Cystic Neoplasms of the Liver: Biliary Cystadenoma and Cystadenocarcinoma

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Cystic diseases of the liver occur in about 5% to 10% of the population. Although simple cysts are most common, biliary cystic tumors (BCTs), specifically biliary cystadenoma (BCA) and cystadenocarcinoma (BCAC), can arise in a subset of patients. Hueter first reported BCA in 1887 and Keen reported the first BCA resection 5 years later. Since that time, there has been a relative paucity of data published on the surgical management of BCTs. Because of the rarity of BCTs, many clinicians are unfamiliar with the diagnostic features, therapeutic management, and natural history of these liver neoplasms. We provide an evidence-based review of BCTs with particular emphasis on early recognition and approach to management. A search of available electronic databases, including MEDLINE/Pubmed, using the terms biliary cystadenoma, biliary cystadenocarcinoma, and non parasitic hepatic cysts was conducted. Additionally, we searched the MeSH database under the heading “Liver Neoplasm” in combination with the terms mentioned and Boolean operator AND or OR. Criteria for inclusion included English-language articles using human subjects (Fig. 1).

INCIDENCE AND EPIDEMIOLOGY

Biliary cystic tumors, such as BCA and BCAC, constitute <5% of all liver cysts. Although BCA occurs predominantly in females (90%), BCAC is more evenly distributed between males and females. Mean age at presentation of BCA is 45 years, and BCAC typically presents a decade later. Although 10% of reported BCTs originate in the extrahepatic biliary tree, the overwhelming majority arise from the intrahepatic biliary system. Biliary cystic tumors are typically slow-growing lesions with a reported size that can range in diameter from 1.5 to 35 cm.

Based on the mesenchymal stroma and the epithelium resembling endodermal (primitive hepatobiliary) cells that can be seen in BCTs, Wheeler and Edmondson hypothesized that BCAs arise from ectopic rests of embryonic bile ducts. However, 50% of BCTs contain endocrine cells, suggesting that they might originate from intrahepatic peribiliary glands. Biliary cystadenocarcinoma is thought to originate either de novo from formed biliary ducts induced by ischemia and carcinogens, or from malignant transformation of a pre-existing BCA. The latter theory is supported by the multiple reports of benign epithelium within BCAC specimens. For example, Devaney and colleagues reported malignant transformation in 6 of 18 BCACs in their series, as well as cytological atypia/dysplasia in a number of BCAs.

PRESENTATION AND DIFFERENTIAL DIAGNOSIS CONSIDERATIONS

The clinical presentation of BCTs can vary considerably. Many patients with BCTs will be asymptomatic; other patients with BCTs might present with nonspecific symptoms, most commonly abdominal pain and distention (55% to 90%). Although laboratory values are normal in most patients, approximately 20% of patients present with elevated liver function tests, such as an abnormal bilirubin level. Obstructive jaundice and cholangitis are rare and do not correlate with malignant disease, and typically occur with extrahepatic BCT. Hemorrhage and cyst rupture are other recognized, yet very uncommon, presenting complications of these lesions.

The differential diagnosis of patients with complex cystic lesions of the liver includes BCT, as well as hydatid cyst, post-traumatic cyst, liver abscess, polycystic disease, hemorrhagic cyst, embryonal sarcoma, primary or metastatic necrotic neoplasm, atypical simple cyst, and biliary intraductal papillary mucinous neoplasm (IPMN). Biliary IPMN is a recently recognized entity characterized...
by mucin production and prominent intraductal papillary proliferation.34,35 Biliary IPMN occurs equally in both sexes, with a mean age of 58 years.34,36 Some authors now advocate classifying BCTs with biliary tree communication and no ovarian stroma as biliary IPMNs.23,35-37 In one study, Zen and colleagues35 identified 9 cases of BCT, noting 5 tumors with direct communication to the biliary tree. None of these 5 cases contained ovarian-like stroma and all had clinicopathologic characteristics different from BCT.35 In a separate study, Nakagawa and colleagues24 were able to preoperatively differentiate BCA from biliary IPMN based on 3 main imaging characteristics. First, BCAs typically have a smooth tumor wall with septa inside the tumor, resulting in a “cysts-in-cyst” appearance. Second, papillary projections are typical in IPMN and less so in BCA. Third, demonstration of cystic tumor and intrahepatic bile duct communication along with the presence of distal mucin is more characteristic of biliary IPMN.24,36,38 Despite management principles similar to BCT, preoperative identification of cystic biliary IPMN can alert the surgeon to its typical superficial spreading tumor growth pattern.24,38

**IMAGING**

Cross-sectional imaging with either CT or MRI can accurately define and characterize simple cystic lesions. In contrast, the accuracy of cross-sectional imaging to diagnose the different types of complex cystic lesions can be relatively low. Certain radiographic findings can be helpful when trying to differentiate BCT from other non-neoplastic pathologies. Specifically, ultrasound, CT, and MRI combined with clinicopathological features can aid in preoperative differentiation and characterization of hepatic cystic tumors.

Sonographically, BCTs are anechoic with thickened irregular walls and internal septations (Table 1).8,16,39 Septal thickening, papillary infolding, and mural nodules are characteristic of BCTs.40,41 Biliary cystadenocarcinoma is more likely to contain mural or septal nodules and papillary projections.42-44 In the series by Seo and colleagues,45 the authors described their experience with 20 BCT and 19 resected simple cysts that mimicked
Biliary cystadenocarcinoma was associated with intracystic debris, bile duct dilation, and mural nodules. Computed tomography and ultrasound are complementary modalities in evaluating BCTs. Sonography is more sensitive for detecting septa in cystic lesions, and CT more accurately demonstrates size and anatomic extent of these lesions. On CT, BCT lesions are isodense to water (<30 HU) with nodular areas enhanced with IV contrast. Biliary duct dilation, single cysts, and lesions in the left lobe of the liver can be predictive of BCT on CT. In fact, multiple investigators have suggested that BCTs are more often located in the left hemi-liver and, therefore, BCT should be considered for suspicious cystic lesions in the left liver, especially in the setting of an increase in alkaline phosphatase.

Although there have been anecdotal reports on its potential usefulness in identifying malignancy with BCAC, no definitive conclusions can be made about the use of PET-CT for BCTs.

Magnetic resonance imaging is another useful tool for evaluating BCT. Biliary cystic tumors are typically multilocular with irregular thick walls on MRI. A homogeneous low-intensity T1 signal and high-intensity T2 signal are both characteristic. Magnetic resonance imaging characterizes cyst fluid content by varying signal intensities on T1-weighted images, depending on cyst fluid protein content. Linear low-signal intensity within high-intensity cysts identifies septations on T2-weighted images. Magnetic resonance imaging demonstrates the anatomic relationships within the liver and can aid surgical planning. The addition of diffusion-weighted MR to conventional MRI sequences aids in the qualitative and quantitative assessment of hepatic lesions with improved cyst characterization and detection of malignancy. Additional studies will be required to establish the relative additional utility of diffusion-weighted MR. Magnetic resonance cholangiopancreatography can also be helpful because it can demonstrate cyst communication with the biliary tree as well as identify internal septations. Magnetic resonance cholangiopancreatography can better visualize the biliary tree proximal to an obstruction, unlike ERCP. Endoscopic retrograde cholangiopancreatography is not commonly used for BCTs, but in select circumstances can be helpful obtaining tissue samples and identifying biliary tree communications and extrahepatic BCTs.

On cross-sectional imaging, BCTs need to be differentiated from other entities, such as simple hepatic cysts.

### Table 1. Radiologic Characteristics of Cystic Liver Lesions

<table>
<thead>
<tr>
<th>Biliary cystic tumors</th>
<th>Hepatic simple cysts</th>
<th>Echinococcal cysts</th>
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<tbody>
<tr>
<td>Multiloculated cyst</td>
<td>Anechoic</td>
<td>Daughter cyst within main cyst</td>
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<tr>
<td>Internal septation</td>
<td>Smooth borders</td>
<td>Intracystic debris</td>
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<tr>
<td>Enhancing cyst wall</td>
<td>No perceptible wall</td>
<td>Low signal intensity rim on T2-weighted MRI</td>
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<tr>
<td>Calcifications</td>
<td>No septations</td>
<td></td>
</tr>
<tr>
<td>Papillary wall nodules</td>
<td>No enhancement on CT with IV contrast</td>
<td></td>
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<tr>
<td>Thickened irregular wall</td>
<td>Water attenuation on CT</td>
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</table>

Enhancement on CT with IV contrast

Water attenuation on CT

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**Figure 2.** Biliary cystadenoma in a 53-year-old female patient. (A) T2 and (B) post-contrast T1-weighted images in the axial plane reveal a 3.5-cm multiseptated mass in the left lobe of the liver, with associated ductal dilation in the left lobe. Left lobectomy revealed biliary cystadenoma.
and infectious or hemorrhagic cysts. Cysts without internal septa or papillary projection are most likely simple hepatic cysts and can be managed conservatively. Simple hepatic cysts have a better-defined tissue/fluid interface on imaging than BCTs (Fig. 3). In addition, Choi and colleagues, in their series of 31 patients, identified the presence of a septum and septal thickening as important predictors of BCA vs simple hepatic cysts. Echinococcal cyst and liver abscesses are the 2 other cystic liver lesions that can sometimes be difficult to differentiate from BCTs. Echinococcal cysts present with debris or daughter cysts within the cysts. Grisolia and colleagues reliably differentiated nonparasitic cysts from echinococcal cysts without using fine-needle aspiration by developing sonographic criteria in conjunction with routine serology. Their criteria included border irregularity, irregular shape, calcification, sex, ultrasound appearance at follow-up, and presence of septa (Fig. 4A). Liver abscesses typically have a septated or multilocular appearance along with an irregular thicker wall than that seen with BCT (Fig. 4B). The combination of imaging characteristics and serologic analysis along with culture of cyst fluid can aid in making an accurate diagnosis.

Differentiating hemorrhagic cysts from BCT can similarly be accomplished using multimodality imaging. Ultrasound displays intracystic clots as irregular and nodular septal images. Computed tomography is less sensitive at detecting intracystic contents and generally demonstrates a homogeneous cyst even in the presence of blood within the cyst. On MRI, high-signal T1 and low-intensity T2-weighted images from clot formation are also seen. This characteristic discrepancy in ultrasound and CT, along with the MRI findings, suggest the diagnosis of hemorrhagic cyst. Mesenchymal hamartoma and undifferentiated embryonal sarcoma have a multiloculated cystic appearance, but are generally seen in children and young adults and without the characteristic female predominance of BCT.

Although cross-sectional imaging can more reliably differentiate BCTs from other non-BCT lesions, BCA and BCAC cannot be reliably differentiated with preoperative imaging. Both BCA and BCAC can appear as solitary, multilocular cystic lesions. Mural nodule and irregular cyst-wall thickness on CT might suggest a higher likelihood of malignancy, but these features are not pathognomonic for BCAC. Wang and colleagues proposed a novel scoring system using age, sex, duration of symptoms, and lesion location to differentiate BCA and BCAC. The authors validated this scoring system using 2 separate patient cohorts and demonstrated a >90% sensitivity and specificity. This scoring system, however, necessitates additional validation before being adopted on a more widespread basis.

Image-guided aspiration of suspected BCTs is sometimes undertaken. Aspiration of BCT often demonstrates bile-tinged mucin and can allow differentiation from parasitic cysts, hematoma, or hemorrhagic cysts. With adequate tissue sampling, cytology can reveal malignant glandular cells and mucin suggesting BCAC, although these cells are rarely identified. The role of cyst fluid analysis with carbohydrate antigen 19-9 (CA 19-9) and CEA has been studied. Pinto and Kaye used intracystic CEA levels to differentiate BCTs from benign (nonmucinous) cysts and abscesses with 100% sensitivity and 94% specificity. Other case reports demonstrated elevated serum and cystic fluid CA 19-9 levels in patients with BCTs. These findings, however, have not been replicated in other studies. The role of cyst fluid CEA, CA 19-9, and serum tumor markers remains controversial. Although many providers might obtain a CA 19-9, the sensitivity and specificity are not high enough to differentiate a BCA from a BCAC. In addition, fine-needle aspiration of suspected BCAC has

Figure 3. Incidental simple cyst in a 60-year-old female patient. (A) Axial and (B) coronal CT images obtained in the portal venous phase show a large 7-cm well-defined simple cyst, with no internal septations or enhancing mural nodules. (C) Coronal T2-weighted MR image shows a well-defined hyperintense lesion compatible with simple cyst.
been associated with pleural and peritoneal dissemination.\textsuperscript{7,8,56} Because of this risk, along with the low likelihood of identifying malignant cells, routine fine-needle aspiration and core-needle biopsy of suspected BCTs should generally be avoided.\textsuperscript{23,65,66}

**PATHOLOGICAL CHARACTERISTICS**

On gross pathology, BCTs are generally solitary, multilocular cystic lesions with fluid contents\textsuperscript{10}; multifocal BCT are rare.\textsuperscript{21,47,71} Cyst walls (BCA or BCAC) can have dense fibrosis and even calcifications. Biliary cystadenomas exhibit pure biliary immunophenotype with a single layer of cuboidal to columnar epithelium supported by fibrous connective tissue (Fig. 5).\textsuperscript{3,19,36,72} Classically, BCA has been characterized by ovarian-type stroma that typically expresses estrogen and progesterone receptors (60\% to 100\%).\textsuperscript{36} This finding might explain the high incidence among females,\textsuperscript{10,19,36,73,74} as well as the reported finding that BCAs occurred more often in patients on hormonal therapy and are known to increase in size during pregnancy.\textsuperscript{19,23,74,75} Wheeler and Edmondson\textsuperscript{18} have defined BCA based on the presence of mesenchymal stroma.\textsuperscript{76} This description of BCA is characterized by 3 distinct layers: an epithelial layer of mucin-producing cells; a layer of undifferentiated mesenchymal cells under the epithelial lining, and a dense layer of collagenous connective tissue outside the stromal bands of cells.\textsuperscript{18,24} All
13 patients with ovarian-like stroma BCTs reported in Wheeler and Edmondson’s case series were female and had a mean age of 41.7 years. A subsequent review of the literature identified an additional 27 cases of BCA with mesenchymal stroma. These patients were similarly all female and had a mean age of 44.1 years. In contrast, patients with BCA lacking mesenchymal stroma had a mean age at diagnosis of 53.4 years and were distributed equally between the sexes. Defining BCA based solely on the presence of ovarian stroma might be insufficient because a number of reported BCTs lack this stromal component.

In contrast to BCA, BCAC is identified by the presence of proliferating cytologically malignant epithelium. Multilayered epithelium, frequent mitotic figures, loss of polarity, and nuclear pleomorphism are features that suggest a BCA has become a BCAC. The presence of mesenchymal (ovarian-like) stroma has important prognostic implications. Specifically, BCAC without mesenchymal stroma appears to progress to malignancy more often, disseminate more rapidly, and is associated with a worse prognosis than BCAC without this distinctive component.

In one of the largest BCT series, Devaney and colleagues noted a median survival of 3 years in patients with BCAC without mesenchymal stroma vs no deaths in 4 to 8 years of follow-up in all cases of mesenchymal-associated BCAC. Biliary cystadenocarcinoma without ovarian stroma occurs in both men and women as opposed to the almost exclusive female incidence seen in BCAC with ovarian-type stroma. Biliary cystadenoma ovarian stroma can regress during malignant transformation. Both BCA and BCAC epithelial cells are characterized by mucin production and immunoreactivities to cytokeratins (CAM5.2, AE1, AE3), CA 19-9, epithelial membrane antigen, and CEA. Immunohistochemical analysis has demonstrated CEA and CA 19-9 in the epithelial component of BCT. Carcinoembryonic antigen is localized to the luminal surface of BCA epithelial cells, and BCAC epithelium have CEA diffusely within the cytoplasm. D’Errico and colleagues noted markedly increased albumin messenger RNA in all 8 BCAC lesions they investigated, and noted its absence in BCA and other benign biliary cysts.

MANAGEMENT

The risk of malignant transformation of BCA to BCAC can be as high as 20%. Appropriate management is critical, given the malignant potential and propensity for the lesion to recur. Therefore, in general, attempts at percutaneous aspiration, ethanol injection, and unroofing are unnecessary and generally inappropriate. When aspiration is undertaken for a presumed simple cyst, rapid recurrence can denote misdiagnosed BCT. In fact, fenestration, aspiration, sclerosis, internal drainage, marsupialization, or partial resection with or without cavity ablation can result in recurrence rate as high as 80% to 90%. As such, formal surgical resection with negative margins is recommended. Appropriate surgical therapies can include liver resection or enucleation, depending on the individual patient; anatomic position of the cyst; and surgeon experience. Although peripheral lesions or those BCTs relegated to one side of the liver should be treated with formal resection, centrally located lesions that involve central vascular or biliary structures can require enucleation. Extrahepatic BCTs will require complete resection in conjunction with resection of the bile duct followed by biliary diversion. A few case reports of laparoscopic BCT management have been published and reveal acceptable morbidity with recurrence rates similar to open series. In the largest BCT laparoscopic resection series, Koffron and colleagues resected 22 BCTs laparoscopically, performing lobectomy (n = 1), segmentectomy (n = 8), marsupialization with remnant cyst fulguration (n = 10), enucleation (n = 2), and subtotal resection (n = 1). With a mean follow-up of 16 months, they reported 1 recurrence (5%) in a patient who had undergone marsupialization and noted less morbidity in the laparoscopic cohort compared with their open cohort of 12 patients. Whether the operative approach is open or laparoscopic, the surgeon should strive for complete extirpation of the BCT. Leaving residual BCA based on intraoperative frozen-section analysis that shows no evidence of overt malignancy (eg, BCAC) should generally be avoided. Intraoperative frozen-section analysis is often insensitive, most likely due to small focus of neoplastic cells that can be missed by partial sampling.

In addition to surgical resection, a few single case reports of orthotopic liver transplantation for BCT management have been reported. For example, Romagnoli and colleagues reported successful outcomes after performing orthotopic liver transplantation in a symptomatic BCT deemed unresectable. Given BCAC’s indolent course, the use of liver transplantation should be used in rare circumstances only, when formal resection is not possible and only after multidisciplinary review by an experienced team.

OUTCOMES AND PROGNOSIS

Due to the rarity of these lesions, relatively little is known about the prognosis of BCTs. Based on the few small series that have been published, recurrence rates for BCAs seem to be extremely low after appropriate surgical
management (5% to 10%). Most often BCAC recurs locally in the liver, perhaps due to initial inadequate local management. Recurrent extrahepatic disease is relatively uncommon, but does occur in a subset of patients (20%). However, at least one series reported that even some patients with metastatic disease, depending on the subtype, can live beyond 10 years. For overall survival, surgical resection of BCTs is associated with improved prognosis compared with other malignant liver tumors, such as hepatocellular carcinoma and cholangiocarcinoma (57% vs 40% vs 22% 5-year survival, respectively).

In fact, overall survival for patients with benign BCA in one large series was >90%, with 18 years of follow-up. Among patients with BCAC, the prognosis is worse than BCA, but still better than other primary malignancies of the liver. Several series have reported that complete surgical resection of BCAC can result in 5-year survival of 65% to 70%. In contrast, at least one series noted that partial excision of BCAC had a considerably worse 5-year survival of only 36%.

Prognostic factors associated with survival after resection of BCAC include tumor subtype as well as the type of stroma found in the tumor. Nakajima and colleagues classified BCAC into 2 subtypes: invasive and noninvasive based on the carcinoma extension into the liver. The authors reported a significant difference in survival. Specifically, there were no recurrences or deaths noted among patients with noninvasive BCAC compared with a median survival of only 7 months among those patients who had the invasive subtype. In a separate study, Deva-ney and colleagues reported that patients with a BCAC characterized by ovarian-like stroma on pathology had

<table>
<thead>
<tr>
<th>First author, year</th>
<th>Patients</th>
<th>Operative procedure</th>
<th>Outcomes</th>
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<tbody>
<tr>
<td>Martel, 2012⁶⁷</td>
<td>n=13 (12 F); BCA (n=11); BCAC (n=2); mean age 51 y</td>
<td>Liver resection (3); enucleation (6); unroofing (4)</td>
<td>No mortality; median follow-up 23.1 mo; no BCA or BCAC recurrences</td>
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<tr>
<td>Pillai, 2012²³</td>
<td>n=13 (BCA, 11 F); mean age 46 y</td>
<td>Hepatectomy (5); segmentectomy (5); enucleation (3)</td>
<td>No mortality; median follow-up 22 mo; no recurrence</td>
</tr>
<tr>
<td>Ratti, 2012⁶¹</td>
<td>n=12 (BCA, 12 F); mean age 45 y</td>
<td>Hepatectomy (12)</td>
<td>No mortality; morbidity 16.7%; median follow-up 16 mo; no recurrence</td>
</tr>
<tr>
<td>Sang, 2011⁸</td>
<td>n=33; BCA (n=19, 17 F), Mean age 44.2 y; BCAC (n=14, 5 F), Mean age 57.0 y</td>
<td>Enucleation (3) (BCA 3); fenestration (2) (BCA 1); segmentectomy (6) (BCA 5) (BCAC 1); sectionectomy (7) (BCA 5) (BCAC 2); hemihepatectomy (7) (BCA 5) (BCAC 4); trilobectomy (1) (BCAC 1)</td>
<td>BCA: 1 patient death 8 mo postoperatively, secondary to metastatic disease after BCA resection; median follow-up 2.5 mo; no recurrence in fenestration patient, 14.4 mo of follow up</td>
</tr>
<tr>
<td>Erdogan, 2010⁷⁷</td>
<td>n=15 (13 F); BCA (n=12); BCAC (n=3)</td>
<td>Hepatectomy (6); enucleation (9)</td>
<td>No mortality; morbidity 13%; no follow-up recorded</td>
</tr>
<tr>
<td>Teoh, 2006⁶⁵</td>
<td>n=20 (BCA, 16 F); mean age 58 y</td>
<td>Hepatectomy (10); enucleation (10)</td>
<td>Mortality rate not reported; mean follow-up 5.5 y (1 lost to follow-up); no recurrences after resection</td>
</tr>
<tr>
<td>Daniels, 2006⁷⁴</td>
<td>n=12; BCA, 12 F, mean age 50.5 y</td>
<td>Hepatectomy (2); enucleation (4); combination hepatectomy enucleation (7)</td>
<td>25% morbidity rate; wound infection (1); pulmonary embolus (1); bile fistula (1); no mortality</td>
</tr>
<tr>
<td>Thomas, 2005⁵⁰</td>
<td>n=19 (18 F); BCA (n=18); BCAC (n=1); mean age 48 y</td>
<td>Hepatectomy (12); enucleation (6); fenestration (1)</td>
<td>No mortality; mean follow-up 4 y; no recurrences</td>
</tr>
<tr>
<td>Vogt, 2005⁶⁶</td>
<td>n=22; BCA (n=18, 18 F); mean age 48 y; BCAC (n=4, 3 F); mean age 60 y</td>
<td>Hepatectomy (14); enucleation (8)</td>
<td>2 deaths at 6 and 12 mo secondary to metastatic BCAC</td>
</tr>
<tr>
<td>Koffron, 2004⁷⁰</td>
<td>n=34; BCA (n=33); BCAC (n=1)</td>
<td>Open resection (11); open enucleation (1); laparoscopic resection (10); 1 subtotal resection; laparoscopic enucleation (2); laparoscopic marsupialization (10)</td>
<td>No mortality; 1 recurrence after BCA laparoscopic marsupialization</td>
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BCA, biliary cystadenoma; BCAC, biliary cystadenocarcinoma; F, female.
a considerably better long-term prognosis compared with patients who had BCAC with tumors that lacked ovarian-like mesenchymal stroma. Vogt and colleagues\(^6\) similarly noted a worse prognosis among men with BCAC, probably due to the higher likelihood of BCAC without mesenchymal stroma in males.

Very few reports exist on the role of chemotherapy and radiation for BCAC. Anecdotal case studies have reported salvage or adjuvant chemotherapy, radiation, or transcatheter arterial chemoembolization.\(^8\) Laufer and colleagues,\(^7\) in their meta-analysis of 112 invasive BCAC cases, reported a 33% five-year survival for patients with unresectable BCAC who were treated with definitive chemotherapy and radiation. Due to the lack of data, no evidence-based recommendations can be made about the role chemotherapy and radiation therapy in BCAC. However, given the excellent long-term outcomes after surgery alone for BCAC, there is probably no role for adjuvant therapy.

**CONCLUSIONS**

Improving imaging modalities and increased abdominal imaging has led to higher detection rates of cystic lesions of the liver. Biliary cystic tumor incidence correlates significantly with sex and age and should be the primary diagnostic consideration in a middle-aged woman with a well-encapsulated, multilocular cystic liver mass. Diagnostic modalities, such as ultrasound, CT, MRI, and ERCP, yield important information about the nature of the cyst, but cannot reliably distinguish BCA from BCAC. As such, suspected BCTs should be extirpated surgically with complete removal of the lesion when feasible. After excision of BCA, long-term outcomes are good, however, patients with BCAC have a worse long-term prognosis. Due to the rarity of this disease, future multi-institutional studies will be needed to better understand the management, biology, and prognosis of patients with BCTs.

**Author Contributions**

Study conception and design: Soares, Arnaoutakis, Pawlik

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Drafting of manuscript: Soares, Arnaoutakis, Kamel, Anders, Adams, Bauer, Pawlik

Critical revision: Soares, Arnaoutakis, Kamel, Anders, Adams, Bauer, Pawlik

**REFERENCES**


22. Bardin RL, Trupiano JK, Howerton RM, Geisinger KR. Oncocytic biliary cystadenocarcinoma: a case report and


