Intrahepatic cholangiocarcinoma

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Cholangiocarcinomas develop in the bile duct and demonstrate considerable variation in origin, behaviour and pathophysiol ogy . Twenty per cent of cholangiocarcinomas are intrahepatic (ICC), representing 5–30% of primary liver tumours, which is second only to HCC. The y are rare in the West with an incidence of 0.5–2/100 /uni00A0 000, although the incidence is increasing and is significantly higher in South East Asia, where Clonorchis sinensis infection is endemic, and in Thailand the rate is 60/100 /uni00A0 000. There are three morphological subtypes – infiltrating periductal (Figure 69.23a), mass-forming (Figure 69.23b) and intraductal – and ICC is now considered to have multiple cellular origins. Ultrasonography will confirm intrahepatic biliary obstruction and MRCP is prefer red for diagnosis. ERCP will delineate tumour extent and with brush TM cytology or SpyGlass can achieve pathological confirmation of malignancy . ICC was originally staged with HCC but is now classified separately . Prognostic pathological features include vascular invasion, tumour multiplicity , local extension, periductal infil tration and lymph node metastasis. CT scanning is the best staging modality to identify distant metastases and confirm tha the lesion is primary and not metastatic. Angiography is some times required preoperatively to assess vascular involvement. The sensitivity and specificity of PET-CT for diagnosis of cholangiocarcinoma varies by location, being higher for intra hepatic (>90%) than for extrahepatic (60%) tumour s, although detection rates for distant metastases approach 100%. ICC is an aggressive tumour and, even when confined to the liver, only 30% of patients are suitable for resection at the time of presentation. If surgical resection is considered, biopsy should be av oided; in borderline cases diagnostic laparoscopy and intraoperative ultrasonography will exclude additional hepatic or peritoneal disease. Lymph node status (porta hepa tis, common hepatic artery and the gastroduodenal ligament) remains an important prognostic factor and should be sam pled. One-year survival rates have improved to 25%, although Vincenzo Mazzaferro , b. 1957, surgeon, Istituto Nazionale dei Tumori, Milan, Italy . - - - 5-year survival of 3% remains unchanged. Unfortunately , conventional chemotherapy o ff ers limited survival benefit for unresectable or metastatic disease. -

Figure 69.23 Hilar cholangiocarcinoma (Klatskin

tumour) demonstrat

ing tight stricture and intrahepatic dilatation of intrahepatic ducts due to in /f_i ltrating tumour (a) and an apparent space-occupying lesion due to the mass-forming variety (b) .

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