Case report

Squamous cell carcinoma arising within choledochal cyst managed by pancreaticoduodenectomy: A case report

Sujan Shrestha a,⁎, Ramesh Singh Bhandari a, Bibek Man Shrestha b, Suraj Shrestha b, Nirajan Subedi a, Sumita Pradhan a

a Department of GI and General Surgery, Tribhuvan University Teaching Hospital, Institute of Medicine, Kathmandu, Nepal
b Maharajgunj Medical Campus, Institute of Medicine, Kathmandu, Nepal

A R T I C L E   I N F O

Article history:
Received 17 March 2021
Received in revised form 17 April 2021
Accepted 19 April 2021
Available online 27 April 2021

Keywords:
Choledochal cyst
Pancreaticoduodenectomy
Squamous cell carcinoma

A B S T R A C T

Introduction and importance: Choledochal cysts (CC) are rare cystic dilation of the biliary trees. Malignancy is one of the important significant findings in CC and its incidence increases with age. Associated squamous cell carcinoma of the choledochal cyst is an uncommon pathological finding.

Case presentation: We present a 21-year-old male diagnosed with type 1 CC and planned for excision of the cystic dilated extrahepatic biliary tract with hepaticojejunal anastomosis in Roux-en-Y but underwent pancreaticoduodenectomy due to intraoperative palpable mass of CC which was adhered to the duodenum and pancreatic head. Histopathology of the excised specimen revealed squamous cell carcinoma (SCC) of the choledochal cyst.

Discussion: CC represents a rare biliary cystic disease. Though infrequent, malignant transformation in CC includes cholangiocarcinoma, adenocarcinoma, and rarely SCC. The post-operative management for SCC in CC is ill-defined and carries a grave prognosis.

Conclusion: The choledochal cyst should be completely excised wherever possible to avoid the possible malignant transformation in CC including SCC.

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1. Introduction

Choledochal Cysts (CC) are infrequent congenital cystic dilation of the biliary tract, with an approximate incidence of 1:1000 in the Asian population. Choledochal Cysts are four times more common in females and are usually diagnosed in infants and young children within the first decade of their life [1].

Todani and colleague’s modification of Alonso-Lej types is the most validated classification for CC used in general practice which divides the CC into five major types. Type I is dilatation of extrahepatic biliary duct. Type II is the formation of diverticulum of the common bile duct, choledochocoele involving an intraduodenal portion of common bile duct represents Type III. Type IV is intra and extrahepatic duct dilatation and Caroli’s disease is Type V [2]. Cholelithiasis, common bile duct stone, cholangitis, pancreatitis, portal hypertension, and liver function test abnormalities are common complications in patients with CC [3]. The management of CC is surgical and varies according to the type; ranging from cyst excision to orthotopic liver transplant [4].

However, the most concerning about CC is its malignant transformation. The incidence of malignancy has been reported in 7.6% of patients with type I, 4.3% with type II, 4% with type III, 9.2% with type IV, and 2.5% with type V CC [5]. Cholangiocarcinoma, adenocarcinoma, carcinoma gall bladder are the common malignancies. The finding of squamous cell carcinoma (SCC) in CC is rare with very few cases reported in the literature [6].

Herein, we present a case of large CC managed by pancreaticoduodenectomy. To our surprise, histopathological examination of the specimen revealed SCC of CC. This case has been reported in line with SCARE guidelines [7].

1.1. Case presentation

A 21-year-old male presented to our hospital with complaints of intermittent, mild epigastric pain for one year and progressive yellowish discoloration with pruritus of the body for the past two months. He had also noticed a lump at the right hypochondrium around 45 days before admission while taking a bath. There was no history of anorexia, significant weight loss, fever, and other constitutional symptoms or similar illness among any family members.

On examination, he was of average built with normal vital signs as well as normal systemic and physical examinations. He was icteric and there were scratch marks with petechial hemorrhage present over his body. Abdominal examination revealed a palpable non-tender gallbladder.
His routine hemogram was normal including total counts and C-reactive protein except for his liver function tests and coagulation profile. His total and direct bilirubin were 297 and 120 mmol/L respectively, INR of 3.75, and alkaline phosphatase (ALP) of 672 IU/L. Ultrasound abdomen and pelvis revealed 14.9*8.7*9.6 cm choledochal cyst with distended gallbladder with multiple stones. Contrast-Enhanced Computed Tomography (CECT) was planned for better delineation of cyst anatomy, which revealed large type 1 CC with distended gallbladder with multiple cholelithiases (Fig. 1). Considering a possible choledochal cyst complication including cholangiocarcinoma, CA19-9 was sent but was only marginally raised to 45 U/mL (Range: <37 U/mL). Considering the poor financial situation of the patient and evident diagnosis on CECT, MRI or Magnetic resonance cholangiopancreatography (MRCP) was not performed.

With the diagnosis of type 1 CC, the patient was prepared for Choledochal Cyst excision with Roux-En-Y hepaticojejunostomy after the correction of the coagulation profile by a team of gastrointestinal surgeons. Open surgery was performed. Intraoperatively, there was a distended gallbladder and large CC with a hard mass on its lower part along with dense adhesion of the distal part of CC with the second part of the duodenum and the pancreatic head was noticed (Fig. 2). Due to above mentioned intraoperative findings, pancreaticoduodenectomy was performed along with dunking pancreaticojejunostomy, interrupted hepaticojejunostomy, and gastrojejunostomy (Fig. 3). The histopathological examination of the surgical specimen revealed moderately differentiated SCC with atypical squamoid cells arranged in cords and nests (Fig. 4) within the CC along with an extension of tumor to duodenum and pancreas with perineural invasion. However, lymphatic and vascular invasions were absent and the resected margins were free of tumor. Additionally, eighteen lymph nodes were sampled, and all nodes and
other margins were negative for tumor cells with pathological cancer stage of pT2pN0.

His postoperative period was uneventful. The patient recovered well and was discharged on the ninth postoperative day. The patient is receiving paclitaxel-based chemotherapy and is doing well after 3 months of surgery. He is on close follow-up and has no issues to date.

2. Discussion

CC represents a rare biliary cystic disease. The incidence of abdominal pain in pediatric and adult patients ranges from 78% to 90%, jaundice, and cholangitis being in 40 to 50% [8]. The classical triad of abdominal pain, mass, and jaundice in adults with CC is found in a minority of patients and ranges from 0 to 17% [9]. Our patient was a young male who presented with a triad of abdominal pain, a palpable lump in the right hypochondrium, and obstructive jaundice.

Cystic dilatation of bile ducts due to an anomalous pancreaticobiliary ductal union is the most accepted hypothesis stated in Babbitt’s theory in the formation of CC with anomalous pancreaticobiliary ductal union seen in 0–17% of all CC [2]. There was no evidence of anomalous pancreaticobiliary ductal union in our case. Well-performed USG is very sensitive (71 to 97%) in the detection of CC but MRCP is the gold standard and the choice of diagnostic tool preoperatively for CC. MRCP is a highly sensitive and specific, safe, and non-invasive diagnostic preoperative technique for the diagnosis of CC [10]. We preferred CECT as a diagnostic tool in our case with the possibility of another complex hepatobiliary lesion and taking into consideration the financial status of the patient.

Cho et al. mentioned complete excision of the extrahepatic component of CC combined with cholecystectomy, followed by Roux-en-Y biliary reconstruction as the treatment of choice for type I and IV CC [11]. The excision of the extrahepatic component of CC in type IV is used if only the extrahepatic biliary tree is involved. If additive intrahepatic ducts are involved, they are treated additively with intrahepatic dilatation or hepatic lobectomy. Diverticulectomy followed by primary CBD closure and endoscopic sphincterotomy is the treatment of choice for type II and type III CC respectively [12]. Type V management consists of liver resection or orthotopic liver transplant [4]. We also planned for complete excision of the extrahepatic component of CC combined with cholecystectomy, followed by Roux-en-Y biliary reconstruction but, the distal part of the cyst was difficult and densely adhered to the duodenum and posterosuperior part of the pancreas, a pancreaticoduodenectomy was done in our case.

Malignant transformation occurs in about 10–30% of adults with CC [13]. In the patients with CC, in addition to the 5% risk of gallbladder cancer, the risk of cholangiocarcinoma is 14% [14]. A significant number of patients with type IVA develop bile duct cancer and type I develop gallbladder cancer [15]. However, primary SCC of the extrahepatic bile duct is rare, and only 5% of biliary carcinomas are of squamous cell origin, with only a few cases reported in approximately the past 100 years [6]. The inflammation-cancer transformation is the most accepted theory for this rare cancer to occur in CC. Cabot et al. has suggested an inflammatory stimulation as the culprit behind the metaplasia in the epithelial mucus membranes [16]. Other probable pathophysiological mechanisms are genetic deletions and hereditary predisposition [17].

This type of malignancy often shows a high grade of malignancy, early nodal involvement along with systemic metastasis, and thus a poor prognosis [18]. CT scan in our case did not reveal any nodal or organ metastasis. Also, the histopathological examination of the specimen had free tumor margins with no nodal, lymphatic or vascular involvement. In a study, patients with malignancy in choledochal cyst staged I/II and who underwent curative resection, the 5-year survival rates of patients with cholangiocarcinoma arising from a choledochal cyst in stage Ia, Ib, and Ila bile duct cancer were 90.4%, 40.0%, and 25.1%, respectively which was comparable with that of patients with cholangiocarcinoma in general [15].

There are only a few case reports on this rare occurrence of SCC in choledochal cyst managed by pancreaticoduodenectomy. Because of the rare occurrence of SCC in the biliary tree, post-operative management is not well defined. After the completion of careful multidisciplinary discussion on this case with the pathologist and oncologist, he was started on paclitaxel-based chemotherapy and kept in close surveillance and on regular follow-up. However, this type of cancer is often high-grade, and chemotherapy does not significantly prolong the patient’s survival [18].

3. Conclusion

We report a rare case of squamous cell carcinoma within choledochal cyst managed by Pancreaticoduodenectomy. Our findings suggest that SCC in the biliary tree can originate from congenital cystic disease. To avoid possible complications of CC including malignant transformation, it should be completely excised wherever possible. As postoperative management is not well defined for SCC of CC, a multidisciplinary approach can help provide the best treatment options for the patient.

Ethical approval

Not required.

Funding

None.

Authors’ contribution

Sujan Shrestha (SS), Ramesh Singh Bhandari (RSB), Nirajan Subedi (NS), and Sumita Pradhan (SP) = Study concept, Data collection, and surgical therapy for the patient.

SS, Bibek Man Shrestha (BMS) = Writing, original draft preparation.

SS, BMS, Sujan Shrestha and Nirajan Subedi (NS) = Editing and writing.

RSB and SP = senior author and manuscript reviewer.

All the authors read and approved the final manuscript.
Guarantor

Sujan Shrestha.

Registration of research studies

Not applicable.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Declaration of competing interest

None.

Acknowledgment

None.

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