A 55-year-old woman had been admitted to our hospital because of the swelling above the ampulla of Vater and referred to our department for further examination. No abnormality was found on physical examination or in laboratory data. However, duodenoscopy revealed a soft cystic lesion above the ampulla of Vater (**A**; *left*). EUS showed that the common bile duct and the main pancreatic duct communicated with the cyst located in the duodenal wall (**A**; *right*). The diagnosis was choledochocele. Ten years after the diagnosis of choledochocele, she was readmitted for abdominal pain, at age 65. Her serum CA19-9 was 53.4 U/mL (normal <37 U/mL). Duodenoscopy showed the enlarged choledochocele (**B**; *left*). EUS revealed an echogenic mass in the choledochocele (**B**; *right, arrowheads*). An incision was made to the roof of the choledochocele, and peroral cholangioscopy revealed villous tumor in the choledochocele. Biopsy of the tumor revealed villous adenoma. She underwent local resection of the ampulla of Vater. Microscopic examination revealed villous adenoma with moderate to severe dysplasia (**C**; *left* [H&E, orig. mag. × 40], *right* [H&E, orig. mag. × 400]). The postoperative course was uneventful. In 5-years follow-up, no clinical evidence of reccurence is found.