Adenocarcinoma in Choledochal Cyst—a Case Report

Deepak Verma • Kamlesh Damor • Neeraj Kumar •
Nirupama Kothari

Received: 13 September 2013 / Accepted: 3 April 2014 / Published online: 27 April 2014
© Association of Surgeons of India 2014

Introduction

Choledochal cyst is a rare congenital condition and can present at any age with female preponderance. Incidence is high in Asian as compared to Western countries [1]. Clinical presentation is nonspecific, and complications include pancreatitis, cholangitis, biliary cirrhosis, spontaneous rupture, and malignancy [2]. Incidence of malignancy increases with age. Imaging with contrast enhanced computerized tomography (CECT) and magnetic resonance cholangiopancreatography (MRCP) has facilitated the preoperative suspicion of malignancy, and surgical treatment has evolved from cystoenterostomy to primary cyst excision with bilioenteric drainage [2]. We are reporting a preoperatively suspected case of choledochal cyst with malignancy.

Case Report

A 19-year-old male presented with pain and lump in the abdomen of 6-month duration. Patient was icteric, and abdominal examination revealed a mass of 10×10 cm in the right hypochondrium. Investigations revealed raised bilirubin and enzyme levels including alkaline phosphatase. Coagulation profile and CEA was normal.

Ultrasonography (USG) of the abdomen revealed a cystic lesion with intraluminal echogenicity in the subhepatic region and distended gall bladder with dilated cystic and hepatic ducts diagnosed as choledochal cyst. Contrast enhanced

computerized tomography (CECT) revealed a choledochal cyst (Todani type IVa) complicated by polypoidal intraluminal SOL. MRCP also confirmed the findings of CECT (Fig. 1). Preoperative, there was a type IVa choledochal cyst extending below the second part of the duodenum. Excision of the cyst with Roux-en-Y hepaticojejunostomy was done. Two polypoidal masses were attached to the posterior wall of the cyst (Fig. 2). Histopathology revealed early infiltrative moderately differentiated papillary adenocarcinoma developing in the tubulovillous adenoma of choledochal cyst with papillomatous area. Monthly follow-up for the last 9 months had been normal.

Discussion

Choledochal cysts are uncommon congenital anomalies with relatively high incidence in Asian countries specially Japan as compared to Western population [1]. Besides cholangitis, pancreatitis, and cholelithiasis, malignant changes within the duct are reported complications.

The incidence of carcinoma for different age groups is closely related to the increase in age and in patient over 60 years of age; the possibility of malignant change is high [3]. However, it is difficult to develop precise risk based on age alone.

Malignancy may be associated with any type of choledochal cyst, but the greatest prevalence is said to be with type I, type IV (66.7 and 33.35 %, respectively), and type V [3]. Type II cysts are at minimal risk of malignant degeneration [4].

Choledochal cysts are supposed to be caused by an APBDJ, allowing pancreatic juice reflux leading to increased pressure and dilatation. Neoplastic changes in choledochal cysts occur as a result of chronic inflammation, cell regeneration, and DNA breaks leading to dysplasia with or without

D. Verma (✉) • K. Damor • N. Kumar
Department of Surgery, Dr. Sampurnanand Medical College,
Jodhpur, Rajasthan, India
e-mail: drdeepakv@gmail.com

N. Kothari
SRL Ranbaxy Laboratory, Jodhpur, Rajasthan, India

Fig. 1 MRCP showing papillary filling defect on the posterior wall of the choledochal cyst

intestinal metaplasia and invasive carcinoma [5]. Pancreatic reflux is also supposed to cause K-ras mutation, cellular atypia, P₅₃ overexpression, and carcinogenesis.

Various neoplastic lesions arising from choledochal cyst include adenomyoma, benign biliary papillomatosis, papillomatosis with adenocarcinoma, papillary adenocarcinoma, tubular adenocarcinoma [6], squamous cell carcinoma adenocanthoma, anaplastic carcinoma, and bile duct sarcoma. Site of malignant lesion may be in the extra- or intrahepatic biliary tract, gall bladder, liver, and pancreas.

There are no specific or sensitive clinical indicators of malignant transformation. Weight loss was more common in patients with malignancy [7]. Imaging techniques helps in early diagnosis. USG has a sensitivity of 71–97 %, whereas CTCP has a 90 % sensitivity [2]. MRI and MRCP are gold standard with a sensitivity of 90–100 %. It avoids ionizing radiation and complications like cholangitis and pancreatitis. Papillary structure in dilated biliary duct increases suspicion of malignancy.

Fig. 2 Cut specimen of excised cyst showing papillomatous growth from mucosal surface of the cyst

Standard surgical treatment of choledochal cyst with malignancy is excision of the cyst hepaticojejunostomy [8] to avoid complications such as cholangitis, biliary cirrhosis, portal hypertension, lithiasis, rupture, and pancreatitis frequently encountered in cases with malignant change. Pancreaticoduodenectomy is another modality but has higher morbidity and mortality.

The prognosis for malignancy in choledochal cyst is grim with medial survival of 6–21 months [7], and the 2-year survival rate is < 5 %.

References

1. O'Neill JA Jr (1992) Choledochal cyst. *Curr Probl Surg* 29:361–410
2. Bhavsar MS, Vora HB, Giriappa VH (2012) Choledochal cysts: a review of literature. *Saudi J Gastroenterol* 18(4):230–236
3. Feng JF, Chen WY, Chen DF, Zhou S, Liu J (2011) Choledochal cysts with malignancy in adults: a retrospective study with an experience of twenty–two years. *Pak J Med Sci* 27(1):6–10
4. Tajiri H (1996) Choledochocoele-containing stones. *Am J Gastroenterol* 91(5):1046–1048
5. Benjamin IS (2003) Biliary cystic disease: the risk of cancer. *J Hepato-Biliary Pancreat Surg* 10(5):335–339
6. Kraus I, Rubinic M, Uravic M, Kovac D, Brncic N, Kraus D, Voinikovic B (2003) Early cancer in congenital choledochal cyst. *Coll Antropol* 27(2):677–683
7. Nagorney DM, McIlrath DC, Adson MA (1984) Choledochal cysts in adults. *Clin Manag Surg* 96:656–663
8. Tan SS, Tan NC, Ibrahim S (2007) Management of adult choledochal cyst. *Singap Med J* 48(6):524–527