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Staging of extrahepatic cholangiocarcinoma

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Abstract Preoperative staging of extrahepatic cholangiocarcinoma is important in determining the best treatment plan. Several classification systems have been suggested to determine the operability and extent of surgery. Longitudinal tumor extent is especially important in extrahepatic cholangiocarcinoma because operative methods differ depending on the tumor extent. The Bismuth-Corlette classification system provides useful information when planning for surgery. However, this classification system is not adequate for selecting surgical candidates. Anatomic variation of the bile duct and gross morphology of the tumor must be considered simultaneously. Lateral spread of the tumor can be evaluated based on the TNM staging provided by American Joint Committee on Cancer (AJCC). However, there is a potential for ambiguity in the distinction of T1 and T2 cancer from one another. In addition, T stage does not necessarily mean invasiveness. Blumgart T staging is helpful for the assessment of resectability with the consideration of nodal status and distant metastasis as suggested by the AJCC cancer staging system. Computed tomography (CT) and magnetic resonance imaging (MRI) are the primary tools used in the assessment of longitudinal and lateral spread of a tumor when determining respectability. Diagnostic laparoscopy and positron emission tomography (PET) may play additional roles in this regard.

Keywords Bile duct cancer · Extrahepatic cholangiocarcinoma · Staging

Introduction

An extrahepatic cholangiocarcinoma is a malignant tumor that arises from the ductal epithelium of the extrahepatic bile duct (EBD) and is typically classified as an adenocarcinoma. There are several risk factors for extrahepatic cholangiocarcinoma including choledochal cysts, cholangitis, biliary cirrhosis, cholelithiasis, thyrotoxicosis, and obesity [1, 2]. About two-thirds of EBD cancers arise at the hepatic hilum with one third arising from the distal common bile duct [3, 4]. EBD cancer arising from the hepatic hilum is also called Klatskin tumor [5]. An extrahepatic cholangiocarcinoma is a rare tumor, and its prognosis is very poor [3]. The only curative treatment is complete resection with a negative surgical margin [3, 4, 6–8]. Preoperative evaluation of the tumor is important in order to evaluate resectability and the extent of surgery. Several methods are proposed to evaluate tumor extension. Bismuth-Corlette classification has been used to define the longitudinal tumor extension in hilar cholangiocarcinoma.

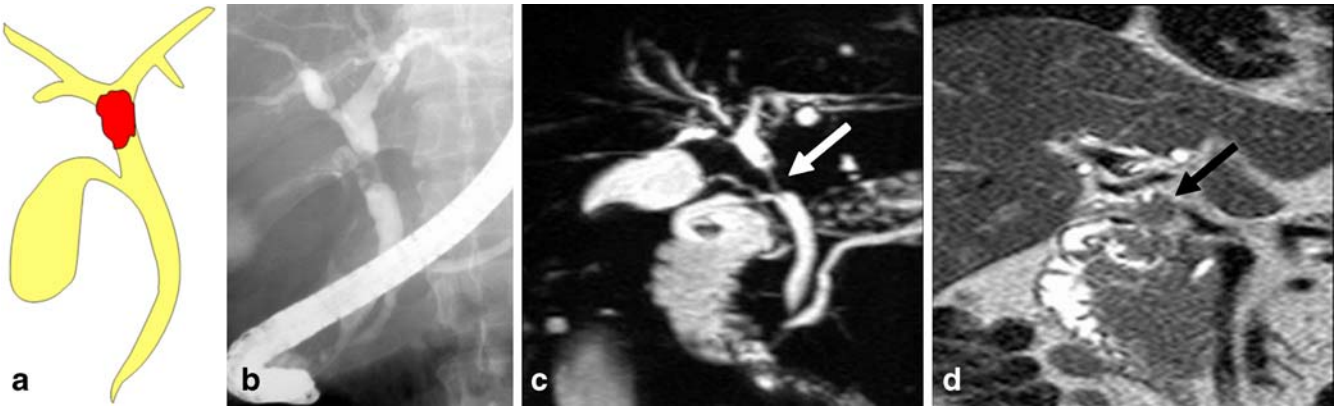

Fig. 1 **a** Drawing illustrates Bismuth-Corlette type I tumor that is confined below the confluence of the right and left hepatic duct. **b** ERCP, **c** MRCP, and **d** coronal T2-weighted image show extrahepatic cholangiocarcinoma in the upper third of the EBD without invasion of the hilum (arrows)

Lateral tumor extension can be defined by using T stages of the AJCC cancer staging system. Resectability of the tumor can be evaluated by the Blumgart T-staging system combined with the AJCC cancer staging system.

In this review, we will discuss the evaluation of ductal extension according to the Bismuth-Corlette classification and will also discuss the assessment of soft tissue extension based on TNM staging as well as the determination of resectability.

Evaluation of longitudinal spread

In extrahepatic cholangiocarcinoma, determining tumor location and its longitudinal extent is important because surgical method and survival rate are dependent upon these factors [3, 4, 8, 9]. Traditionally, extrahepatic cholangiocarcinoma has been classified into upper, middle, and lower third cancer by its location [4, 9, 10]. However, in recent literature, extrahepatic cholangiocarcinoma has been simply classified into perihilar and distal cancer, which is due to the relatively low incidence of middle bile duct cancer, and its operative method is similar to the Bismuth-Corlette type I hilar cholangiocarcinoma [3, 8, 11]. Perihilar tumors can be defined as those involving or requiring resection of the hepatic duct bifurcation and are typically located in the extrahepatic biliary tree proximal to the origin of the cystic duct [12]. Perihilar cholangiocarcinomas can be categorized by the modified Bismuth-Corlette classification into four types, which has been adopted from an original classification with three types [13, 14]. More sophisticated classification has been suggested, but it has not been widely adopted [15].

The Bismuth-Corlette type I tumor is defined by the presence of a lesion confined below the confluence of the right and left hepatic ducts (Fig. 1). This type of tumor can be treated with segmental resection of the EBD and regional lymph node dissection. Hepatectomy is unnecessary if the resection margin is microscopically confirmed to be negative by frozen pathology during surgery [16]. However, in some cases, it is not always easy to define type I on imaging. This is due to either the variation of the cystic duct origin or to poor demarcation of the upper and lower margins of the tumor. If a bile duct tumor spread across the origin of the cystic duct across the upper and middle third of the EBD without involvement of the confluence of the

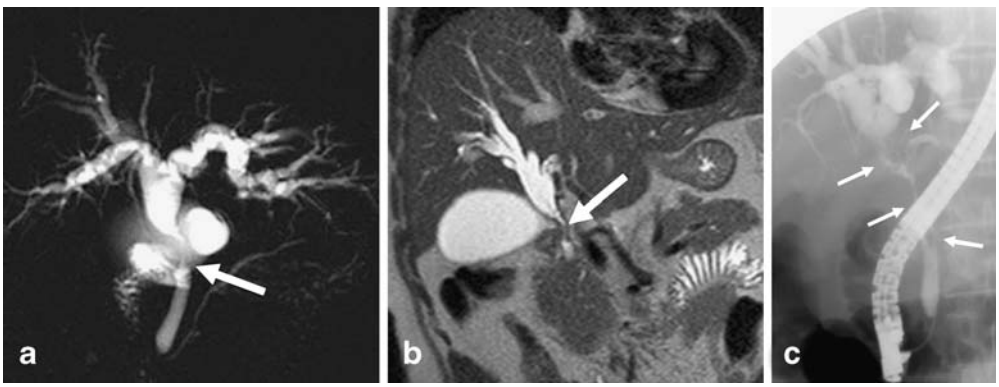

Fig. 2 **a** MRCP and **b** T2-weighted images show a perihilar or middle bile duct carcinoma sparing the confluence of the left and right hepatic duct (arrows). **c** On endoscopic retrograde cholangiography, the tumor seems more extended to the hilum and mid CBD, because of the incomplete filling of contrast media in the proximal duct (small arrows)

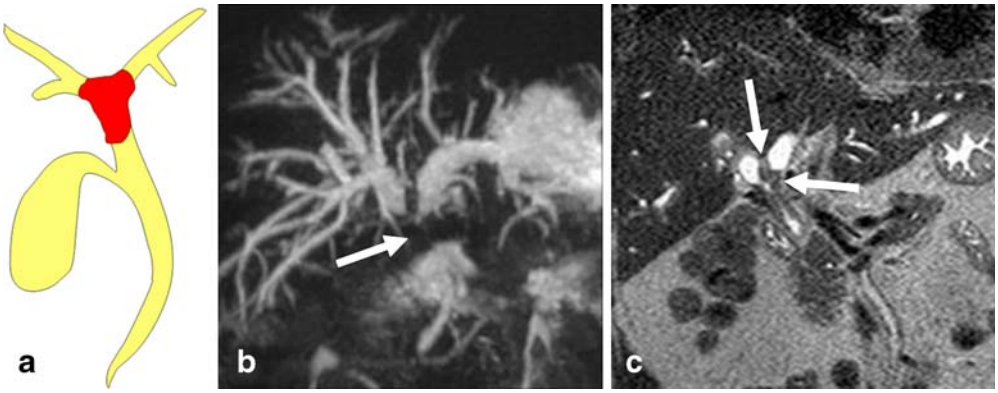

Fig. 3 **a** Drawing illustrates Bismuth Corlette type II tumor. **b** MRCP and **c** coronal T2-weighted images show the tumor in the hepatic hilum without invasion of the second confluence of the intrahepatic bile duct (arrows)

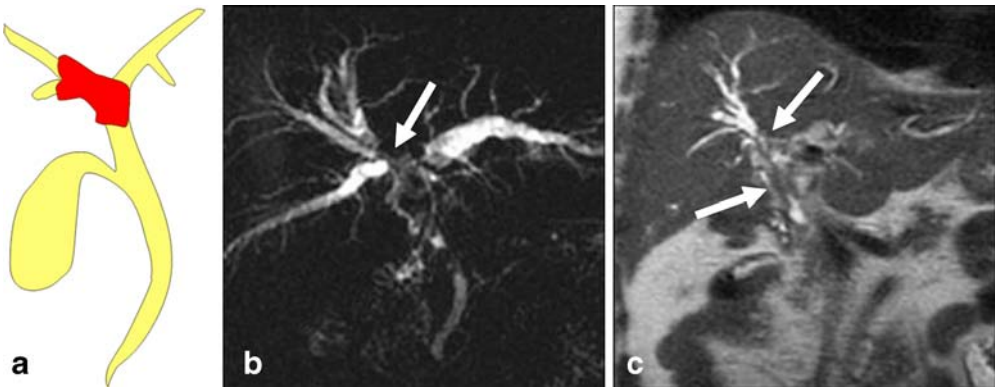

Fig. 4 **a** Drawing illustrates Bismuth Corlette type IIIa tumor. **b** MRCP and **c** coronal T2-weighted images show that the hilar tumor extends to the second confluence of the right hepatic duct (arrows)

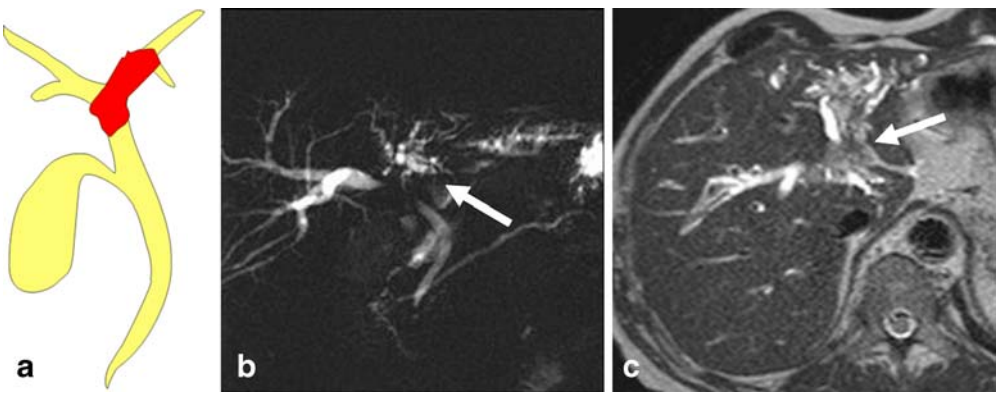

Fig. 5 **a** Drawing illustrates Bismuth Corlette type IIIb tumor. **b** MRCP and **c** axial T2-weighted images show that the hilar tumor extends to the second confluence of the left hepatic duct (arrows)

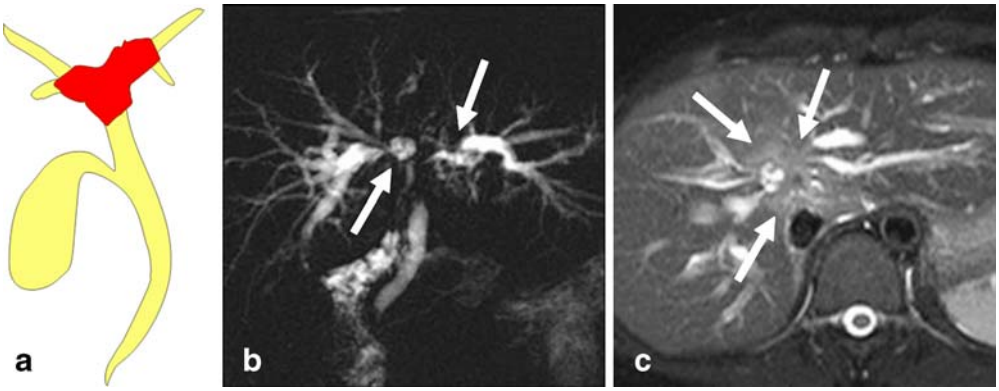

Fig. 6 **a** Drawing illustrates Bismuth Corlette type-IV tumor. **b** MRCP and **c** axial T2-weighted images show that the hilar tumor extends to the second confluences on both the right and left hepatic ducts (arrows)

right and left bile ducts, it would be classified as either type I perihilar cholangiocarcinoma or middle EBD cancer (Fig. 2).

Bismuth type II tumors extend to the confluence of the right and left hepatic ducts, which can be readily seen on cross-sectional imaging (Fig. 3). Bismuth type II tumor can be treated by bile duct resection with hepaticojejunostomy and regional lymph node dissection. Caudate lobectomy is mandatory when the tumor infiltrates caudate bile duct branches [16, 17]. A type 3a tumor extends to the bifurcation of the right hepatic duct (Fig. 4), and a type 3b tumor extends to the bifurcation of the left hepatic duct (Fig. 5). Hilar bile duct resection with hemihepatectomy including the caudate lobectomy and regional lymph node dissection is the standard surgical method for type III tumor [16–18]. Type 4 tumors extend to the bifurcation of both the right and left hepatic ducts and have been generally regarded as inoperable except for liver transplantation (Fig. 6). Recently, with advances in surgical technique, curatively intended surgery is attempted in the cases with type IV tumor extending less than 2 cm from the hilum

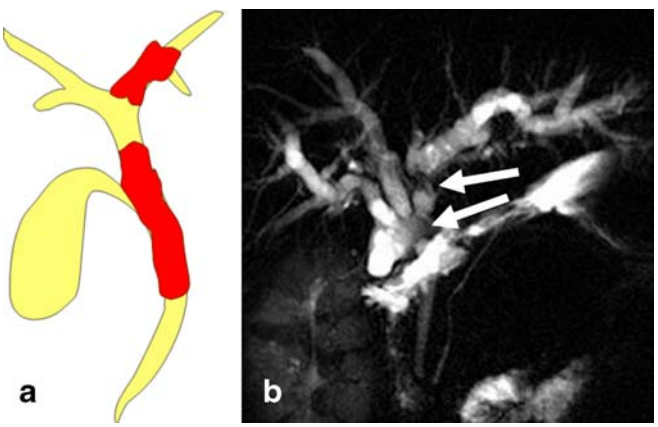

Fig. 7 **a** Drawing illustrates multicentric cholangiocarcinomas, which are also classified into Bismuth Corlette type IV. **b** MRCP shows skipped luminal narrowing in the EBD, suggesting multicentric EBD tumor (arrows)

[19]. Multicentric tumors are also included in this category (Fig. 7).

For surgical planning based on the Bismuth-Corlette classification, variations of the bile duct anatomy that may affect the classification demand attention. Anatomical variation includes insertion of the right anterior or posterior duct to the left hepatic duct and trifurcation of the right anterior, posterior and left hepatic ducts (Fig. 8). These anatomical variations let patients diagnosed even as type IV to still undergo definitive surgery. For example, if the right anterior and right posterior bile ducts drain separately into the CBD, such as in trifurcation or if the right posterior bile duct drains to left bile duct (Fig. 9), type IV hilar cholangiocarcinoma can be treated by hilar bile duct resection with left hepatectomy and hepaticojenunostomy. Similarly, when left segment 4 duct drains directly into the CBD, even Bismuth type IV tumor can be completely removed by hilar bile duct resection with extended right hepatectomy [18]. In some cases, tumors can be classified as the incorrect Bismuth-Corlette type if anatomic variation is not considered carefully (Fig. 10).

Bismuth-Corlette classification provides an anatomical description of the tumor and is useful in determining resection or palliative treatment and the type of surgery, but surgical candidates cannot be determined solely by this classification, and it is not indicative of survival [7]. Another aspect to consider is that longitudinal spread pattern of a tumor can be related to gross morphology [20]. Papillary tumors frequently present with long-range mucosal spread, while infiltrating tumors tend to show subepithelial extension. The subepithelial infiltration may readily be depicted on CT or MRI by showing thickening or increased enhancement of the ductal wall, but the mucosal spread may hardly be visible on CT or MRI. Therefore, determination of longitudinal spread must be made more cautiously when a papillary or polypoid tumor is seen on imaging. Abe et al. [21] presented an illustrative case in which the utility of choledocoscopy is demonstrated. In that case, cholangiography showed a polypoid tumor in the middle CBD, but choledochoscopy demonstrated multifocal superficial spreading tumors along the

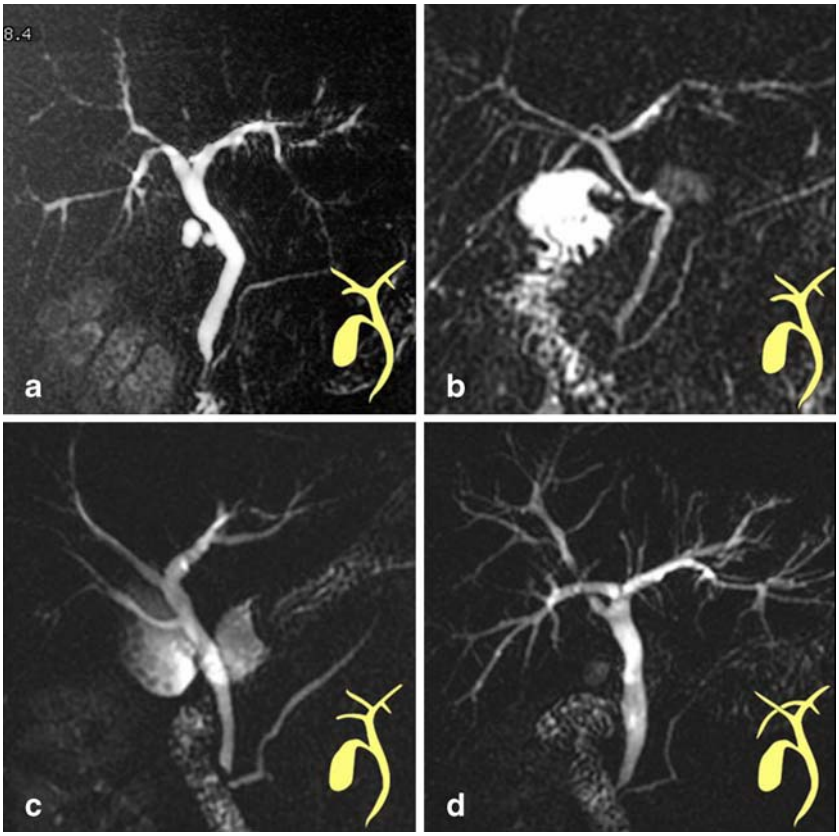

Fig. 8 Bile duct variations. MRCP shows **a** normal, **b** trifurcation, **c** right anterior duct drains to the left hepatic duct, and the right posterior duct drains to the common bile duct, and **d** the right posterior duct drains to the left hepatic duct

entire bile duct necessitating more extensive surgery than was expected from the cholangiography alone.

Evaluation of lateral extension

Assessment of the lateral spread and soft tissue extension can be evaluated based on the TNM staging system (Tables 1 and 2) [22].

A T1 tumor is confined to the bile duct, and most of the T1 tumors are papillary or polypoid. Papillary tumors frequently present during the T1 stage, even large ones [23] (Fig. 11). A T2 tumor extends beyond the wall of the bile duct and frequently presents as a periductal infiltrative or nodular mass showing irregular ductal wall thickening and increased enhancement (Fig. 12). T3 lesions include locally invasive lesions involving the liver, gall bladder, pancreas, or ipsilateral portal vein or hepatic artery (Fig. 13). In a

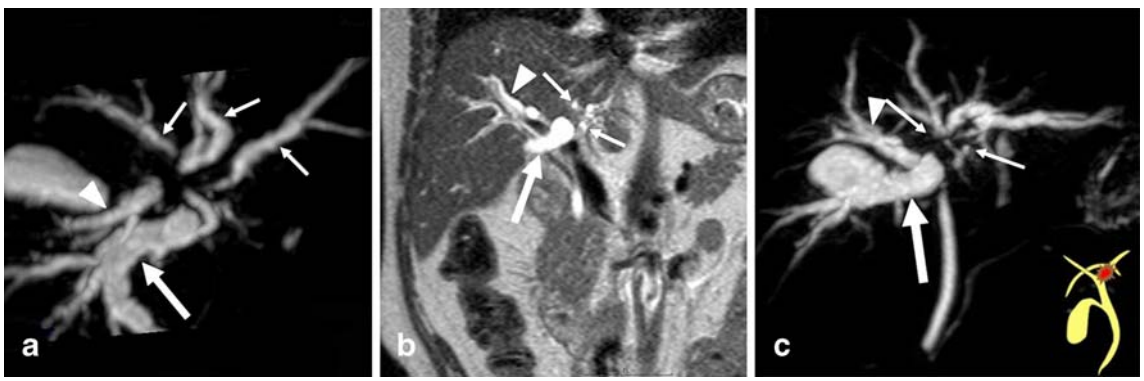

Fig. 9 **a** On craniocaudal view of MRCP, left segmental hepatic ducts (small arrows) and right anterior (arrow head) and posterior hepatic ducts (large arrow) are separated, suggesting Bismuth-Corlette type IV tumor. **b** T2-weighted coronal image and **c** MRCP show that the right posterior duct (large arrow) drains to the left hepatic duct (small arrows), whereas the right anterior duct (arrowhead) drains to the common bile duct. This patient underwent left lobectomy and choledochojejunostomy as for a Bismuth-Corlette type IIIb tumor

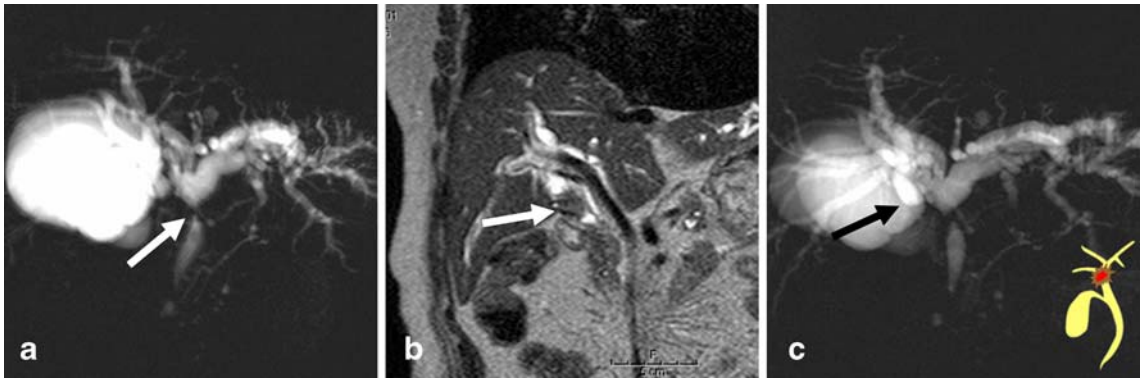

Fig. 10 **a** On coronal view of MRCP, a tumor (arrow) at the typical location of Bismuth-Corlette type I appears to involve the upper third of EBD without invasion of confluence. **b** On coronal T2-weighted and **c** oblique view of MRCP images show that the right posterior duct is also obstructed (black arrow), suggesting a type IIIa lesion. This patient shows anatomic variation of the right posterior hepatic duct being inserted into the common hepatic duct

Bismuth-Corlette type 3a cancer, tumor encasement of the right hepatic artery is frequently depicted as well as liver invasion. If a T3 lesion is seen in the distal CBD, enhancing the ductal wall mass may show periductal infiltration into the surrounding pancreas (Fig. 14).

On cross-sectional imaging, vascular involvement is regarded as present if vascular occlusion, ipsilateral hepatic atrophy, stenosis or contour deformity, or tumor contact where more than 50% of the perimeter of the vessel is observed (Fig. 15) [19]. Dynamic CT and MRI are useful to determine vascular involvement, but the findings may be equivocal between two examinations or even between different vascular phases in the same examination. In our experience, high-resolution T2-weighted images are useful to define the preserved fat plane between the tumor and vascular structures, which may be difficult to identify on dynamic images either on CT or MRI (Fig. 16). Intraportal endovascular ultrasonography (IPEUS) can be helpful in determining portal vein invasion. Disruption of echogenic band around the portal vein on IPEUS suggests tumor invasion with high diagnostic accuracy up to 95%–100% [24, 25]. But IPEUS is not performed widely because of its invasive nature and evolution of cross-sectional imaging modalities.

T4 stage includes widely invasive tumors involving the bilateral or main portal vein (Fig. 17), common hepatic artery, or invading adjacent organs, such as the colon, stomach, duodenum, or abdominal wall (Fig. 18).

Problems in the current T staging

According to the current T staging by the Sixth AJCC Cancer Staging System, there is a potential for ambiguity in distinguishing T1 and T2 extrahepatic cholangiocarcinomas [10, 26, 27]. T1 cancer is defined as a tumor confined to the bile duct histologically, and T2 cancer is defined as a tumor that extends beyond the bile duct. However, the determination of the border of the bile duct can be difficult even histologically in some cases [10, 26, 27]. The outer border of the bile duct is defined by the outer smooth layer;

Table 1 T Staging (AJCC 6th edn)

Stage	Criteria
Tis	Carcinoma *in situ*
T0	No evidence of primary tumor
T1	Tumor confined to the bile duct histologically
T2	Invades beyond the wall of the bile duct
T3	Tumor invades the liver, gall bladder, pancreas, and/or ipsilateral branches of the portal vein (right or left), or hepatic artery (right or left)
T4	Tumor invades any of the following: main portal vein or its branches bilaterally, common hepatic artery, or other adjacent structures, such as the colon, stomach, duodenum, or abdominal wall

Table 2 Blumgart staging system

Stage	Criteria
T1	Tumor involving biliary confluence with or without unilateral extension to second-order biliary radicles
T2	Tumor involving biliary confluence with or without unilateral extension to second-order biliary radicles and ipsilateral portal vein involvement with or without ipsilateral hepatic lobar atrophy
T3	Tumor involving biliary confluence + bilateral extension to second-order biliary radicles or unilateral extension to second-order biliary radicles with contralateral portal vein involvement or unilateral extension to second-order biliary radicles with contralateral hepatic lobar atrophy or main or bilateral portal vein involvement

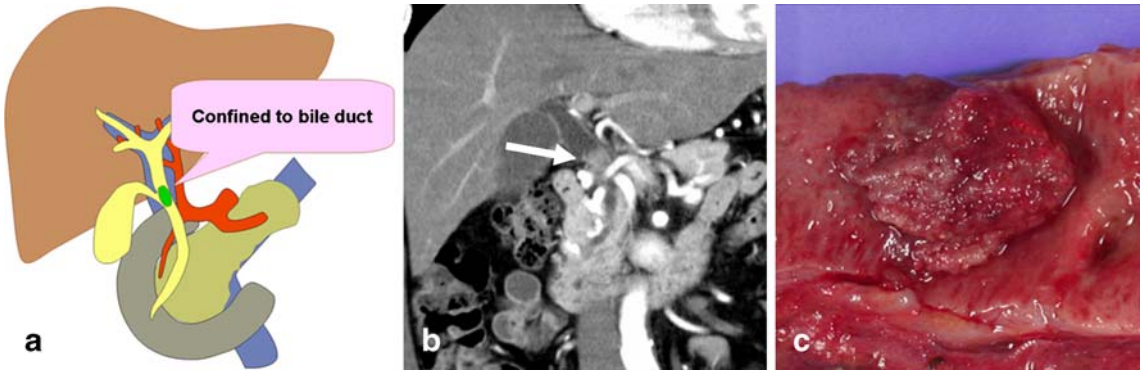

Fig. 11 **a** Drawing illustrates T1 extrahepatic cholangiocarcinoma confined to bile duct histologically. **b** Coronal CT shows a polypoid enhancing mass in the common bile duct (arrow). **c** Photograph of a gross specimen shows a polypoid mass comparable with that seen on a CT

however, it is well known that the distribution of the smooth muscle varies along the vertical length of the EBD: a continuous muscle layer may be seen in the distal EBD, but only an interrupted or scattered muscle layer is present in the middle EBD, and scant muscle fibers are seen in the proximal EBD [10]. This histologic characteristic of EBD and frequent desmoplastic stromal reaction accompanied in extrahepatic cholangiocarcinomas may lead to confusion in the distinction between T1 and T2 stages. In addition, because of the differences of surrounding organs, a middle CBD cancer with extended pericholedochal soft tissue can be staged as T2, while a tumor of the same depth of invasion may be staged as T3 in the distal bile duct because of minimal invasion of the pancreas [28] (Fig. 14). Recently, Hong et al. [27] proposed a new staging system based on direct measurement of the tumor's depth of invasion. However, it will be important for this system to be further evaluated in a larger scale study.

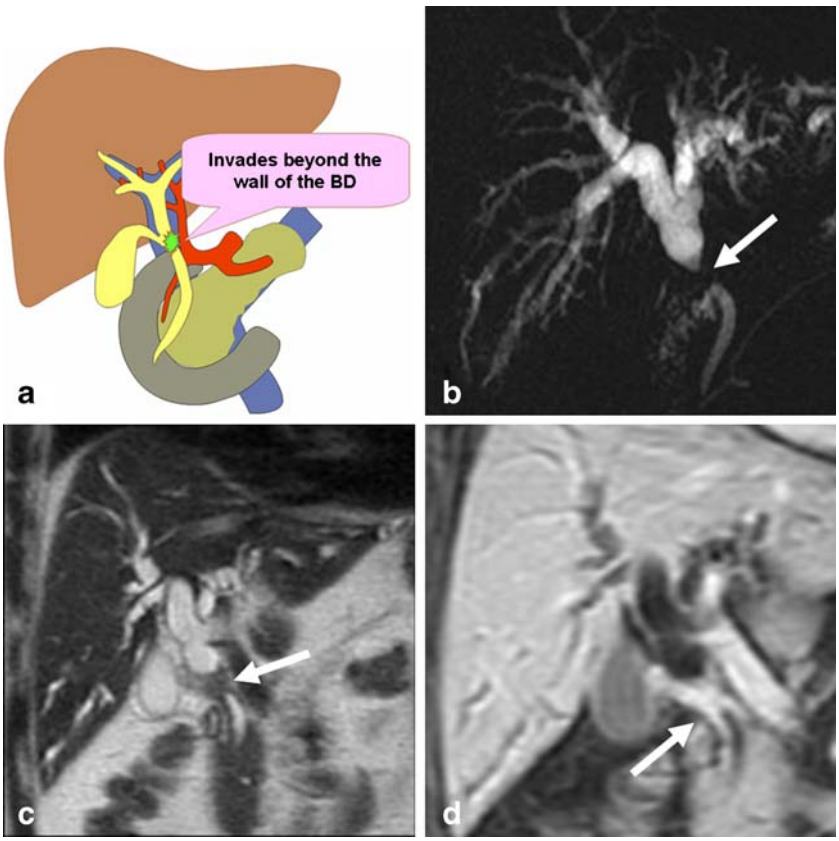

Fig. 12 **a** Drawing illustrates T2 extrahepatic cholangiocarcinoma extending beyond the wall of the bile duct. **b** MRCP shows an abrupt luminal narrowing at the level of middle CBD (arrow). **c** Coronal T2-weighted and **d** T1-weighted contrast-enhanced MR images show irregular ductal wall thickening and enhancement (arrows), suggesting a periductal, infiltrating, extrahepatic cholangiocarcinoma

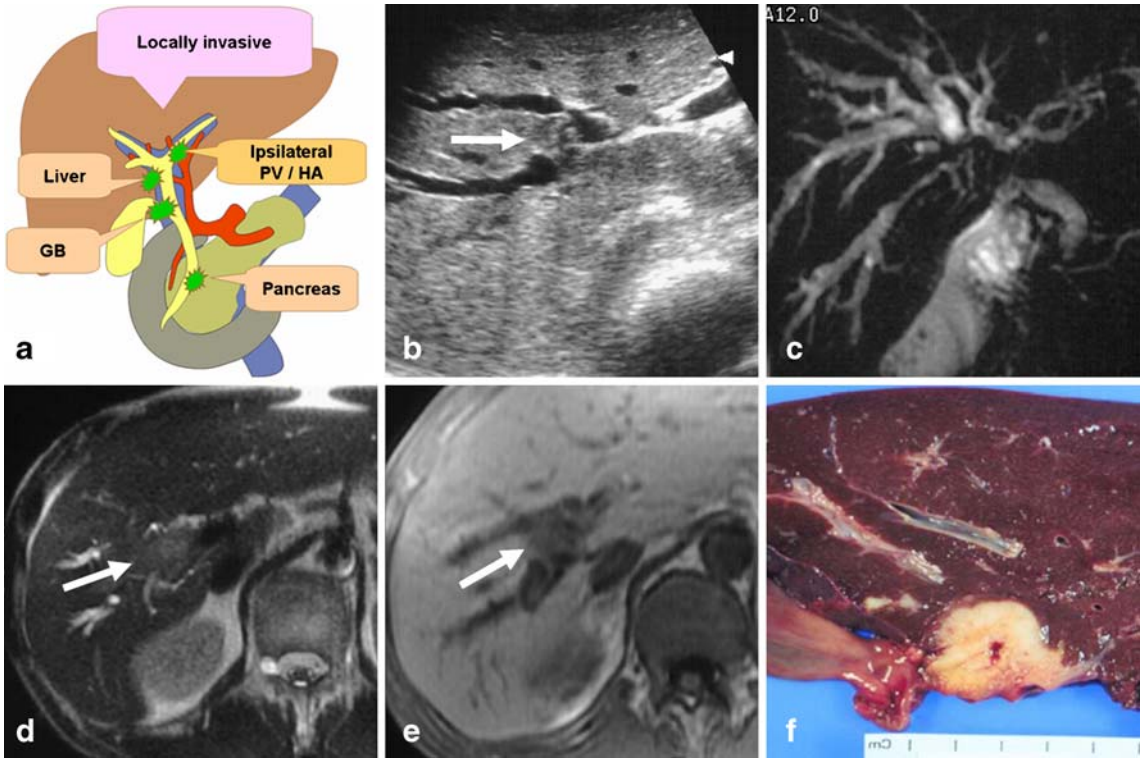

Fig. 13 **a** Drawing illustrates T3 extrahepatic cholangiocarcinoma invading the liver. **b** Sonography shows a hypoechoic mass around the hilum (arrow) with intrahepatic ductal dilation. **c** MRCP, **d** transverse T2-weighted, and **e** T1-weighted MR images show a hilar mass with adjacent liver invasion. **f** Photograph of the gross specimen shows a nodular lesion invading the liver, which agrees with the findings of sonography and MR

There is also some problem in classifying T3 and T4 stages. In the TNM system, the same criteria are used for staging from proximal to distal EBD cancer. It appears reasonable to classify both proximal cancer involving liver and distal cancer involving the pancreas into the same category of T3. However, if a cancer in the middle third of EBD extends to the proximal or distal EBD and invades the liver or pancreas, then this cancer is also classified as T3, although the actual lateral invasion depth may be far more advanced than those of the proximal or distal EBD cancers invading the adjacent organs. In addition, with regard to the invasion of the main PV or duodenum, invasion of these structures indicates T4 tumor by definition (Fig. 17). However, in current surgical management, a focal invasion of less than 2 cm in length of the main PV may not be a contraindication for the curative resection and could be the indication for curative resection of involved portal vein with venous graft placement [19, 29–31]. Meanwhile, tumors of the middle CBD origin extending to the liver or pancreas at the T3 stage may actually be more widely extended than those

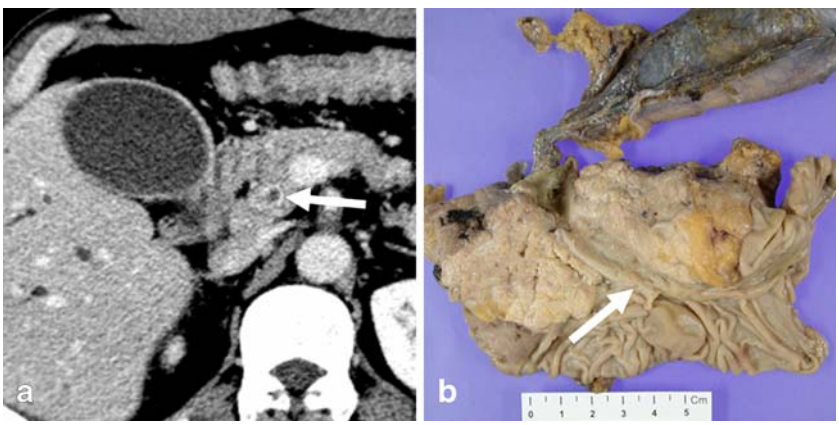

Fig. 14 **a** On contrast-enhanced CT, ductal wall thickening with increased enhancement is seen in the distal bile duct. **b** Photograph of gross specimen shows ductal wall thickening with luminal narrowing. Microscopic view shows that the tumor infiltrates the surrounding pancreas (not shown). Extrahepatic cholangiocarcinomas in the distal bile duct are frequently T3 lesions when they are depicted on CT or MRI

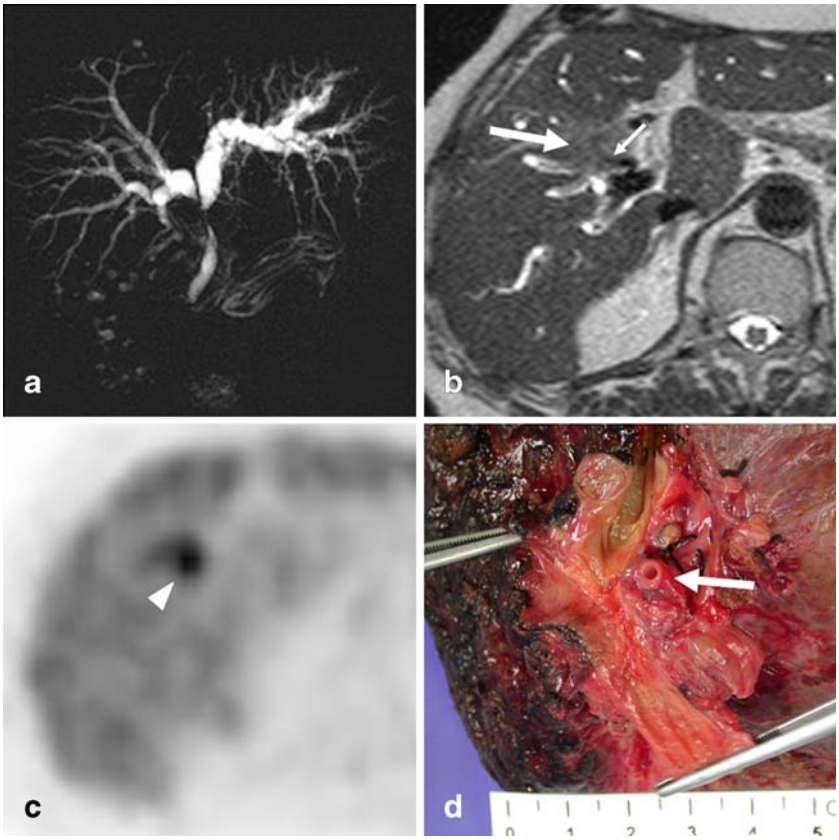

Fig. 15 **a** MRCP shows a hilar cholangiocarcinoma invading the confluence of the right intrahepatic bile duct (Bismuth Corlette type IIIa). **b** T2-weighted transverse MR image shows a hilar mass (arrow) invading the liver and encasing the right hepatic artery (small arrow). **c** FDG-PET shows increased uptake in the hilar mass (arrowhead). **d** Photograph of a gross specimen shows a hilar cholangiocarcinoma with liver and right hepatic artery invasion (arrow)

tumors and may be unresectable. Therefore, stage T3 does not necessarily mean less invasive than stage T4 in this condition.

Evaluation of resectability

Determination of resectability can be helped by considering Blumgart T-staging system (Table 2) [32].

Although the descriptions might seem quite complex, they can be simplified as in the illustration (Fig. 19). In this T staging system, T3 tumors corresponding to the Bismuth-Corlette type 4, bilateral, or main portal vein invasion, or Bismuth-Corlette type 3 tumors with contralateral portal vein invasion or hepatic atrophy are considered poor candidates for surgery. However, some Bismuth type 4 tumors with favorable anatomy or short segmental invasion

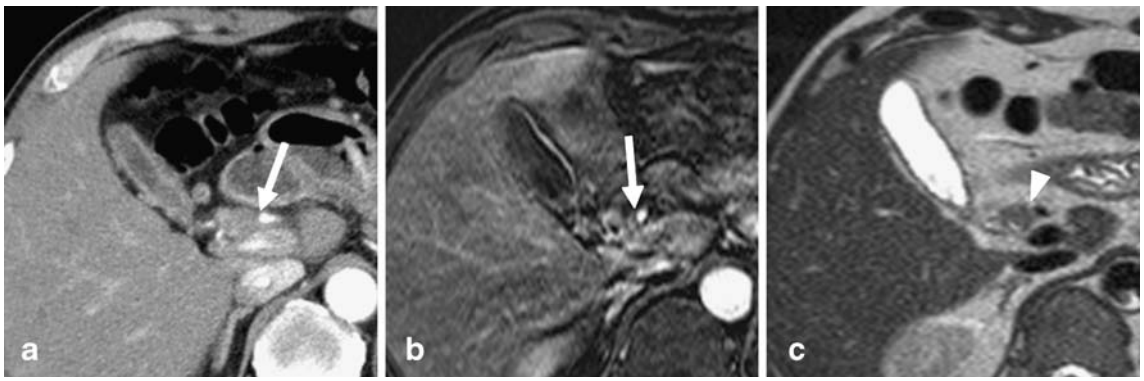

Fig. 16 **a** Contrast-enhanced CT and **b** MR images taken on arterial phase show obliteration of the fat plane around the proper hepatic artery, but the fat plane is seen preserved on **c** high-resolution T2-weighted imaging. Surgical findings confirmed the absence of vascular involvement

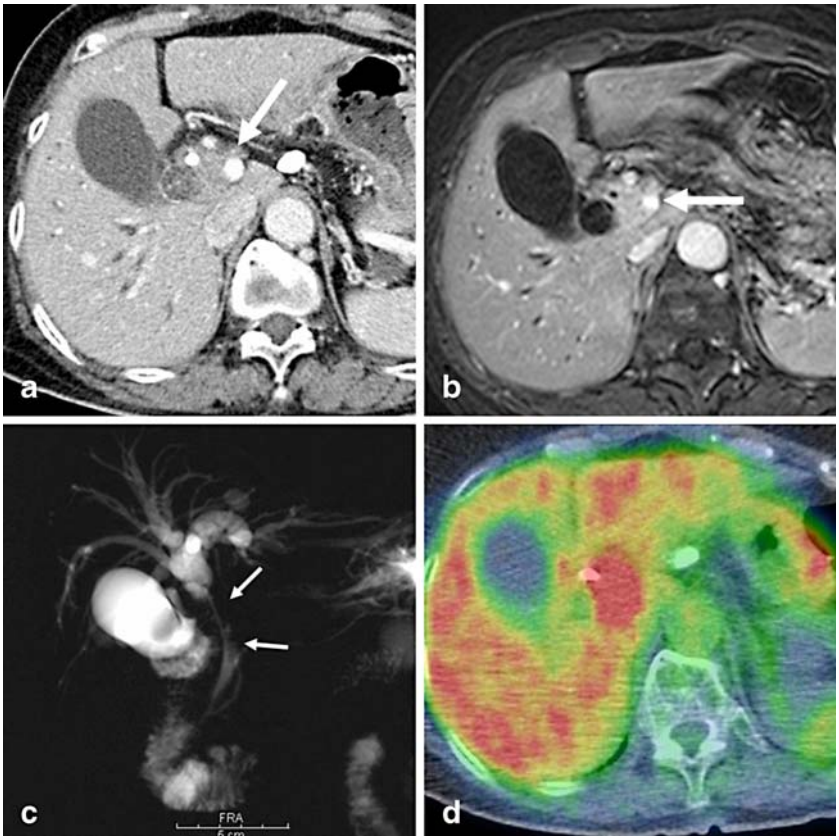

Fig. 17 **a** On contrast-enhanced CT and **b** MR images taken on portal venous phase, the tumor is abutting on main portal vein more than 180 degree (arrows), suggesting main portal vein invasion. **c** MRCP shows luminal narrowing (small arrows) in the proximal CBD without hilar involvement (Bismuth Correlet type IV). **d** PET-CT image shows increased FDG uptake in the bile duct tumor abutting on main portal vein

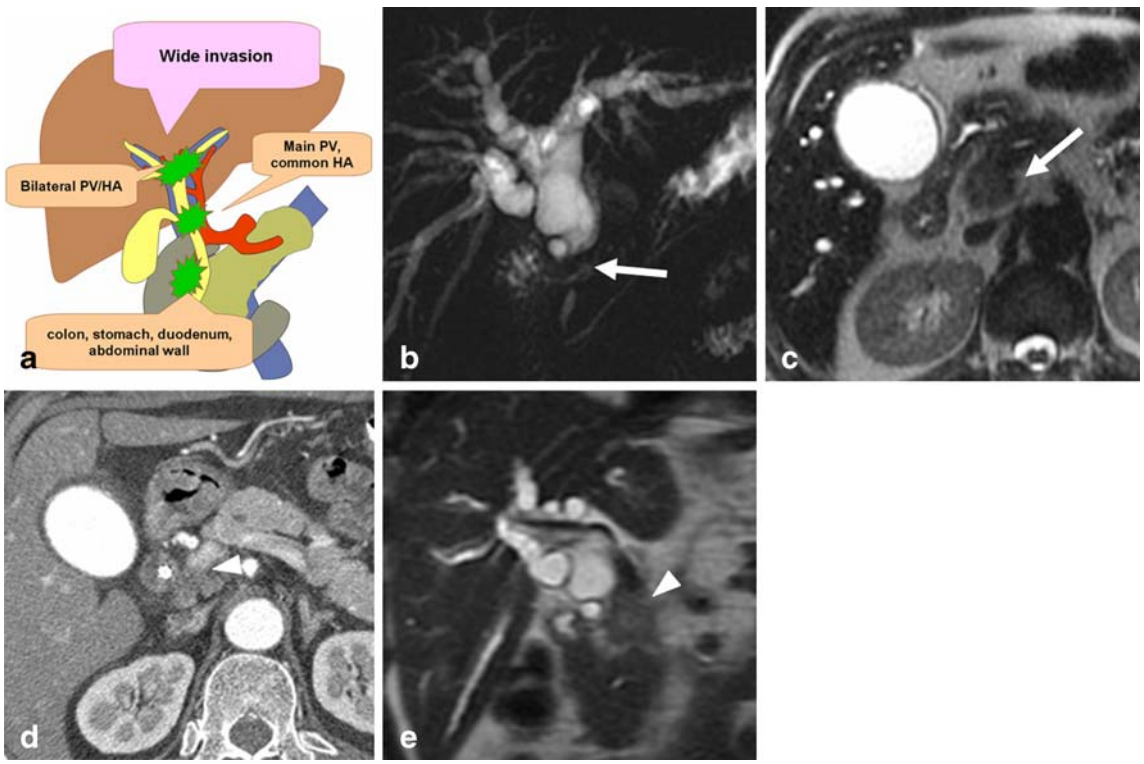

Fig. 18 **a** Drawing illustrates T4 extrahepatic cholangiocarcinoma invading the main portal vein. **b** MRCP shows a bile duct obstruction due to a tumor (arrow). **c** Transverse T2-weighted imaging shows an irregular nodular mass in the common bile duct (arrow). **d** Contrast-enhanced CT and **e** coronal T2-weighted MR images show a tumor invading the main portal vein (arrowheads)

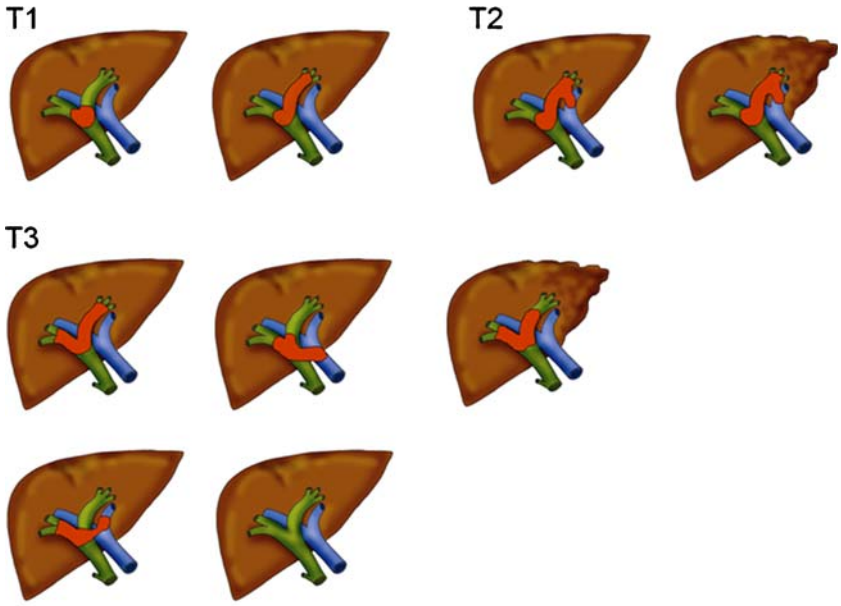

Fig. 19 Drawing illustrates Blumgart T-staging system of hilar cholangiocarcinoma

of main PV may not be an absolute contraindication to attempt curative resection [19, 29, 30]. This staging system corresponds to what can be assessed by evaluating the longitudinal and lateral spread pattern based on the Bismuth classification and TNM staging system.

According to the Sixth AJCC Cancer Staging System, N1 is defined as regional lymph node metastasis (Table 3). The regional lymph nodes include hilar, celiac, periduodenal, peripancratic, and superior mesenteric lymph nodes [22]. The lymph node numbering system proposed by the Japanese Research Society for Gastric Cancer is being used by surgeons and pathologists [33]. However, identification of each numbering group of the lymph nodes on cross-sectional imaging is difficult and is not practical. The proposed CT criteria for identification of metastatic lymph node are as follows: larger than 10 mm in short-axis diameter, presence of central necrosis, and hyperattneuating the liver in the portal phase (Fig. 20) [19]. The sensitivity and specificity for nodal involvement are not so high on current cross-sectional imaging, and false-negative cases are frequently encountered on CT, MRI, or FDG-PET [34–36]. Therefore, lymph node status cannot be reliably determined on the current imaging system, and the presence of equivocal lymph nodes cannot be used as a criterion for unresectability. Some surgeons prefer paraaortic lymph node sampling before proceeding to an intended curative resection [37, 38].

Table 3 Regional lymph nodes (N) and distant metastasis (M) staging (AJCC 6th edn)

Regional lymph nodes (N)	
NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Regional lymph node metastasis
Distant metastasis (M)	
MX	Distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis

In terms of distant metastases, unexpected metastases are becoming less commonly seen with the advance of CT/MRI, and FDG-PET and diagnostic laparoscopy may play a complementary role. It is generally accepted that a FDG-PET is useful to detect distant metastases and may lead to change in management in up to 30% of patients (Fig. 20) [39]. Although a FDG-PET is not regarded as suitable for the detection of regional LN metastasis [35], it may provide higher specificity compared with a CT [40] (Fig. 21).

Staging laparoscopy may be another useful method when resectability of the EBD cancer cannot be determined from the imaging study. Diagnostic laparoscopy can reduce unnecessary laparotomy, but it has limitations in evaluating vascular or nodal involvement, especially in hilar cholangiocarcinoma [41]. One long-term study in the UK showed that laparoscopy detected unresectable cases in 42% of presumed resectable hilar cholangiocarcinomas. Among the patients who underwent laparotomy, 51% were actually resectable, and the overall sensitivity for unresectability was 65% [42].

Resectability also depends on the patient factor. Patient factor is essential for operation even in technically feasible case. Curative intend surgery cannot be performed in patients who have comorbidity and cannot tolerate a major operation [43]. Patient age also affects the decision for

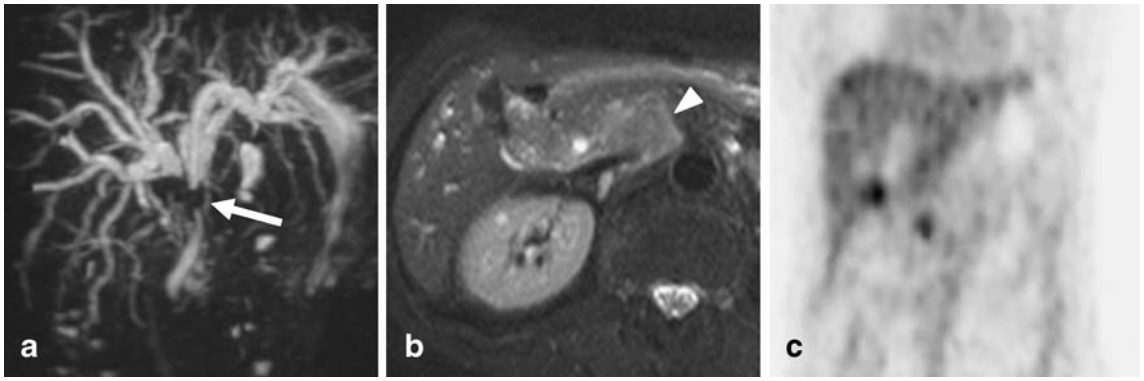

Fig. 20 **a** MRCP shows Bismuth-Corlette type IIIa cancer (arrow). **b** Axial T2-weighted MR image shows an enlarged lymph node in the retropancraetic area (arrowhead: number 13 lymph node by numbering system proposed by the Japanese Research Society for Gastric Cancer). **c** FDG-PET shows increased uptake both in primary hilar cancer and the retropancreatic lymph node

operation, but recently, old age has not been considered as an absolute contraindication for the major operation [44].

Conclusion

In summary, longitudinal extension can be assessed according to the Bismuth classification. For this purpose, anatomical variation should also be considered and well documented. Lateral extension in terms of soft tissue extension and vascular invasion can be assessed based on T staging, but the T stage itself may not reflect the actual depth of extension. Further study to reduce the ambiguity of T staging is necessary. To determine the resectability of the tumor, evaluation of a tumor based on the Blumgart T-staging system can be useful.

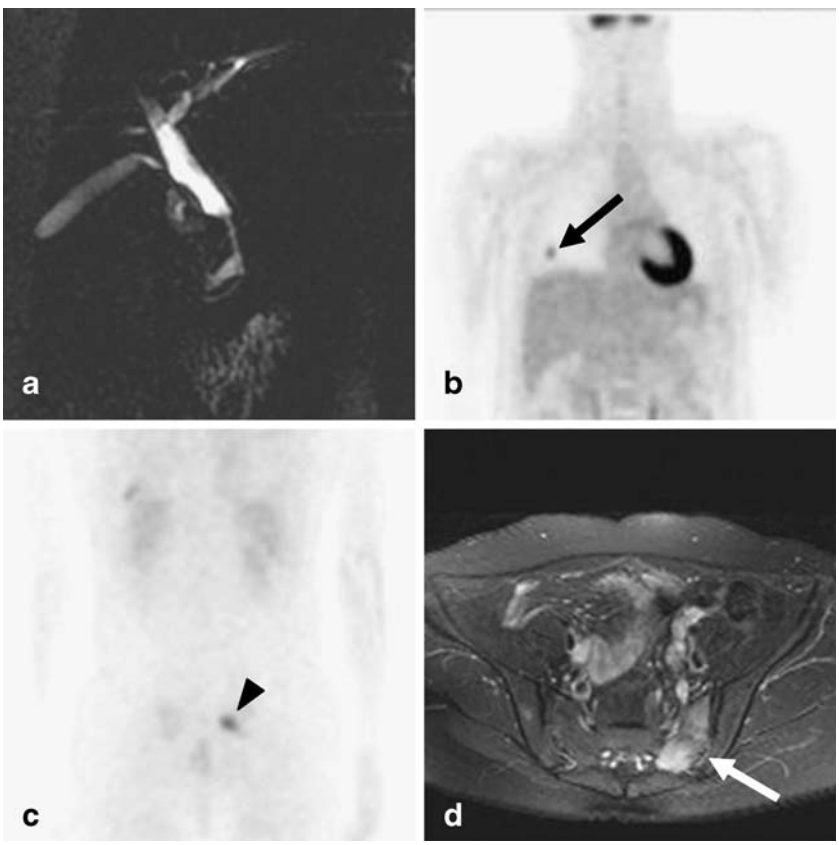

Fig. 21 **a** MRCP shows distal EBD cancer. FDG PET depicted two unsuspected metastases **b** in the right lower lung (arrow) and **c** left sacral bone (arrowhead). **d** Axial T2-weighted MR imaging shows bone metastasis in the sacrum (arrow)

The N and M staging accuracy of CT and MR is still not sufficiently high. FDG-PET and diagnostic laparoscopy may help with diagnosis, especially for detection of distant metastasis.

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