Choledochal Cyst Disease in Adults: 14-year Experience with 204 Patients in a Single Institution

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Purpose: We present our 14-year experience with 204 adult patients with choledochal cyst disease (CCD), focusing on clinical outcomes after surgical treatment.

Materials and Methods: The medical records of 204 adult patients (≥18 years old) treated for CCD at the Asan Medical Center from July 1995 to June 2009 were reviewed retrospectively.

Results: Median patient age was 40.2 years and 157 (77%) were female. Abdominal pain was the most common symptom. Eleven patients (5.4%) had undergone prior biliary surgery. Todani classification was type I in 116 patients (56.9%), type II in 1 (0.5%), type IVa in 86 (42.2%) and type V in 1 (0.5%). Extrahepatic cyst excision and hepaticojejunostomy were performed in 185 patients (90.7%). Major perioperative complications occurred in 6 patients (2.9%), resulting in no mortality. Late complications occurred in 48 patients (23.6%): intrahepatic duct stone formation in 25 (14.4%), recurrent cholangitis in 31 (17.8%), overt anastomotic stricture requiring treatment in 11 (6.3%), and recurrent pancreatitis with or without pancreatic duct stone in 10 (5.7%). Overt late stenosis at the hepaticojejunostomy was detected in 11 patients; 6 underwent radiological intervention with or without cholangioscopy, 2 underwent surgical revision, 1 underwent left hepatectomy, and 2 were closely observed while receiving supportive care. Refined surgical techniques reduced the incidences of anastomotic stricture and recurrent pancreatitis. No recurrent pancreatitis occurred in patients who received complete removal of the intrapancreatic cyst.

Concurrent cancer was diagnosed in 20 patients (9.8%). Anomalou s union of the pancreaticobiliary duct was more frequently associated with gallbladder cancer than with bile duct cancer. Two of the 184 patients (1.1%) without biliary cancer at the time of surgery developed de novo malignancies at the remnant cyst within the pancreatic parenchyma, 6 months and 13 months, respectively, after cyst excision and hepaticojejunostomy. The survival outcomes in CCD patients with concurrent biliary cancer were comparable to those in cancer patients without CCD.

Conclusions: Due to diverse features of CCDs, various clinical problems can occur after surgical excision in adult patients. Surgical treatment for patients with CCDs having complex features should be individualized to maintain a balance between the risk of surgery and the potential risk of late complications. We emphasize the importance of well-experienced surgeons teaching less-experienced hands in performing surgery for other than type I CCD.