surgical treatment. [7, 8, 9, 10, 11], In our series, preoperative imaging played a pivotal role in therapeutic decision-making, significantly reducing the risk of unnecessary laparotomies, which could otherwise negatively impact patient outcomes. The

study aimed to evaluate the surgical outcomes of EHC, emphasizing the critical impact of early diagnosis and timely intervention on survival. Delays between symptom onset and treatment initiation often compromise prognosis. [12-16]. Optimal preoperative preparation, including nutritional support and correction of severe jaundice, is essential for improving surgical outcomes. The primary objective of EHC resection is complete tumor excision (R0 resection) with negative margins, while preserving sufficient liver volume to maintain postoperative hepatic function. Despite achieving R0 resection, the 5-year survival rate for cholangiocarcinoma remains low at 5–10%, reflecting the aggressive nature of the disease. [17-22]. In our cohort, the resectability rate was limited to 40%, primarily due to delayed diagnoses. Advances in surgical techniques, including resections involving vascular structures with appropriate repair methods, have improved outcomes in select cases. Commonly performed procedures include resection of the bile duct with lymph node dissection for proximal EHC (Figure 2) and cephalic duodenopancreatectomy (CBD) with lymph node dissection for distal EHC (Figure 3) [23-26]. Prognostic factors influencing outcomes include lymph node involvement, vascular invasion, and microscopic residual disease (R1 status).

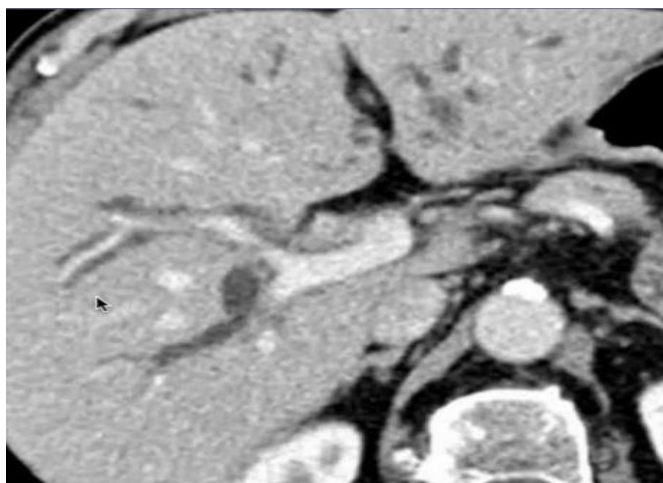


Figure 1: Extrahepatic cholangiocarcinoma

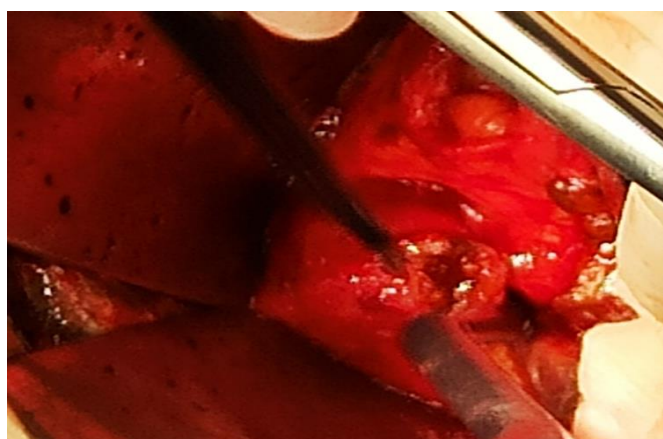


Figure 2: Common bile duct resection (Cholangiocarcinoma of the middle 1/3 of the CBD)



Figure 3: Pièce opératoire d'une Duodéno-Pancréatectomie Céphalique (Cholangiocarcinome du 1/3 inférieur de la VBP)

However, the presence of a single poor prognostic factor does not inherently contraindicate surgical intervention. Surgical exploration with intraoperative histological assessment of resection margins remains an essential practice. [27-29]. Major hepatic or pancreatic resections for EHC are associated with higher morbidity and mortality rates compared to similar procedures for other indications, primarily due to patient age, comorbidities, and jaundice-related complications. In our study, the morbidity and mortality rates were high (50% and 37.5%, respectively), reflecting the challenges posed by delayed management and the technical expertise required for these complex procedures [30-32]. The presence of tumor adenopathies beyond the first lymph node relay represents a contraindication to curative resection. The morbidity and mortality of major hepatic (or pancreatic) resections for biliary cancer is higher than that of the same resections performed for other indications and of biliary resections alone [33-36]. In particular due to the often advanced age or an altered general condition, in particular by jaundice [37-40]. Biliary drainage before curative surgery seems to improve the results of surgery [41-45]. In our series, the morbidity and mortality rate is high (50% vs 37.5%). This increase can be explained in large part by a delay in management and a certain expertise that the surgeon must acquire to guarantee an adequate resection procedure. Patient selection is essential in order to reduce operative mortality.

CONCLUSION

The curative treatment of cholangiocarcinoma primarily involves oncological surgical resection. MRI is the most effective tool for operative indications and therapeutic strategy selection. This cancer is often diagnosed at an advanced stage, and the outcomes of radical surgery remain poor. Multidisciplinary discussions should be encouraged for better patient management. Improvements in techniques and expertise in hepatobiliary surgery at specialized centers have led to lower morbidity and mortality rates.

Conflicts of Interest: The authors declare that they have no conflicts of interest concerning this article.

Authors' Contributions: Abdelkrim Chetibi designed the study and wrote the manuscript. All authors read and approved the final version of the manuscript.

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