tation of the liver (with Clonorchis sinesis or Opisthorchis viverrini), following Thorotrast administration or anabolic steroid therapy or in association with intrahepatic lithiasis. The age of diagnosis has ranged from 23 to over 90 years, and the median age has varied from 59 to 69 years. Clinically, these tumors usually cause early obstruction resulting in jaundice, which can progress or fluctuate. Other symptoms include right upper quadrant pain, malaise, weight loss, pruritus, anorexia, nausea, and vomiting. Less than 20% of these tumors are associated with choledocholithiasis. In fact, the association of extrahepatic bile duct carcinoma with gallstones is no greater than expected from the prevalence of gallstones in the general population.

For prognostic and therapeutic purposes, it is useful to divide the extrahepatic ductal system into 3 parts: upper, middle, and lower thirds. Almost 50% of all carcinomas are located in the upper third, 25% in the middle third, and 19% in the lower third. In less than 10%, it is not possible to specify the origin because of diffuse involvement of the bile ducts.[2]

Grossly, 3 patterns of growth are described: polypoid, sclerosing, and infiltrative. Most carcinomas of the extrahepatic ducts appear as poorly defined, gray-white tumors with thickening of the bile duct wall and narrowing of the lumen, resembling fibrous strictures or sclerosing cholangitis radiographically.[2] They tend to grow slowly, infiltrate the wall of the ducts, and dissect along tissue planes. Perineural invasion and metastasis to regional lymph nodes are common.

Problems in differential diagnosis of bile duct cancer are more common than that of gallbladder cancer. Carcinoma of the extrahepatic and intrahepatic biliary ducts can extend to the liver, pancreas, ampulla, duodenum, or gallbladder. Since all of these organs give rise to malignant epithelial neoplasms similar to those originating in the bile ducts, identification of their exact origin may be difficult, especially with large cancers. In many patients, a diagnosis can only be reached after careful clinicopathologic correlation, including a review of the radiographic and operative findings. Also, the pathologist should recall that the clinical presentation and biologic behavior vary according to the origin of the tumor. Nevertheless, in some cases it may be impossible to specify with certainty the origin of the tumor. This uncertainty has led to the use of nonspecific terms such as carcinoma of the periampullary region or hilar carcinoma of the liver, which reflect the difficulties involved in locating the primary site. Immunohistochemically there are consistent reactivities for keratin, EMA, and CEA, features of importance in the different diagnosis of liver carcinomas.

Pathological Diagnosis

Cholangiocarcinoma, hilar type, measuring 7 × 6 × 4 cm, with multiple intrahepatic metastatic nodules, status post stent insertion and radiotherapy, with direct invasion to the diaphragm, first portion of duodenum, antrum and low body of stomach, and with metastases to bilateral lungs, bilateral pleura, hilum of spleen, body of pancreas, small and large intestines, omentum, mesentery, and rectus abdominis muscle.

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