Klatskin Tumors

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Editorial

Carcinoma of the hepatic duct bifurcation was first described by Altemeir in 1957. Subsequently, a series of 13 patients with this diagnosis was reported by Klatskin in 1965. Since that time, hilar cholangiocarcinomas at this location have carried the eponym of Klatskin tumors. Cholangiocarcinoma accounts for less than 2% of all human malignancies but is the second most common primary liver tumor [1]. Hilar Cholangiocarcinoma (HC) comprises up to 50–70 % of all cases of bile duct carcinomas. Approximately 3–5 new cases per 100,000 are diagnosed annually in Europe, but the incidence and mortality are increasing worldwide. HC is approximately 3,000 cases annually in the United States alone [2]. The absence of early symptoms leads to the diagnosis of most Klatskin tumors at an advanced incurable stage. Only painless obstructive jaundice indicates rarely earlier stages, but it is nevertheless often a sign of advanced disease. There has been a growing recognition that hilar vs. distal cholangiocarcinoma have a distinct biological behaviour and natural history, as well as a therapeutic treatment strategy. This has led the latest (seventh) edition of the American Joint Committee on Cancer (AJCC) Cancer Staging Manual to divide perihilar and distal cholangiocarcinoma into distinct staging subgroups. Less than one half of HC are resectable. A variety of risk factors have been associated with HC, most notably primary sclerosing cholangitis, biliary stone disease and parasitic liver disease. Patients typically present with abdominal pain, pruritis, weight loss, and jaundice. Computed topography, magnetic resonance imaging, and ultrasound are used to characterize biliary lesions. Endoscopic retrograde cholangiopancreatography and percutaneous transhepatic cholangiography assess local ductal extent of the tumor while allowing for therapeutic biliary drainage. Magnetic resonance cholangiopancreatography has demonstrated similar efficacies to transhepatic cholangiography and retrograde cholangiopancreatography in identifying anatomic extension of tumors with less complications. Serum tumor markers, specifically carcinoembryonic antigen and CA19-9, are used for the diagnosis, treatment, and monitoring of HC with 89% sensitivity and 86% specificity when combined with other diagnostic modalities. Over 90% of extra hepatic epithelial bile duct tumors are adenocarcinomas. Treatment consists of surgery, radiation, chemotherapy and photodynamic therapy. Biliary drainage of the future liver remnant should be performed to decrease bilirubin levels thereby facilitating future liver hypertrophy. Standard therapy consists of surgical margin-negative (R0) resection with extra hepatic bile duct resection, hepatectomy and en bloc lymphadenectomy. Local resection should not be undertaken. Lymph node invasion, tumor grade and negative margins are important prognostic indicators. In instances where curative resection is not possible, liver transplantation has demonstrated acceptable outcomes in highly selected patients. Exclusion criteria include patients with a mass lesion below the level of cystic duct, tumors greater than 3 cm, evidence of intrahepatic or extra hepatic metastases, or previous history of transperitoneal biopsy. Despite the limited data, chemotherapy is indicated for patients with unresectable tumors and adequate functional status. Criteria for unresectability include bilateral spread to secondary biliary radicals, involvement of the portal vein main trunk, bilobar involvement of hepatic arterial and/or portal venous branches, and unilateral hepatic artery involvement with evidence of extensive contralateral duct spread. Five-year survival after surgical resection of HC ranges from 10% to 40% however, recurrence can be as high as 50-70% even after R0 resection. Due to the complexity of this disease, a multi-disciplinary approach with multimodal treatment is recommended for this complex disease.

References