Management of Adult Choledochal Cyst Coexisting with Gallbladder Carcinoma: A Case Report and Review of Literature

Olusegun Isaac Alatise, Olatunbosun Ayokunle Oke, Abdulrasheed Kayode Adesunkanmi, Olaejinrin O Olaofe[1], Christianah Mopelola Asaleye[2]

Departments of Surgery, [1]Morbid Anatomy and [2]Radiology, Obafemi Awolowo University, College of Health Sciences, Ile-Ife, Osun State, Nigeria

Abstract

Choledochal cyst is a relatively rare condition. Even rarer is a choledochal cyst in association with a gallbladder carcinoma. This study reports a rare case of gallbladder carcinoma coexisting with a choledochal cyst in a Nigerian patient. Clinical records of the patient including preoperative evaluation, intraoperative findings, and postoperative care were reviewed. A 38-year-old woman presented with the recurrent right upper abdominal pain of 3 years duration associated with progressive weight loss, anorexia, recurrent vomiting, as well as, low-grade fever with chills and rigors. Physical examination revealed an anicteric woman with tenderness in the right hypochondrium and a positive Murphy's sign. A combination of abdominal ultrasound and computed tomography scan suggested a Type IV choledochal cyst and a distended gallbladder with thickened walls containing a heterogeneous hyperdense mass. Preoperative serum alkaline phosphatase was elevated while endoscopic retrograde cholangiopancreatography was inconclusive. At laparotomy, extrahepatic biliary dilatation and enlarged, the nodular gallbladder was found with a diffusely fibrotic pancreas. Intraoperative cholangiogram confirmed Type IV choledochal cyst. Excision of the common bile duct and radical cholecystectomy was performed, and a Roux-en-Y hepaticojejunostomy. Histopathology confirmed the diagnosis of gallbladder adenocarcinoma. She had adjuvant chemotherapy and is presently on follow-up. No evidence of recurrence after 5 years of follow-up. A high index of suspicion is required to detect a combination of these two rare entities. When detected, both conditions should be surgically addressed at the same sitting, and when combined with adjuvant chemotherapy, may increase the chances of achieving a cure.

Keywords: Adult, choledochal cyst, gallbladder cancer, jaundice, surgery, Nigeria

Address for correspondence:
Dr. Olusegun Isaac Alatise,
Department of Surgery, Obafemi Awolowo University Teaching Hospital Complex, PMB 5538, Ile-Ife, Osun State 220005, Nigeria.
E-mail: segunalatishe@yahoo.co.uk

Access this article online

Quick Response Code:

Website: www.nigerianjsurg.com

DOI:
10.4103/1117-6806.169870

Introduction

Choledochal cyst disease is a pathologic condition characterized by varying degrees of congenital dilatation of the biliary system, including the common, intrahepatic, and intrapancreatic bile ducts.[1] It is a rare congenital anomaly which is, for unknown reasons, quite common among Asians population and very rare in Africans.[2,3] At least 60% of patients are diagnosed during the first decade of life, but 20% remain undiagnosed until adulthood.[2,4] Due to the recent improvement in noninvasive hepatobiliary imaging technique majority of cases of choledochal cysts are now detected in adult in most developed countries.[4-6]

These cysts are clinically important because of their attendant complications of recurrent cholangitis, biliary stricture, choledocholithiasis, recurrent acute pancreatitis, and malignant transformation.[7] The risk of malignant transformation is well-documented to be age-related and believed to be the result of chronic inflammation.[8] The malignant transformation usually arises from the cyst or along the bile duct, especially when there is anomalous pancreaticobiliary junction.[9,10] Very rarely, the cancer

How to cite this article: Alatise OI, Oke OA, Adesunkanmi AK, Olaofe OO, Asaleye CM. Management of Adult Choledochal Cyst Coexisting with Gallbladder Carcinoma: A Case Report and Review of Literature. Niger J Surg 2016;22:43-7.

can occur in the gallbladder or in the head of the pancreas. The frequent occurrences of cancers in adult choledochal cyst make surgical management more challenging.[6]

In Nigeria, only a few cases of choledochal cyst have been reported.[3,11,12] All these cases occurred in children below 3 years. None of the patients had a coexisting malignant transformation. We presented a case of adult choledochal cyst coexisting with gallbladder cancer highlighting the clinical presentation and the management option for such patients in an economic poor resource setting.

Case Report

A 38-year-old woman presented with recurrent right upper quadrant abdominal pain of 3 years duration. The pain was colicky and radiated to the epigastrium and right lumbar region. Episodes were associated with transient jaundice, low-grade fever, anorexia, nonbilious vomiting, and weight loss. Physical examination revealed acutely ill woman anicteric, not pale with right hypochondrial tenderness and a positive Murphy's sign. Complete blood count showed leukocytosis with predominant neutrophilia. Alkaline phosphatase was markedly elevated, but other liver enzymes were essentially normal.

Abdominal ultrasound revealed hepatomegaly (17.3 cm) with fusiform dilatation of the extrabiliary duct [Figures 1 and 2]. The common bile duct (CBD) was also dilated (18.8 mm). The gallbladder was distended with inspissated bile. Its wall was thickened (3.4 mm) and had a lobulated outline. The pancreas and other intra-abdominal organs were essentially normal, and there were no ascites. She was placed on parenteral antibiotics cefuroxime and metronidazole, nil per oral and analgesics. She responded to the conservative management and was discharge home; however, the pain recurred within 1-month which necessitated readmission. The intra-abdominal computed tomography (CT)-scan revealed dilatation of intrahepatic ducts, and the CBD which measured 20 mm in diameter. The gallbladder was dilated and filled with a heterogeneous, hyperdense mass which enhanced positively with contrast [Figures 3 and 4]. Endoscopic retrograde pancreatography (ERCP) was done, but cannulation of the bile duct failed, probably due to anomalous pancreatobiliary junction. A preoperative diagnosis of Type IV choledochal cyst with suspected associated gallbladder tumor was made.

At laparotomy, there was mild hepatomegaly, an enlarged, hard, nodular, gallbladder, and a markedly dilated CBD, extending proximally to the common hepatic duct [Figures 5 and 6]. The diagnosis of Type IVa choledochal cyst was confirmed by intraoperative cholangiogram. Cholecystectomy, Segment 4b and 5 hepatic resection, and complete resection of the extrahepatic cyst up to the confluence of the right and left hepatic ducts, followed by an end-to-side Roux-en-Y hepaticojejunostomy, was performed [Figure 7]. She had an uneventful recovery from the surgery and was discharged on postoperative day 5.

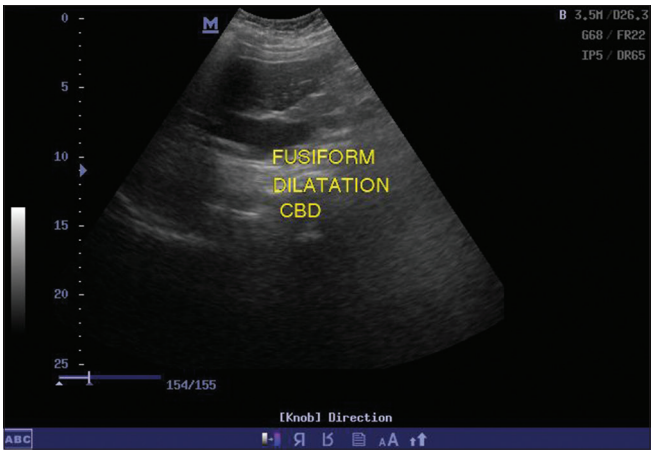

Figure 1: Ultrasound scan picture showing the fusiform dilatation of the common bile duct

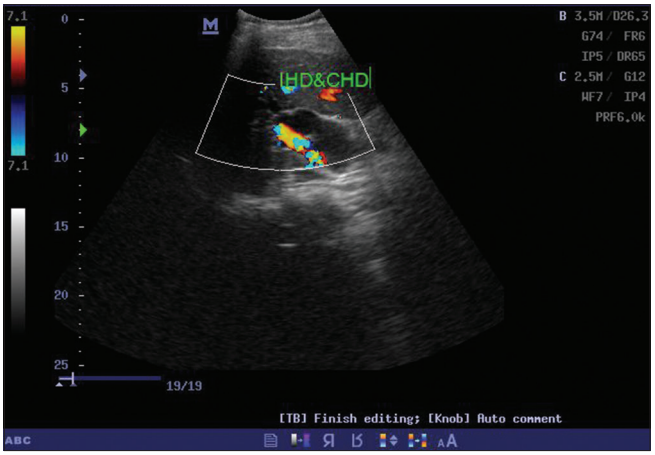

Figure 2: Doppler ultrasound scan picture showing the fusiform dilatation of the common bile and the hepatic artery

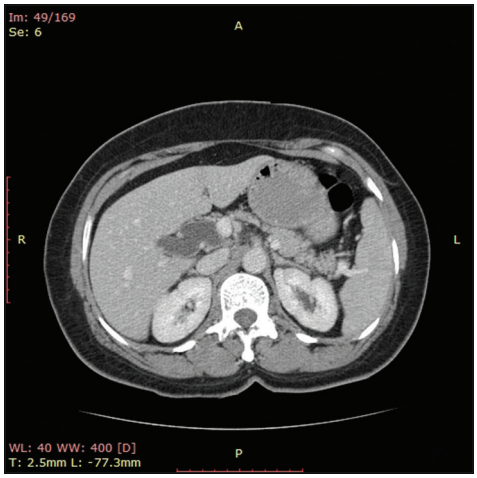

Figure 3: Computerized tomography scan showing fusiform dilatation of the common bile duct

Histopathology confirmed the diagnosis of adenocarcinoma of the gallbladder [Figure 8] there was no tumor in the cyst wall. She had adjuvant chemotherapy with a gemcitabine/cisplatin

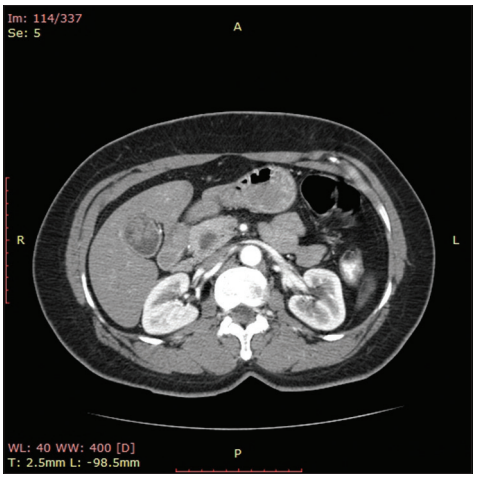

Figure 4: Computerized tomography scan showing thicken gallbladder wall and heterogeneous, hyperdense mass within the lumen

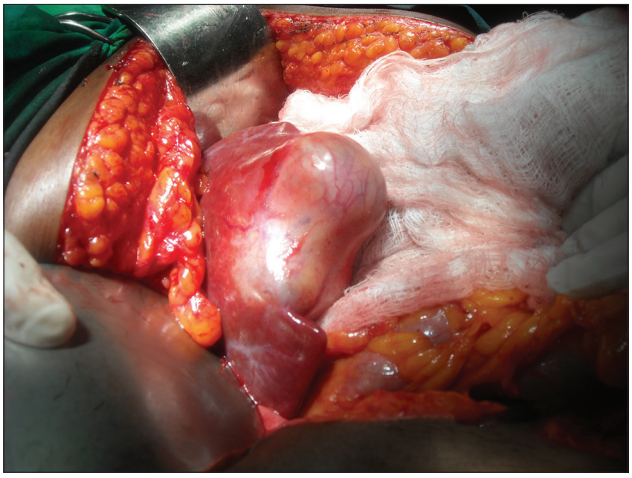

Figure 5: Intraoperative picture showing enlarged, hard, and nodular gallbladder

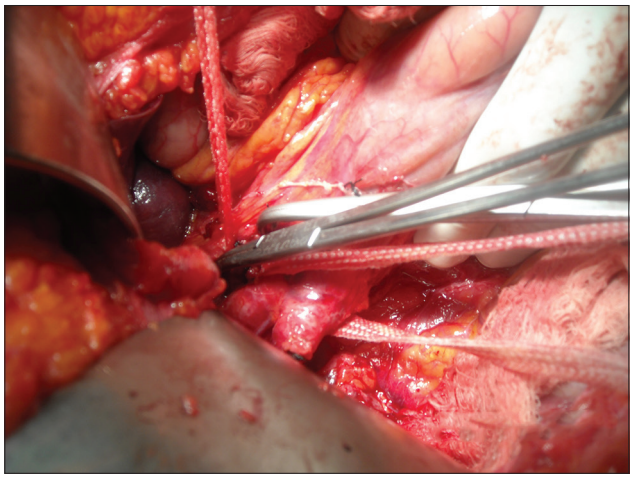

Figure 6: Intraoperative picture showing dilated common bile duct

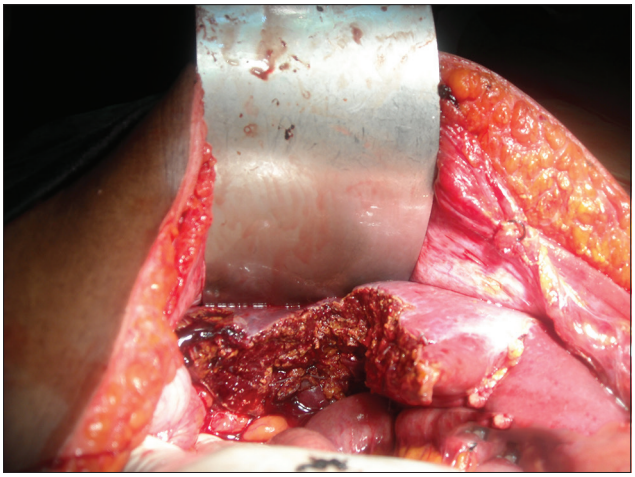

Figure 7: Intraoperative picture showing resection of liver segment 4b and 5

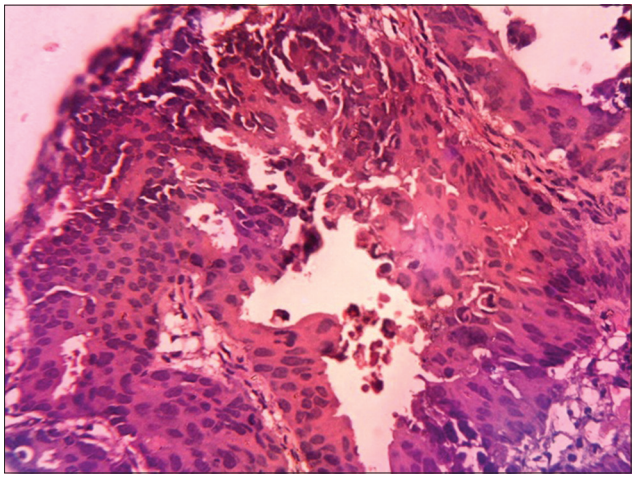

Figure 8: Photomicrography showing tumor infiltration of the entire wall of the gallbladder

combination, and there is no evidence of recurrence during the first 5 years of follow-up.

Discussion

Choledochal cyst is defined as an isolated or combined dilation of the extrahepatic or intrahepatic biliary tree which anatomic detail was first described by Vater and Ezler in 1723.[13] Douglas is credited with the first clinical description of a case of a choledochal cyst.[14] In 1959 Alonso-Lej *et al.* reviewed the literature and proposed the first widely used classification system of choledochal cysts.[15] In 1977 Todani *et al.* modified their classification to account for the combination of intra and extrahepatic cystic dilation, and proposed the five types of congenital choledochal cysts which have gained widespread acceptance.[4]

Choledochal cyst is a very rare congenital anomaly and has been established to occur in 1 in 13,000 to 1 to 2 million live births.[16] More than two-third of all reported cases have originated in Asia. In most series as in this index case report, there is a female preponderance of the condition with the female/male ratio ranging from 3:1 to 8:1.[17,18] The diagnosis of choledochal

cyst is usually made in the first few years of life, and more than 60% of all cases are diagnosed in the first decade.[19] However, many recent reports have emphasized that this disease also is seen in adults.[20]

No clearly defined or scientifically accepted etiology of bile duct cysts currently exists. However, based on clinical and experimental data, several potentially important pathophysiological observations have been offered. Of all these, two main etiologic hypothesis have been favored which include weakness of the wall of the bile duct due to pancreaticobiliary malunion, and obstruction of the distal part of the bile duct.[6] The possibility of anomalous pancreatobiliary ductal union which may be responsible for failed ERCP in our patients may also be the possible predisposing factor in our patient. The frequent occurrence of anomalous pancreatobiliary ductal malfunction in patient with choledochal cyst increase the occurrence of acute pancreatitis following ERCP, hence some authors propose that ERCP should not be done for patients with a choledochal cyst.[21]

While cholangiocarcinoma is the most common malignant transformation in patients with the choledochal cyst, gallbladder carcinoma can also occur (especially in cases like ours) due to the possible occurrence of anomalous pancreatobiliary ductal malfunction.[6-10] Reflux of activated pancreatic enzymes, amylase, bile stasis, and an increased intraductal concentration of bile acids contribute to the proliferative activity of bile duct wall in patients with choledochal cysts.

Clinical presentation of adult choledochal cyst differs from that of neonates or children.[22] Presentation in adults is nonspecific and often leads to delay in diagnosis. The classic triad of abdominal pain, right upper quadrant mass, and jaundice are more common in the pediatric population than adult (85 vs. 25%, respectively).[14] Most adult like our patient usually present with abdominal pain, with or without jaundice, or symptoms suggesting pancreatic, or biliary origin which prompts hepatobiliary imaging before the diagnosis is made. About 50% of the patients do have associated stone which often distract the attention of the attending physician or radiologist from the cyst.[23,24] Unfortunately, cholecystectomy alone does not resolve the symptomatology in such patients. The occurrence of choledochal cyst should always be sorted for in patients with biliary stone.

Fortunately for surgeon working in less developed countries who have limited access to high-tech imaging tools, ultrasound scanning can diagnose choledochal cysts with a specificity of 97%.[17] Ultrasound is, therefore, an excellent first-line investigation of persistence jaundice, and may help to differentiate choledochal cysts from other cause of obstructive jaundice in pediatric or adult patients. However, the gold standard for diagnosing and classifying choledochal cyst is now magnetic resonance cholangiopancreatography.[25] The recent development of multidetector CT has proven useful and more accurate in imaging the pancreaticobiliary system.[26] With more widespread availability of this technique, its role will become clearer.[27]

The aim of treating adult patients with choledochal cyst coexisting with gallbladder cancer is to excise the bile duct and to ensure oncological clearance of the cancer. Resection of the cyst together with cholecystectomy and reconstruction using a standard Roux-en-Y loop for an end-to-side-hepaticojejunostomy is the mainstay of the operative approach.[6,24,28] As opposed to cystojejunostomy, this reduced the risk of malignant transformation, and it also helps to maintain the long-term quality of life.[28,29] Liver resection is not often recommended for the uncomplicated, benign choledochal cyst.[6] However, limited liver resection and lymph node dissection was offered to our patients because of the coexisting gallbladder cancer. We recommend such limited liver resection in setting where most advanced liver sealing agents are not available.

As we found in this patient, excision of bile duct cysts are regarded as a safe procedure, and a number of authors have reported an operative mortality rate of zero over the past decade.[6] Adjuvant chemotherapy is usually required in patients with associated gallbladder cancer. Combination of gemcitabine and cisplatin has been found to be effective in treating gallbladder cancer.[30]

Conclusion

We reported an adult Type IVa choledochal cyst with associated gallbladder carcinoma in Nigerian woman who was successfully managed in our hospital. We suggest high index of suspicion on the part of surgeon managing such adult patient presenting with upper abdominal pain, jaundice and weight loss. Early diagnosis and aggressive surgical resection may provide the only hope of prolonging survival.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Cho MJ, Hwang S, Lee YJ, Kim KH, Ahn CS, Moon DB, *et al.* Surgical experience of 204 cases of adult choledochal cyst disease over 14 years. World J Surg 2011;35:1094-102.
2. Lee SE, Jang JY, Lee YJ, Choi DW, Lee WJ, Cho BH, *et al.* Choledochal cyst and associated malignant tumors in adults: A multicenter survey in South Korea. Arch Surg 2011;146:1178-84.
3. Akinyinka OO, Falade AG, Akinbami FO, Alli T, Atalabi M, Irabor D, *et al.* Choledochal cysts in African infants: A report of 3 cases and a review of the literature. Trop Gastroenterol 2005;26:34-6.
4. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977;134:263-9.
5. Liu CL, Fan ST, Lo CM, Lam CM, Poon RT, Wong J. Choledochal cysts in adults. Arch Surg 2002;137:465-8.

6. Söreide K, Körner H, Havnen J, Söreide JA. Bile duct cysts in adults. Br J Surg 2004;91:1538-48.
7. Wiseman K, Buczkowski AK, Chung SW, Francoeur J, Schaeffer D, Scudamore CH. Epidemiology, presentation, diagnosis, and outcomes of choledochal cysts in adults in an urban environment. Am J Surg 2005;189:527-31.
8. Kobayashi S, Asano T, Yamasaki M, Kenmochi T, Nakagohri T, Ochiai T. Risk of bile duct carcinogenesis after excision of extrahepatic bile ducts in pancreaticobiliary maljunction. Surgery 1999;126:939-44.
9. Matsumoto Y, Fujii H, Itakura J, Matsuda M, Yang Y, Nobukawa B, *et al*. Pancreaticobiliary maljunction: Pathophysiological and clinical aspects and the impact on biliary carcinogenesis. Langenbecks Arch Surg 2003;388:122-31.
10. Song HK, Kim MH, Myung SJ, Lee SK, Kim HJ, Yoo KS, *et al.* Choledochal cyst associated the with anomalous union of pancreaticobiliary duct (AUPBD) has a more grave clinical course than choledochal cyst alone. Korean J Intern Med 1999;14:1-8.
11. Okoromah CN, Odelola MA, Etukudo FZ. Complicated massive choledochal cyst: A case report. Niger J Clin Pract 2002;5:150-2.
12. Gall BM, Madziga AG, Hamid AU. Choledochal cyst: A case report and review of literature. Niger J Med 2004;13:295-9.
13. Dabbas N, Davenport M. Congenital choledochal malformation: not just a problem for children. Annals of The Royal College of Surgeons of England 2009;91:100-5.
14. Douglas AH. Case of dilation of common bile duct. Mon J Med Sci (Lond) 1852;14:97-101.
15. Alonso-Lej F, Rever WB Jr, Pessagno DJ. Congenital choledochal cyst, with a report of 2, and an analysis of 94, cases. Int Abstr Surg 1959;108:1-30.
16. Nagorney DM. Bile duct cysts in adults. In: Blumgart LH, Fong Y, editors. Surgery of the Liver and Biliary Tract. 3rd ed., Vol. 2. London: WB Saunders; 2000. p. 1229-44.
17. Melton GB, Lillemoe KD. Choledochal cysts. In: Zinner MJ, Ashley SW, editors. Maingot's Abdominal Operations. 11th ed. New Delhi: McGraw Hill; 2009. p. 889-97.
18. Shi LB, Peng SY, Meng XK, Peng CH, Liu YB, Chen XP, *et al.* Diagnosis and treatment of congenital choledochal cyst: 20 years' experience in China. World J Gastroenterol 2001;7:732-4.
19. Sela-Herman S, Scharschmidt BF. Choledochal cyst, a disease for all ages. Lancet 1996;347:779.
20. Liu CL, Fan ST, Lo CM, Lam CM, Poon RT, Wong J. Choledochal cysts in adults. Arch Surg 2002;137:465-8.
21. Sheridan MB. Endoscopic retrograde cholangiopancreatography should no longer be used as a diagnostic test: The case in favour. Dig Liver Dis 2002;34:370-4.
22. Lipsett PA, Pitt HA, Colombani PM, Boitnott JK, Cameron JL. Choledochal cyst disease. A changing pattern of presentation. Ann Surg 1994;220:644-52.
23. Atkinson H, Fischer C, de Jong C, Madhavan KK, Parks RW, Garden OJ. Choledochal cysts in adults and their complications. HPB Surg 2003;5:105-10.
24. Jordan PH Jr, Goss JA Jr, Rosenberg WR, Woods KL. Some considerations for management of choledochal cysts. Am J Surg 2004;187:434-9.
25. Irie H, Honda H, Jimi M, Yokohata K, Chijiiwa K, Kuroiwa T, *et al.* Value of MR cholangiopancreatography in evaluating choledochal cysts. AJR Am J Roentgenol 1998;171:1381-5.
26. Sugiyama M, Haradome H, Takahara T, Abe N, Tokuhara M, Masaki T, *et al.* Anomalous pancreaticobiliary junction shown on multidetector CT. AJR Am J Roentgenol 2003;180:173-5.
27. Nino-Murcia M, Jeffrey RB Jr, Beaulieu CF, Li KC, Rubin GD. Multidetector CT of the pancreas and bile duct system: Value of curved planar reformations. AJR Am J Roentgenol 2001;176:689-93.
28. Lipsett PA, Pitt HA. Surgical treatment of choledochal cysts. J Hepatobiliary Pancreat Surg 2003;10:352-9.
29. Watanabe Y, Toki A, Todani T. Bile duct cancer developed after cyst excision for choledochal cyst. J Hepatobiliary Pancreat Surg 1999;6:207-12.
30. Malik IA, Aziz Z, Zaidi SH, Sethuraman G. Gemcitabine and cisplatin is a highly effective combination chemotherapy in patients with advanced cancer of the gallbladder. Am J Clin Oncol 2003;26:174-7.