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Large Cell Neuroendocrine Carcinoma Of The Extrahepatic Bile Duct: A Case Report Of Two Cases

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Background : Neuroendocrine carcinoma originating from extra hepatic bile duct is very rare and only a few cases have been reported. Because of its scarcity of incidence, not much is known about the disease but for its aggressiveness and poor prognosis. Herein we report two cases of large cell neuroendocrine carcinoma (LCNEC) originating from extrahepatic bile duct (hilum and common bile duct (CBD)).

Methods : CASE 1. A 60–years–old woman was preoperatively diagnosed a perihilar cholangiocarcinoma, and a left hepatectomy and caudectomy with hepaticojejunostomy was performed. From the histopathological findings, we diagnosed the tumor as a LCNEC (pT2aN1Mx, pStage IIIB) with focal proportion of an adenocarcinoma component. The postoperative course was uneventful, and she was administered etoposide and cisplatin every 3 weeks (6th cycles) as an adjuvant chemotherapy. She has remained recurrence–free for 7 months.

Results : CASE 2. A 67–years–old man was diagnosed a cholangiocarcinoma of mid–CBD and underwent laparoscopic pylorus–preserving pancreatoduodenectomy (PPPD). The pathological findings showed a LCNEC (pT1N1Mx, pStage IIB) with focal proportion of an adenocarcinoma component in the extrahepatic bile duct with lymph node metastases. After recovery, he was administered etoposide and cisplatin every 3 weeks (currently 6th cycle) as an adjuvant chemotherapy. At 7 months after surgery, there was no recurrence of the disease.

Conclusions : Neuroendocrine carcinoma of the extrahepatic biliary tracts is a very rare and highly malignant disease with a poor prognosis. To establish the definite treatment approach, more cases should be found and reviewed. Until then, a multidisciplinary approach could improve the prognosis for this neoplasm.

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