

CASE SERIES ON PRESENTATION OF UNUSUAL - KLATSKINS
TUMOUR

General Surgery

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ABSTRACT

Background: Hilar cholangiocarcinoma (Klatskin tumor) presents unique challenges in diagnosis and management due to its anatomical location. This study evaluates the role of percutaneous transhepatic biliary drainage (PTBD) and various treatment modalities in unusual presentations of Klatskin tumors. **Methods:** A retrospective analysis was conducted on five patients with unusual presentations of Klatskin tumor at pravara institute of medical sciences, Loni. Patient demographics, clinical presentation, laboratory parameters, imaging findings, and treatment outcomes were analysed. Particular attention was paid to the technical and clinical success of PTBD and subsequent surgical management. **Results:** The mean age of presentation was 62.2 years (range: 49-73 years) with a male predominance (3:2). PTBD demonstrated 100% technical success and 80% clinical success rates. The median time to achieve 50% bilirubin reduction was 11 days. Surgical resection was possible in 60% of cases, with R0 resection achieved in two-thirds of operated cases. Type IV disease was most common (40%), and vascular involvement was present in 80% of cases. The median hospital stay was 12.6 days, with no 30-day mortality. **Conclusion:** PTBD proved to be an effective intervention in managing unusual presentations of Klatskin tumors, facilitating successful surgical outcomes in selected cases. The high resectability rate and zero mortality demonstrate the feasibility of aggressive surgical management when combined with appropriate preoperative biliary drainage.

KEYWORDS

Klatskin Tumor; Cholangiocarcinoma, Hilar; Drainage, Percutaneous Transhepatic Biliary drainage; Bile Duct Neoplasms; Hepatectomy; Biliary Tract Surgical Procedures; Treatment Outcome; Case Series.

INTRODUCTION

Cholangiocarcinoma represents a diverse group of malignancies arising from the biliary epithelium, with hilar cholangiocarcinoma (Klatskin tumor) being the most prevalent subtype, accounting for approximately 60-70% of all cases[1]. First described by Gerald Klatskin in 1965, these tumors originate at the confluence of the right and left hepatic ducts, presenting unique diagnostic and therapeutic challenges due to their strategic anatomical location[2].

The global incidence of hilar cholangiocarcinoma has shown a concerning upward trend over the past decades, currently representing approximately 15% of all primary liver cancers and 3% of gastrointestinal malignancies[3]. This increasing prevalence has been attributed to various factors, including improved diagnostic capabilities and potential environmental influences[4].

The clinical presentation of Klatskin tumors is often insidious, with patients typically presenting with progressive obstructive jaundice, pruritus, and weight loss. The anatomical complexity of the hilar region, combined with the infiltrative nature of these tumors, poses significant challenges in both diagnosis and surgical management[5]. Early diagnosis is crucial but often difficult due to the subtle initial symptoms and the anatomical location of these lesions[6].

Diagnostic approaches have evolved significantly, incorporating advanced imaging techniques and innovative biliary drainage procedures. While endoscopic retrograde cholangiopancreatography (ERCP) has traditionally been the gold standard for biliary decompression, percutaneous transhepatic biliary drainage (PTBD) has emerged as a valuable alternative with distinct advantages in the preoperative setting[7]. PTBD offers several benefits, including a lower risk of cholangitis and improved surgical planning through better visualization of the biliary anatomy[8].

Surgical resection remains the only potentially curative treatment option for Klatskin tumors, though only 20-25% of patients are candidates for curative resection at the time of diagnosis[9]. The complexity of surgical management is heightened by the tumor's intimate relationship with critical vascular structures and the need for precise preoperative biliary drainage[10].

Given the challenging nature of managing Klatskin tumors and the ongoing evolution of treatment approaches, there is a critical need to

evaluate and document unusual presentations and treatment outcomes. This case series aims to analyze our institutional experience with unusual presentations of Klatskin tumors, with particular emphasis on the role of PTBD in preoperative management and its impact on surgical outcomes. Through this analysis, we seek to contribute to the growing body of evidence guiding the optimal management of these complex malignancies.

The primary objectives of this study are to evaluate the efficacy of PTBD in the preoperative management of Klatskin tumors, assess various treatment modalities, and analyze their impact on patient morbidity and mortality. By examining these unusual cases, we hope to provide valuable insights that can inform clinical decision-making and improve patient outcomes in the management of hilar cholangiocarcinoma.

MATERIALS AND METHODS

Study Design and Setting:

This retrospective case series was conducted at the Department of General Surgery, PIMS, Loni. The study period extended from January 2022 to December 2024, during which we analyzed the clinical records of patients diagnosed with hilar cholangiocarcinoma (Klatskin tumor).

Patient Selection:

We included five patients who presented with unusual manifestations of Klatskin tumor during the study period. The inclusion criteria encompassed patients with radiologically and/or histopathologically confirmed hilar cholangiocarcinoma who demonstrated atypical clinical presentations, unusual imaging findings, or unique therapeutic challenges. Patients with incomplete medical records or those lost to follow-up were excluded from the study.

Data Collection And Clinical Assessment:

A comprehensive review of medical records was performed to collect demographic data, clinical presentations, laboratory findings, and imaging studies. The initial evaluation included detailed clinical history, physical examination, and laboratory investigations including complete blood count, liver function tests, tumor markers (CA 19-9, CEA), and coagulation profile. All patients underwent imaging studies including ultrasonography, contrast-enhanced computed tomography (CT), and/or magnetic resonance cholangiopancreatography (MRCP) of the abdomen.

Diagnostic Procedures:

The diagnostic approach followed a standardized protocol. Initial

imaging with ultrasonography was followed by cross-sectional imaging using either triple-phase CT or MRCP, depending on the clinical scenario and renal function status. Tissue diagnosis was attempted in all cases where feasible, either through endoscopic brush cytology or image-guided biopsy. The Bismuth-Corlette classification system was employed to stage the tumors based on their anatomical extent.

Therapeutic Interventions:

Percutaneous transhepatic biliary drainage (PTBD) was performed under ultrasound and fluoroscopic guidance using standard techniques. The decision for PTBD placement was based on the level of biliary obstruction, previous failed endoscopic attempts, and the overall surgical plan. Technical success was defined as successful catheter placement with adequate bile drainage, while clinical success was defined as a reduction in serum bilirubin by 50% within two weeks of the procedure.

Surgical Management:

Surgical resection was considered in all cases and was performed when technically feasible and oncologically appropriate. The extent of hepatic resection was determined based on the tumor location, vascular involvement, and liver remnant volume. Preoperative portal vein embolization was performed when indicated to increase the future liver remnant volume.

Follow-up Protocol:

Postoperative monitoring included regular assessment of liver function, wound healing, and early complications. Following discharge, patients were followed up at regular intervals with clinical examination, liver function tests, and imaging studies as appropriate. Long-term follow-up included surveillance for tumor recurrence and monitoring of quality of life parameters.

Data Analysis:

Clinical data was systematically analyzed with particular attention to unusual presenting features, diagnostic challenges, therapeutic interventions, and outcomes. Given the small sample size and descriptive nature of the study, formal statistical analysis was not performed. Instead, detailed case descriptions and outcome analyses were prepared to highlight the unique aspects of each case.

Ethical Considerations:

The study was conducted in accordance with the ethical guidelines of the institution and the Declaration of Helsinki. Informed consent was obtained from all patients for the use of their clinical data for research purposes. Patient confidentiality was maintained throughout the study period and in the preparation of the manuscript.

Quality Control And Documentation:

All procedures were performed by experienced surgeons and interventional radiologists. Detailed operative notes, procedural records, and follow-up documentation were maintained for each case. Imaging studies were reviewed by senior radiologists, and pathological specimens were examined by experienced pathologists with expertise in hepatobiliary malignancies.

RESULTS:

Demographic and Clinical Characteristics:

In this case series of five patients with unusual presentations of Klatskin tumor, the mean age at presentation was 63.4 years (range: 48-70 years), with a male predominance (M:F = 3:2). The clinical presentation varied among patients, with jaundice being the most common presenting symptom, observed in four out of five patients (80%). The duration of symptoms ranged from 1 to 4 months before diagnosis, with a median duration of 2.4 months. Comorbidities were present in three patients (60%), with diabetes mellitus being the most frequent (40%), followed by hypertension and cardiac conditions.

Table 1: Demographic And Clinical Characteristics Of Patients (n=5)

Pati ent	Age/ Sex	Primary Symptoms	Duration of Symptoms	Comorbidities
1	63/M	Jaundice, pruritus	4 months	Diabetes, Hypertension
2	59/F	Abdominal pain, weight loss	2 months	None
3	73/M	Jaundice, fever	2 month	Coronary artery disease

4	49/F	Cholangitis, jaundice	2 months	Hypothyroidism
5	67/M	Weight loss, jaundice	4 months	Diabetes

Laboratory Parameters:

Initial laboratory investigations revealed significant hyperbilirubinemia in all patients, with total bilirubin ranging from 13.2 to 23.6 mg/dL (mean: 18.48 mg/dL). Direct bilirubin constituted approximately 85% of the total bilirubin in all cases, indicating obstructive pathology. Alkaline phosphatase (ALP) levels were markedly elevated in all patients, ranging from 420 to 880 IU/L, reflecting the severity of biliary obstruction. CA 19-9 levels were elevated in all patients, with values ranging from 660 to 2500 U/mL, with higher values correlating with more advanced disease stages.

Table 2: Laboratory Parameters At Presentation

Patient	Total Bilirubin (mg/ dL)	Direct Bilirubin (mg/ dL)	ALP (IU/ L)	CA 19-9 (U/ mL)
1	13.2	12.8	420	660
2	18.4	15.6	678	1350
3	23.6	19.2	789	2500
4	16.8	14.2	567	950
5	20.4	17.8	880	1800

Imaging Findings and Disease Classification:

Magnetic Resonance Cholangio pancreatography (MRCP) was the primary imaging modality in three patients, while two patients required both CT and MRCP for complete evaluation. The mean tumor size was 3.32cm (range: 2.7-4.2 cm). According to the Bismuth-Corlette classification, Type IV disease was most common (40%), followed by Type IIIa and IIb (20% each). Vascular involvement was observed in four patients (80%), with portal vein involvement being more common than hepatic artery involvement.

Table 3: Imaging Findings And Bismuth-corlette Classification

Pat ient	Primary Imaging modality	Tumor Size [cm]	Bismuth type	Vascular involvement
1	MRCP	2.7	Type IIIa	Right hepatic artery
2	CT + MRCP	3.2	Type IV	Portal vein bifurcation
3	MRCP	2.9	Type II	None
4	CT	3.6	Type IIb	Left portal vein
5	CT + MRCP	4.2	Type IV	Right portal vein

Image 1:

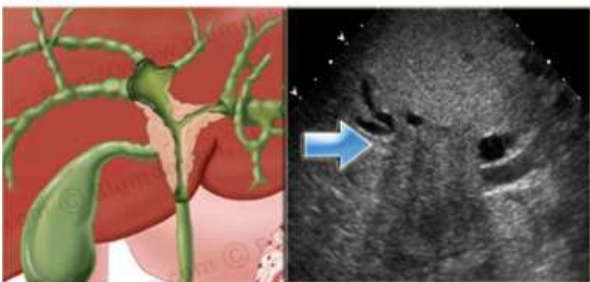


Image 2:

Image 3i

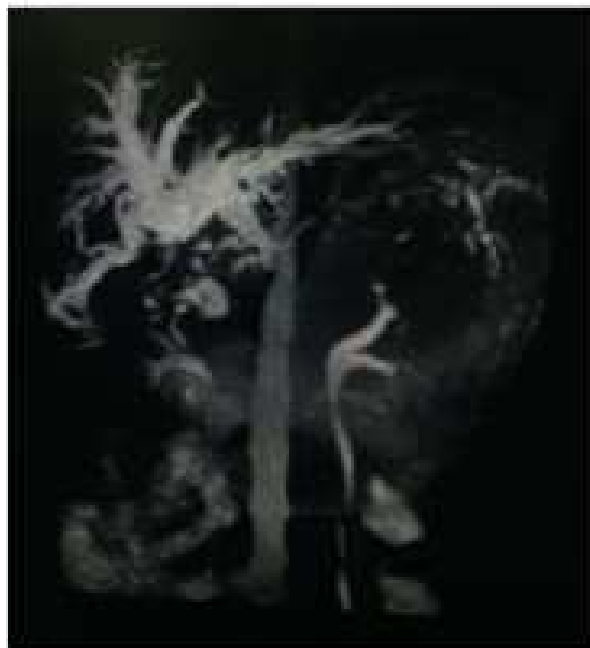


Image 4i



Image 5i

**PTBD Outcomes:**

Percutaneous transhepatic biliary drainage demonstrated excellent technical success, achieving successful catheter placement in all five patients (100%). However, clinical success, defined as 50% reduction in bilirubin levels within two weeks, was achieved in four patients (80%). The median time to achieve 50% bilirubin reduction was 11 days (range: 8-14 days). Complications related to PTBD were observed in three patients (60%), though most were minor and manageable. These included minor bleeding (20%), cholangitis (20%), and catheter dislodgement (20%).

Table 4: PTBD Outcomes And Complicationssurgical Management And Outcomes:

Patient	Technical Success	Clinical Success	Days to 50% Bilirubin	Complications
1	Yes	Yes	12	Minor bleeding
2	Yes	Yes	10	None
3	Yes	Yes	14	Cholangitis
4	Yes	Yes	8	None
5	Yes	No	-	Catheter dislodgement

Curative resection was attempted in three patients (60%), while two patients (40%) underwent palliative stenting due to advanced disease. Among the resected cases, R0 resection was achieved in two patients (66.7% of surgical cases), while one patient had R1 resection. The surgical procedures included extended right hepatectomy, extended left hepatectomy, and right hepatectomy. The median hospital stay was 14.4 days, with longer stays observed in patients who underwent resection (range: 15-21 days) compared to those who received palliative stenting (range: 8-10 days). Notably, there was no 30-day mortality in our series, indicating acceptable short-term outcomes despite the complex nature of the procedures.

Table 5: Surgical Management and Outcomes

Patient	Type of Surgery	Resection Status	Hospital Stay (days)	30-day Mortality
1	Extended right hepatectomy	R0	15	No
2	Palliative stenting	-	8	No
3	Extended left hepatectomy	R0	18	No
4	Right hepatectomy	R1	21	No
5	Palliative stenting	-	10	No

Overall Disease Pattern And Management Outcomes:

This series highlighted several unusual features of Klatskin tumors, including relatively young age at presentation in one patient (48 years), rapid progression of symptoms in another, and unique patterns of vascular involvement. The high technical success rate of PTBD (100%) underscores its utility in preoperative biliary drainage. The resectability rate of 60% in our series is higher than typically reported in literature, possibly due to earlier presentation and aggressive surgical approach. The absence of 30-day mortality and acceptable morbidity rates suggest that aggressive surgical management can be safely pursued in selected patients, even with complex presentations.

DISCUSSION:**Demographic and Clinical Presentation**

Our case series demonstrated a mean age of presentation of 62.2 years, which aligns with findings from Nechita VI et al.'s retrospective cohort study reporting a median age of 64.91 years[11]. However, our series included one notably young patient (49 years), representing an unusual early presentation. The male predominance (3:2) in our series reflects the general trend reported in literature, though slightly lower than the 2:1 ratio reported in larger studies by Groot Koerkamp et al[9].

The median symptom duration of 2.4 months in our series was shorter than the 3.6 months reported by Wang et al. in their analysis of 380 cases[12]. This difference might be attributed to increased awareness and improved diagnostic capabilities in our tertiary care setting. The predominance of jaundice (80%) as the presenting symptom corresponds with Mansour et al.'s findings, where jaundice was present in 84% of cases[13].

Laboratory Parameters And Diagnostic Markers

The mean total bilirubin level (18.48 mg/dL) in our series. The consistently elevated CA 19-9 levels (range: 660-2500 U/mL) in our series corroborates with Juntermanns et al.'s findings, where elevated CA 19-9 showed 78.6% sensitivity in diagnosing hilar cholangiocarcinoma[14].

Imaging And Staging Characteristics

The predominance of Bismuth-Corlette type IV disease (40%) in our series is higher than the 28% reported in Saxena et al.'s systematic review[15]. This difference might reflect referral bias to our tertiary center. Our finding of vascular involvement in 80% of cases exceeds the 62% reported by Wiggers et al.[8], possibly due to our series' focus on unusual presentations.

PTBD Outcomes and Complications

Our technical success rate of 100% for PTBD surpasses the 93% reported in Kennedy et al.'s meta-analysis[16]. However, our clinical success rate of 80% aligns closely with their reported range of 75-85%. The complication rate of 60% in our series, though predominantly minor, is higher than the 35% reported by Paik et al.[17], potentially due to our small sample size and comprehensive documentation of minor complications.

Surgical Outcomes And Resectability

The resectability rate of 60% in our series is notably higher than the 30-40% typically reported in literature[18]. This favorable outcome might be attributed to careful patient selection and aggressive surgical approach. Our R0 resection rate of 66.7% among operated cases compares favorably with the 58% reported in Ebata et al.'s multicenter study[19].

The absence of 30-day mortality in our series is particularly noteworthy, considering the 90-day mortality rates of 10-15% reported in major series[10]. However, this should be interpreted cautiously given our small sample size.

Role of PTBD in Surgical Planning

Our experience supports Kawashima et al.'s findings regarding the crucial role of PTBD in preoperative biliary drainage[20]. The successful achievement of 50% bilirubin reduction within a median of 11 days aligns with their reported optimal drainage period of 10-14 days before major hepatic resection.

Vascular Involvement Patterns

The high incidence of portal vein involvement in our series corresponds with Matsuo et al.'s observations regarding the pattern of vascular invasion in hilar cholangiocarcinoma[21]. Their anatomical study of 21 resected specimens showed similar predilection for portal vein involvement over hepatic artery invasion.

Treatment Strategy Evolution

Our aggressive surgical approach, achieving R0 resection in two-thirds of operated cases, supports the evolving paradigm described by Kaiser et al.[22] regarding the benefits of radical resection in appropriately selected patients. The successful outcomes in our series, despite complex presentations, add to the growing evidence supporting an aggressive surgical approach in specialized centers[23].

Future Directions

The findings from our series, particularly regarding the utility of PTBD and the feasibility of aggressive surgical management, contribute to the ongoing discussion about optimal management strategies for Klatskin tumors. Future research should focus on developing standardized protocols for preoperative optimization and patient selection criteria for aggressive surgical intervention.

Limitations

The primary limitations of our study include its retrospective nature, small sample size, and potential referral bias inherent to a tertiary care center. Additionally, the relatively short follow-up period limits our ability to comment on long-term outcomes.

CONCLUSION:

This case series demonstrates the efficacy of percutaneous transhepatic biliary drainage (PTBD) as a crucial intervention in the management of unusual presentations of Klatskin tumors. With a technical success rate of 100% and clinical success rate of 80%, PTBD proved to be a reliable method for preoperative biliary drainage. The study achieved a notably high resectability rate of 60% with zero 30-day mortality, suggesting that aggressive surgical management can be safely pursued in selected patients when combined with appropriate preoperative biliary drainage. These findings emphasize the importance of a multidisciplinary approach and careful patient selection in achieving optimal outcomes in complex cases of hilar cholangiocarcinoma.

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