Parasitic cysts, hydatid disease

Hydatid cysts, the most common parasitic cysts of the liver, are caused by the cestode Echinococcus granulosus. Hydatid disease is prevalent in South America, the Far East, Southern and Eastern Europe, and Australia. This disease is rare in the United States, but may occur in Alaska and in the southern United States. Echinococcus granulosus is disseminated when the offal of infected sheep or swine is fed to dogs. The dogs’ feces contaminate the ground or water around dwellings, and humans ingest the ova. After digestion, the ova unencyst to pass through the intestinal epithelium into the portal blood and thence to the liver. Although other organs including the lung can be involved, the liver is the most commonly affected site. Sixty to seventy percent of cysts arise in the right lobe, about 20 percent arise in the left lobe, and in 20 to 30 percent of cases, both lobes are diseased. Echinococcal cyst of the liver is a chronic disease that affects both sexes equally and may occur at any age. The cyst may be asymptomatic for long periods and may be detected incidentally. Some patients may experience abdominal discomfort or may detect a mass. Approximately one-third of the patients experience the following complications in a decreasing order of frequency: rupture into the bile ducts, (2) abscess, (3) intraperitoneal rupture, and (4) hepatobronchial fistula. Rupture of echinococcal cyst into a bile duct causes colicky abdominal pain, jaundice, and fever. Plain films of the abdomen may reveal calcification in the cyst wall. Ultrasound is probably the best test for the diagnosis and delineation of an echinococcal cyst. One author stated that the ultrasound findings of the cyst with demonstrable daughter cysts was pathognomonic. CT scans delineate an echinococcal cyst with 98 percent accuracy. False-negatives occur in patients with fatty livers. Immunologic testing may be employed, in one study, the intradermal skin tests (Casoni test) were 69 percent positive; however, some authorities recommend that the Casoni test be abandoned. Immunoelectrophoresis is probably the test of choice. Echinococcal cysts require surgical treatment. Several authors recommend evacuation of the cyst and the application of a sclerosing agent (either 0.5% silver nitrate or 1% cetramide). Formalin should not be used because it can cause bile duct sclerosis or death. When cysts rupture into the bile ducts, common duct exploration and choledochostomy should be performed. There is controversy concerning the treatment of echinococcal cysts. Some recommend excision of the cyst; others advise hepatic lobectomy. Resection must undoubtedly be required on occasion. The cyst fluid is antigenic, and intra-abdominal leakage of the fluid may cause anaphylactic shock.

Liver tumor

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With widespread application of sophisticated radiologic (CT scan, ultrasonography, and angiography) and chemical (CEA and alpha-fetoprotein) diagnostic methods, liver tumors are being detected more often and more accurately than before. Although asymptomatic mass lesions, found incidentally by radiologic examination, are more often benign than malignant, they should be considered as the latter until proved otherwise.

Some benign and malignant tumors have characteristic radiologic features, but the findings are by no means pathognomonic. A small piece of tissue obtained by needle biopsy is often inadequate to establish a definitive diagnosis, particularly in differentiating adenoma from low-grade hepatocellular carcinoma. If the diagnosis is uncertain, the lesion should be excised without delay with an adequate margin. The high mortality following major hepatic resections which existed in the past has been minimized in recent years at many major centers. At our institution, the operative mortality has been less than 3 percent.

In planning how large a resection will be required, a CT scan is most useful to assess the extent of the tumor, but it can be misleading when a large tumor distorts normal anatomic boundaries. If resectability is uncertain after extensive preoperative investigations, the examining physician should refer the patient to a surgeon who is experienced in major hepatic resections instead of subjecting him or her to exploratory celiotomy by someone who is unprepared to undertake a definitive procedure.

Benign liver tumors

Cavernous hemangioma

Hemangiomas are the most common benign tumors of the liver. They are usually small (less than 4 cm in
and solitary. Occasionally, they are, or grow very large. By convention, lesions larger than 4 cm are called giant hemangiomas. Most hemangiomas are called giant hemangiomas and are found incidentally. However, giant hemangiomas can cause various disabling pressure symptoms and pain. Life-threatening spontaneous rupture of a giant hemangioma is uncommon, but this complication has been reported many times. The usual cause of a massive hemorrhage is an ill-advised percutaneous needle biopsy.

Symptomatic cavernous hemangiomas should be excised surgically. The majority of giant hemangiomas resected en bloc or trisegmentectomy, but some, that are located on the surface of the liver or those that are trisected, can be enucleated along pseudocapsular planes without significant loss of normal tissue. We have treated 60 giant cavernous hemangiomas without any complications. An accurate preoperative diagnosis can be made with certainty. However, a needle biopsy usually confirms the diagnosis because of the characteristic histology. Rare large focal nodular hyperplasias cause disabling pressure symptoms and pain, necessitating hepatic resection.

Cysts

Congenital simple cysts of the liver that have an endothelial-like lining have been referred to as "spring-water cysts." They are usually asymptomatic and can safely be observed.

Simple cysts lined with cuboidal epithelium sometimes become large and multilocular. The lining may become fibrous, proliferative, papillary, or mucin-producing. They often communicate with the biliary system, bleed internally, or become infected. These symptomatic congenital cysts should be treated surgically. Although it is tempting to aspirate or drain the cystic fluid percutaneously, those procedures are often diagnostically equivocal and therapeutically ineffective, and may involve risks of introducing infection and of implantation of neoplastic cells or parasites, if used unwisely. Some advocate that the solitary cysts with clear fluid be unroofed and drained into the peritoneal cavity, and that cysts with bile-stained fluid be drained with a Roux-en-Y jejunal limb. We recommend resection of all symptomatic cysts, whenever possible. They can be locally excised or removed by anatomic resection. Although malignant change in the cyst wall is uncommon, we have treated three patients whose cyst lining developed squamous cell carcinoma; two of the three had been previously operated on with Roux-en-Y internal drainage techniques.

A cystadenoma of the liver should be totally excised because of the possibility of malignant degeneration and because it may not be distinguishable histologically from a cystadenocarcinoma. Small lesions may be excised locally with adequate margins, but larger ones should be removed by anatomic resections.

The pressure symptoms of polycystic disease of the liver can sometimes be palliated with multiple needle aspirations. Operative marsupialization, incisional drainage, or aspiration of large cysts cannot relieve the symptoms more effectively than simple percutaneous needle aspirations. Therefore, operative procedures for polycystic disease are rarely justified. However, there are occasional patients who have a dominance of normal tissue in the left lateral segment of the liver. We have treated two such patients by right trisegmentectomy and the result was prolonged symptomatic relief. We have used orthotopic liver transplantation to treat a woman whose huge liver caused uncontrollable pain and multiple rib fractures. She has been pain-free and off narcotics since the transplantation.

Other Benign Lesions

Rare benign tumors, such as fibroma, rhabdomyoma, leiomyoma, fibrous mesothelioma, and myelolipoma, can—
not be easily distinguished from malignant tumors without pathologic examination. If the diagnosis is uncertain, the tumor should be excised with an adequate margin for detailed histologic examination.

MALIGNANT TUMORS

Primary Hepatic Malignant Tumors

Various types of primary malignant tumors develop in the liver; hepatocellular carcinoma (hepatoma) is the most common, followed by cholangiocarcinoma, hepatoblastoma, and various cell types of sarcoma. As long as the lesion is localized in three of the four segments of the liver, curative subtotal hepatectomy theoretically can be performed. Many hepatomas found in noncirrhotic livers grow slowly, and even very large tumors sometimes spare one segment of the normal liver. Results after major hepatic resections justify the efforts, because the 5-year survival rate is 40 to 50 percent for noncirrhotic patients.

However, hepatoma developing in a cirrhotic liver is a different therapeutic problem. The patients with well-compensated cirrhosis (no jaundice, no ascites, and serum albumin greater than 3 mg per deciliter) can usually tolerate a lobectomy, but the operative mortality is as high as 30 percent. Both the progressive nature of underlying cirrhosis and the multiplicity of malignant tumors in cirrhotic livers contribute to a grim prognosis.

The role of orthotopic liver transplantation for primary malignant tumors of the liver is limited. Our experience with 50 such patients, as well as the experience of others, has shown that primary malignant tumors of the liver that cannot be removed by conventional subtotal hepatectomy cannot be cured by total hepatectomy and liver replacement. The tumors have almost always returned if the patients lived long enough. However, good palliation has been achieved for many patients for a year or two.

In contrast, cure usually has been achieved of tumors found incidentally in livers removed for other end-stage hepatic disease.

Metastatic Tumors

The liver is one of the organs most commonly involved by metastatic tumors. An aggressive approach with the resection of hepatic metastases is warranted because long survival has been regularly achieved, particularly when isolated or regional hepatic metastases have been from colorectal primaries. Our usual approach to metastatic liver tumors has been with anatomic resections rather than excisions. We have performed more than 80 major anatomic hepatic resections for metastatic tumors without any mortality and have found unexpected small additional metastases in the resected specimens in nearly 10 percent of the cases. The large size of the lesion, a multiplicity of metastases, or a short interval between resection of the primary and appearance of secondary lesions can adversely affect the survival rate after resection, but not to let any of these factors dissuade us from proceeding. Five-year survival after resection among our 50 patients with metastatic colorectal cancer is approximately 50 percent.

TECHNICAL REFINEMENTS IN HEPATIC RESECTION

Incision

A bilateral subcostal incision with an upper midline extension usually gives adequate exposure for any type of resection, especially if the xiphoid process is removed. In many cases the left subcostal extension is not necessary, particularly for right-sided resection. If a thoracic extension is decided upon, a right seventh intercostal incision is connected to the midportion of the right subcostal incision. The thoracic extension almost never is required, even in physically well-developed adults.

Right Lobectomy and Right Trisegmentectomy

The steps of the operation may vary, depending on the location and size of the lesion. Usually the right triangular ligaments are incised, and the bare area is entered. This allows the right lobe to be lifted into the wound and retracted to the left. Inability to mobilize the liver safely at this time and difficulty in visualizing the area where the right hepatic vein enters the inferior vena cava are the main reasons for considering a thoracic extension.

Before the hilar dissection is begun, anatomic variations of the hepatic artery must be looked for. In the "normal" situation, the common hepatic artery originates from the celiac axis and lies in the left anterior portion of the portal triad. However, the right hepatic or even common hepatic artery may originate from the superior mesenteric artery, and if so, the anomalous vessel will lie posterior to the portal vein. The left hepatic artery can originate from the left gastric artery or separately from the aorta; in which case it will be found in the middle of the gastrohepatic ligament, running toward the umbilical fissure.

The hilar dissection is begun by ligation and division of the cystic duct and artery. The right branches of the structures of the portal triad are isolated. The right hepatic artery is sacrificed first. Arterial anomalies are so numerous that ligation should never be performed without preliminary test occlusion and without being sure that during this occlusion there are pulsations distally in the region of the umbilical fissure.

At a more superior level, the right branch of the portal vein is detached and the stump is tied or closed with sutures. After dividing the right hepatic artery and the right portal vein, a line of demarcation is evident between

A
Almost invariably, the hepatic bile duct is the hilar structure with the most superior bifurcation, and sometimes the division is within the substance of the liver. The duct is ligated and divided where it comes off like the crossbar of a T. Dissection of the hilum is completed for a true right lobectomy.

If the right lobe is retracted, dissection and encirclement of the right hepatic vein is now begun. The vein is doubly clamped with angled vascular clamps, divided, and ligated at their origin. If this left portion of the caudate lobe is to be spared, the ligatures usually should be distal to the posteriorly directed first branch. When those maneuvers are completed, the true left lobe becomes cyanotic. With continuous traction of the left lateral segment anteriorly and to the right, the posterior incision of the eventual specimen is developed through the liver capsule. If a decision has been made to preserve the left part of the caudate lobe, the parenchyma of the liver is entered along the natural line of the obliterated ductus venosus. Alternatively, the caudate lobe can be removed with consequent complete visualization of the anterior and left lateral surface of the retrohepatic vena cava. The elected specimen is developed through the liver capsule. If a decision has been made to preserve the left part of the caudate lobe, the parenchyma of the liver is entered along the natural line of the obliterated ductus venosus. Alternatively, the caudate lobe can be removed with consequent complete visualization of the anterior and left lateral surface of the retrohepatic vena cava. The elected posterior line of transection is continued superiorly until the left hepatic vein is encircled, transected, and either ligated or sewn closed with vascular sutures. Earlier attempts to encircle the left hepatic vein may be dangerous because of posteriorly located tributaries which can be thereby injured. At this point, a left lobectomy is completed by transecting the lobar plane defined by the color demarcation.

The additional requirement of a left hepatic trisegmentectomy is to remove the anterior segment of the right lobe. The main difficulty is to identify the correct intersegmental plane. There are two points at which this intersegmental plane can be properly entered. Some livers have a natural groove near the base of the gallbladder, which delineates the plane between the anterior and posterior segments. This groove, if present, is an excellent landmark. However, the search for this intersegmental plane is best begun superiorly, anterior to the right hepatic cuff. If only the left lateral segment is to be removed, the left hilar structures should be ligated at their origin. If this left portion of the caudate lobe is to be spared, the ligatures usually should be distal to the posteriorly directed first branch. When those maneuvers are completed, the true left lobe becomes cyanotic. With continuous traction of the left lateral segment anteriorly and to the right, the posterior incision of the eventual specimen is developed through the liver capsule. If a decision has been made to preserve the left part of the caudate lobe, the parenchyma of the liver is entered along the natural line of the obliterated ductus venosus. Alternatively, the caudate lobe can be removed with consequent complete visualization of the anterior and left lateral surface of the retrohepatic vena cava. The elected posterior line of transection is continued superiorly until the left hepatic vein is encircled, transected, and either ligated or sewn closed with vascular sutures. Earlier attempts to encircle the left hepatic vein may be dangerous because of posteriorly located tributaries which can be thereby injured. At this point, a left lobectomy is completed by transecting the lobar plane defined by the color demarcation.

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vein. With blunt dissection, the superior end of the previously defined line of posterior parenchymal incision is deepened near the diaphragm, using the point of the transected left hepatic vein as the starting point. The dissection finger is first brought anteriorly, then turned at right angles so that it can be swept transversely in the coronal plane. The fingertip emerges just anterior to the right hepatic vein. The middle hepatic vein is encountered and must be transected and ligated or sutured if this has not already been accomplished. Intraparenchymal tributaries to the right hepatic vein are ligated as encountered.

The superior to inferior scalping maneuver is continued, now aided by downward traction of the specimen. A resistant-free plane is sought, and all strands encountered are clamped or ligated. Inferiorly, the dissecting finger should emerge near the base of, and at right angles to, the gallbladder bed. Since the anterior segment retains its portal venous and hepatic arterial inflow almost until the specimen is out, the blood loss may be excessive. If so, the portal triad can be temporarily cross-clamped safely (the Pringle maneuver) for about one hour.

When the dominant natural transverse groove near the base of the gallbladder is present, the scalping process can be started at the hilum, ligating the arterial, portal venous, and ductal strands which run anteriorly, while protecting the vital residual posterior structures from injury.

The frontally presenting cut surface of the posterior segment after removal of the specimen permits precise visualization of residual bleeding joints and bile leaks. The extent to which ducts are exposed is greater than with other kinds of anatomic resections. An intraoperative cholangiogram through a T-tube should be obtained to check the integrity of the bile duct system before the closure.

**Drainage and Other Care**

The importance of adequate drainage of the subphrenic space after major hepatic resection cannot be overemphasized. Multiple closed-system suction drains can be placed in the huge, dead space, or open drains can be used with multiple one-inch Penrose drains. T-tube biliary drainage is not necessary unless the biliary system has been injured, except after left trisegmentectomy. After left trisegmentectomy, a T-tube should be left in with its upper limb passing into the remaining posterior segmental duct, which tends otherwise to become angulated.

Prophylactic antibiotics are started preoperatively and continued for a few days. Intraoperative correction of coagulopathy is important when a large amount of blood transfusion is required. Fear of hypoglycemia has been overemphasized. Usually, a maintenance infusion of 5% glucose and electrolyte solution is sufficient to maintain an adequate glucose level during and after operation. Transient jaundice and depression of multiple hepatic function tests are often seen after trisegmentectomy, but even after an 85 percent resection, relatively complete recovery can be expected within 1 to 3 weeks.

**HEPATIC ARTERY LIGATION AND ARTERIAL INFUSION CHEMOTHERAPY**

Hepatic artery ligation or radiologic embolization of hepatic artery has been used to treat nonresectable hepatic malignant tumors. The effects are usually temporary, and both procedures carry significant mortality and morbidity.

More recently, infusion chemotherapy through the hepatic artery by an implantable pump has been under investigation. Superiority of this hepatic artery infusion chemotherapy over conventional systemic intravenous chemotherapy is still uncertain. Various complications related to infusion catheter and implantable pumps have been reported at an incidence of 10 to 25 percent.

**COLORECTAL CANCER METASTATIC TO THE LIVER: RESECTION**

**MARTIN A. ADSON, M.D.**

Most of what has been learned clinically about the resective treatment of hepatic metastases has had to do with colorectal cancers because these primary lesions (1) are so common, (2) usually can be removed widely along with regional lymphatic spread, and (3) so often give rise to resectable hepatic metastases that appear to be the only sites of residual or recurrent growth. Also, most of what has been learned about metastases from colorectal cancers can be applied clinically to the management of other visceral cancers that (1) are well differentiated, (2) originate where primary and regional growth can be removed, and (3) can spread to the liver through the portal vein.

What is known about the effect of removing hepatic metastases from such lesions is surprising: about 25 percent of patients so treated survive for 5 years or more! Removal of such secondary sites of tumor growth is more effective than the resective treatment of some primary visceral cancers because of biologic and anatomic factors that are poorly understood. It is true that some other