Malignant tumours of the bile duct

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Summary box 71.7 Bile duct cancer (cholangiocarcinoma) /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Cholangiocarcinoma Incidence Cholangiocarcinoma is an uncommon malignancy . The overall annual incidence is 1–1.5 per 100 /uni00A0 000 with the peak incidence in the eighth decade. The male-to-female ratio is approximately 1.5:1. Anatomically , tumours involving the biliary confluence (hilar cholangiocarcinoma or Klatskin tumours) account for 60% of cases, with the remainder involving the distal bile (20–30%) or intrahepatic ducts (10–20%). Risk factors A minority of patients have a known risk factor; the major risk factor in western practice is PSC. It is estimated that a longstanding history of PSC increases the risk of developing biliary tract cancer 20-fold compared with the normal popu lation; those with concomitant IBD are at significantly higher risk. Cholangiocarcinoma appears to occur at an earlier age in patients with PSC (30–50 years of age) than in the general population. In addition, disease is usually multifocal and detected at an advanced stage with a resultant poor prognosis compared with the general population. Congenital cystic disease, he patolithiasis, oriental cholan giohepatitis, hepatitis C virus infection and infestation with liver flukes have also been associated with an increased risk of cholangiocarcinoma. While the pathophysiology is unclear, it is thought that these parasites cause c hronic inflammation that leads to DNA mutations through production of carcinogens and free radicals; the latter can stimulate cellular proliferation in the intrahepatic bile ducts and ultimately lead to invasive cancer (Summary box 71.8). Gerald Klatskin , 1910–1986, hepatologist, V A Hospital, Newington, CT , USA. Early symptoms are often non-specific, with abdominal pain, early satiety , anorexia and weight loss commonly seen. Symptoms associated with biliary obstruction (pruritus and jaundice) may be present in a minority of patients. In these patients, examination often demonstrates clinical signs of jaundice, cachexia is often noticeable and the gallbladder is palpable if the obstruction is in the distal CBD (Courvoisier’s sign). Summary box 71.8 Risk factors for cholangiocarcinoma /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Investigations Biochemical investigations (elevated bilirubin, ALP and GGTP) will confirm obstructive jaundice. The tumour marker carbohydrate 19-9 (CA19-9) may also be elevated. Imaging studies such as USG, CT and MRI/MRCP are essential for duct diagnosis, staging and assessing the anatomical relationship between the tumour and the major perihilar vascular struc - tures (Figure 71.38). These studies allow the level of biliary obstruction to be defined and determine the locoregional extent of disease and the presence of metastases. Direct cholangiography using ERCP or PTC is also used following non-invasive studies. Both can define the level of obstruction - and allow biopsy (non-diagnostic in 40–80% of patients) and placement of endobiliary stents for biliary drainage. The choice between the modalities depends on local availability and the anatomical site of the tumour, with PTC preferred for more proximal lesions and ERCP favoured for distal tumours. Despite a higher incidence of postoperative infection

with stenting, patients undergoing anticipated major liver resection - need preoperative biliary drainage as hyperbilirubinaemia (bilirubin level >6 /uni00A0 mg/dL) may impair hepatic regeneration and hypertrophy . Cholangioscopy is an adjunct to ERCP , with diagnostic accuracy increased from 78% to 93% and sensitivity from 58% to 100%. Positron emission tomography (PET) is useful in detecting lymph node and distant metastases but has limited value in the assessment of local resectability .

Malignancy arising from the biliary epithelium; histologically of three types: mass forming, intraductal growing and periductal in /f_i ltrating Rare, but incidence increasing Most patients present with abnormal liver function tests or frank jaundice Diagnosis by USG, CT or MRI Majority of patients receive palliative care only Complete surgical excision possible in <10% Prognosis poor: 90% die within 1 year from liver failure or biliary sepsis Adjuvant chemoradiation therapy has a limited role Chronic in /f_l ammatory Chemical agents conditions Thorium dioxide (Thorotrast) PSC Vinyl chloride Oriental cholangiohepatitis Dioxin Hepatitis C infection Asbestos Parasitic infections Post surgical Opisthorchis viverrini Biliary–enteric anastomosis C. sinensis Non-alcoholic fatty liver disease Congenital Choledochal cysts Caroli’s disease

The anatomical extent of the disease is classified according to either the Bismuth–Corlette (Figure 71.39) or the Memorial Sloan Kettering Cancer Center (MSKCC) classification. The MSKCC classification T-stage criteria for hilar chol angiocarcinoma are as follows: /uni25CF T1 tumour involving the biliary confluence without exten sion to second-order biliary radicles. /uni25CF T2 tumour involving the biliary confluence with unilateral extension to second-order biliary radicles and ipsilateral portal vein involvement or ipsilateral hepatic atrophy . /uni25CF T3 tumour involving the biliary confluence with bilateral extension to second-order biliary radicles; or unilateral Henri Bismuth , b. 1934, surgeon, Hôpital Paul Bruce, Villejif, France, and Allen Oldfather Whipple , 1881–1963, surgeon, Columbia-Presbyterian Medical Center, New Y ork, NY , USA. Mayo Clinic , Rochester, MN, USA, established in the 1880s by Dr William Worral Mayo and his sons William and Charles, both surgeons. lateral portal vein involvement; or unilateral exten - sion to second-order biliary radicles with contralateral hepatic lobar atrophy; or main or bilateral portal venous involvement. Treatment A multidisciplinary approach is required. The choice of treatment depends on the site and extent of the disease. Unfortunately , the majority of patients present with advanced disease. However, 10–15% are suitable for surgical resection, which o ff ers the only hope for long-term survival. The aim of surgical resection is to achieve complete resection with negative pathological margins (R0 resection) and safely restore biliary–enteric continuity . Whether or not the disease is resectable depends on patient factors (comorbidities, presence or absence of chronic liver dis - ease) and tumour factors (extent of disease within the biliary tree, vascular involv ement, remnant liver volume, increase in the remnant after portal vein embolisation, presence or absence of metastatic disease). De pending on the site of disease, sur - gery involves either a standard or extended hepatic resection with caudate lobe excision with en bloc lymphadenectomy and reconstruction of the biliary tree. Distal common duct tumours - may require pancreaticoduodenectomy (Whipple pr ocedure). Local resection should be avoided. In selected patients, liver transplantation has been recom - - mended for those with locally unresectable disease without evi - dence of distant metastases. Transplantation is often preceded by neoadjuvant chemoradiation therapy and staging laparos - copy . While emerging data are encouraging, this aggressive appr oach remains controversial and is reserved for selected patients

in specialised centres (Mayo protocol). Marvin Corlette described the classification of cholangiocarcinoma in 1975.

Figure 71.38 Magnetic resonance imaging scan showing a hilar chol angiocarcinoma with dilatation of the intrahepatic biliary tree. Right hepatic duct Common hepatic duct Figure 71.39 Bismuth–Corlette classi /f_i cation of cholangiocarcinoma.

Left hepatic duct Type I Type II Type IIIA Type IIIB Type IV

the T-stage, margin status, metastatic lymph node spread, perineural and perivascular invasion, non-papillary tumour subtypes and poor tumour di ff erentiation. Of these, the only variable in which the surgeon plays a major role is margin-negative resection, and emphasis needs to be placed on achieving R0 resection. Approximately 35% of patients will survive 5 years after surgery . Adjuvant chemotherapy and radiotherapy have a limited role and have not been demonstrated to add survival benefit following surgical resection. However, patients at high risk for recurrence (positive surgical margins or node positive) may benefit from adjuvant therapy and should be referred for medical or radiation oncology opinion. The majority of patients who present with unresectable disease are candidates for palliative chemotherapy – gemcitabine with cisplatin. The aim is to maintain or improve quality of life by relieving symptoms and preventing cholestatic liver failure. Biliary obstruction can be relieved by endoscopic (ERCP) or percutaneous (PTC) methods. Surgical bypass rarely has a role apart from in patients with a distal bile duct lesion found to have unresectable disease at operation.

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