Liver Cysts:
A Practical Approach

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1) Learning Objectives

- Liver cysts are a common radiological finding
- The majority will be simple (biliary) cysts that are easily characterised, although a minority will not resulting in diagnostic uncertainty
- It is therefore imperative that radiologists have a sound understanding of liver cysts
- This poster presentation will explore the different features of liver cysts that will aid their diagnosis

2) Background

- Liver cysts can be divided into congenital, neoplastic, inflammatory, infective, post-traumatic and iatrogenic
- A crucial process in narrowing the differential is to determine the presence or absence of complex features
- The different types of complexity found in liver cysts can often be linked to a specific pathological process
- Figure 1 demonstrates some of the complex features found in liver cysts

2) Background

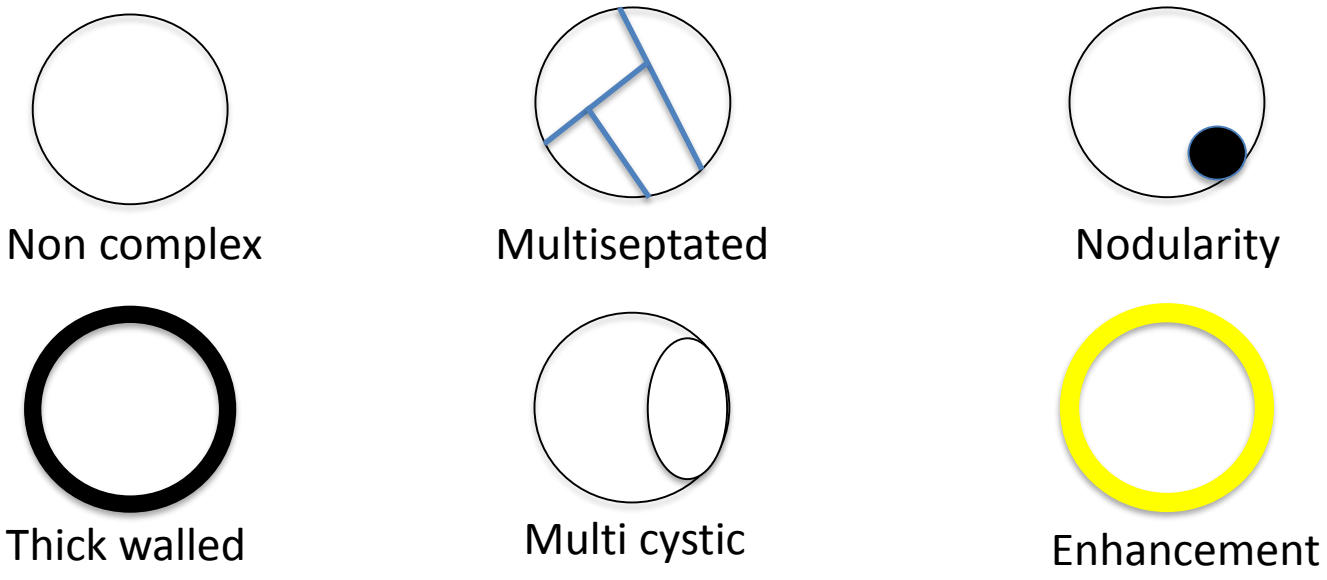

Fig 1: Schematic diagram of the most common types of complexity found in cystic liver lesions.

Adapted from Claret Loaiza S et al EPSO ECR 2014

2) Background

- Due to overlapping radiological features of some liver cysts, it is often necessary to integrate the imaging with additional information

- Pertinent information such as **clinical presentation** and **past medical history** is often helpful in the formation of a differential diagnosis

- Laboratory findings such as raised **inflammatory markers** and positive **serology markers** can also help confirm the presence of an infective or inflammatory process

2) Background

- Table 1 summarises the various types of liver cysts with their associated radiological, clinical and laboratory findings
- A more detailed assessment of these liver cysts with imaging will be presented in the next section
- It is not intended to be an exhaustive list, but highlight some of the more important and interesting types of cysts
- The imaging presented has been collated from a tertiary liver unit within the United Kingdom

2) Background

Subtype	Clinical feature	Imaging
Congenital (non-ductal plate malformations)		
Simple biliary cyst	Asymptotic	Homogenous, round, regular, no wall, no enhancement
Ciliated hepatic foregut cyst	Asymptomatic. Occurs more commonly in women	Solitary cystic lesion in segment 4 of the liver with variable complex features
Congenital (ductal-plate malformations)		
Bile duct hamartoma	Asymptomatic	Homogenous cysts less than 1.5cm with irregular walls. No enhancement
ADPLD	Can develop abdominal pain if liver capsule becomes stretched	Multiple, homogenous, round, regular, no wall, no enhancement
Caroli disease	Can present with cholangitis	Cysts with connection to the biliary tree. Presence of a central dot
Choledochal cysts	Can present with abdominal pain and jaundice	Cystic or fusiform dilatation of the intra or extra hepatic ducts depending on subtype

*Autosomal dominant polycystic liver disease

Fig 2: Clinical and imaging features of cystic liver lesions.
Adapted from Mortele KJ et al Radiographics 2001 21, 895-910

2) Background

Subtype	Clinical feature	Imaging
Neoplastic		
Cystic metastasis	Known primary cancer	Multiple rim enhancing lesions
Cystadenoma and Cystadenocarcinoma	Occurs more commonly in middle aged women	Usually large, solitary, multilocular, mural nodules, fibrous capsule, calcification
Embryonal sarcoma	Occurs in adolescent children and young adults	Large solitary cyst with enhancing solid components and calcification
Infective / inflammatory		
Abscess	Sepsis. Raised WCC and CRP	Presence of air, double target sign, enhancing wall
Hydatid cysts	Recent travel to endemic areas such as South America and Central Asia. Positive serology and eosinophilia	Calcification, daughter cysts, pericysts

Fig 2: Clinical and imaging features of cystic liver lesions.
Adapted from Mortele KJ et al Radiographics 2001 21, 895-910

2) Background

Subtype	Clinical feature	Imaging
Post Traumatic / Iatrogenic		
Haematoma	Trauma / surgery	Fluid density on CT imaging. This can change to high attenuation on unenhanced CT scans. Met-Hb on MRI, no enhancement, signs of trauma
Biloma	Trauma / surgery	Fluid on CT and MRI, no capsule, no septa, no calcification
Miscellaneous		
Parabillary cysts	Asymptomatic finding in cirrhotic patients, particularly those with alcohol related liver disease	Multiple simple looking cysts adjacent to the biliary vessels

Fig 2: Clinical and imaging features of cystic liver lesions.
Adapted from Mortele KJ et al Radiographics 2001 21, 895-910

3) Imaging Findings: Simple biliary Cysts

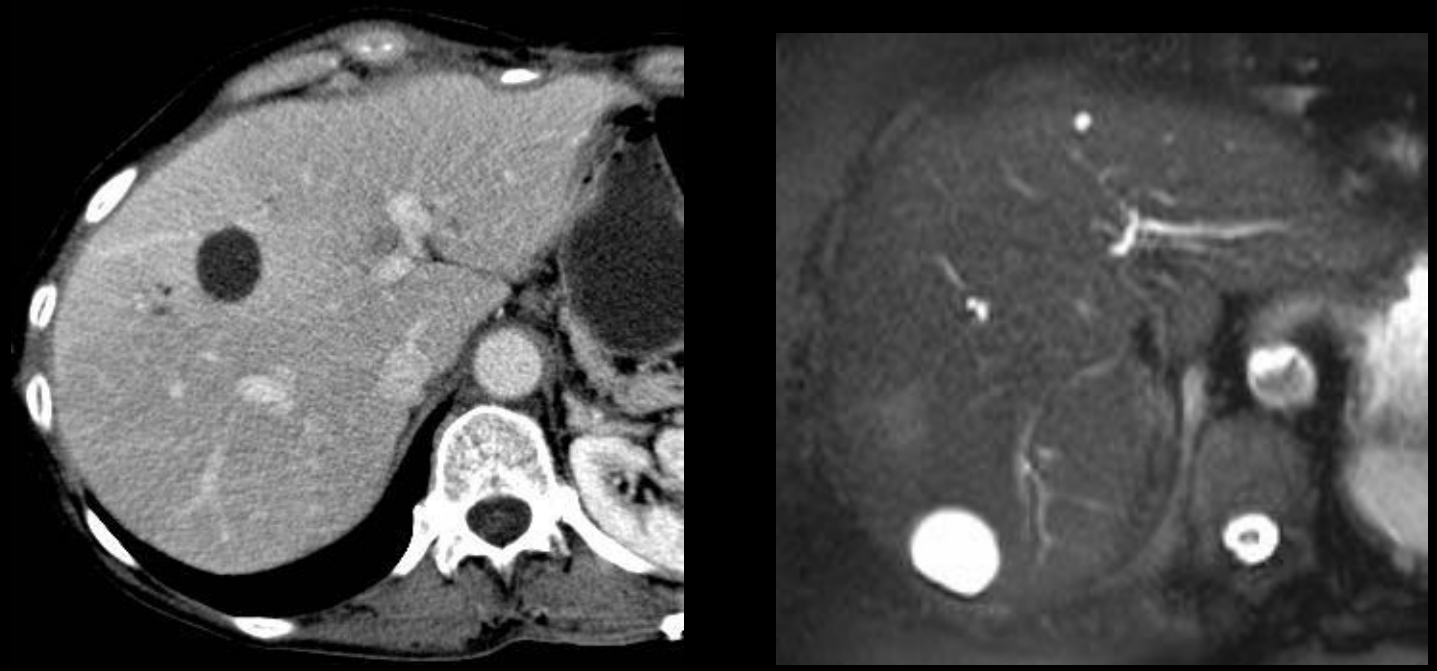

Fig 3: PV Ph CT scan and HASTE FS MRI of different patients with a simple cyst in the right liver lobe. Simple cysts are typically homogenous, well defined and non enhancing.

Simple biliary cysts arise from the biliary endothelium with no communication with the biliary tree. They occur more often in middle aged women. The lesions are benign and almost always asymptomatic. On CT cysts are well defined with homogenous low attenuation. At MRI they have homogenously low T1 and high T2 signal. The T2 signal becomes more intense on heavily weight T2 imaging, which helps differentiate them from cystic liver metastases. No enhancement is seen on either imaging modality.

3) Imaging Findings: Ciliated Hepatic Foregut Cysts (CHFC)

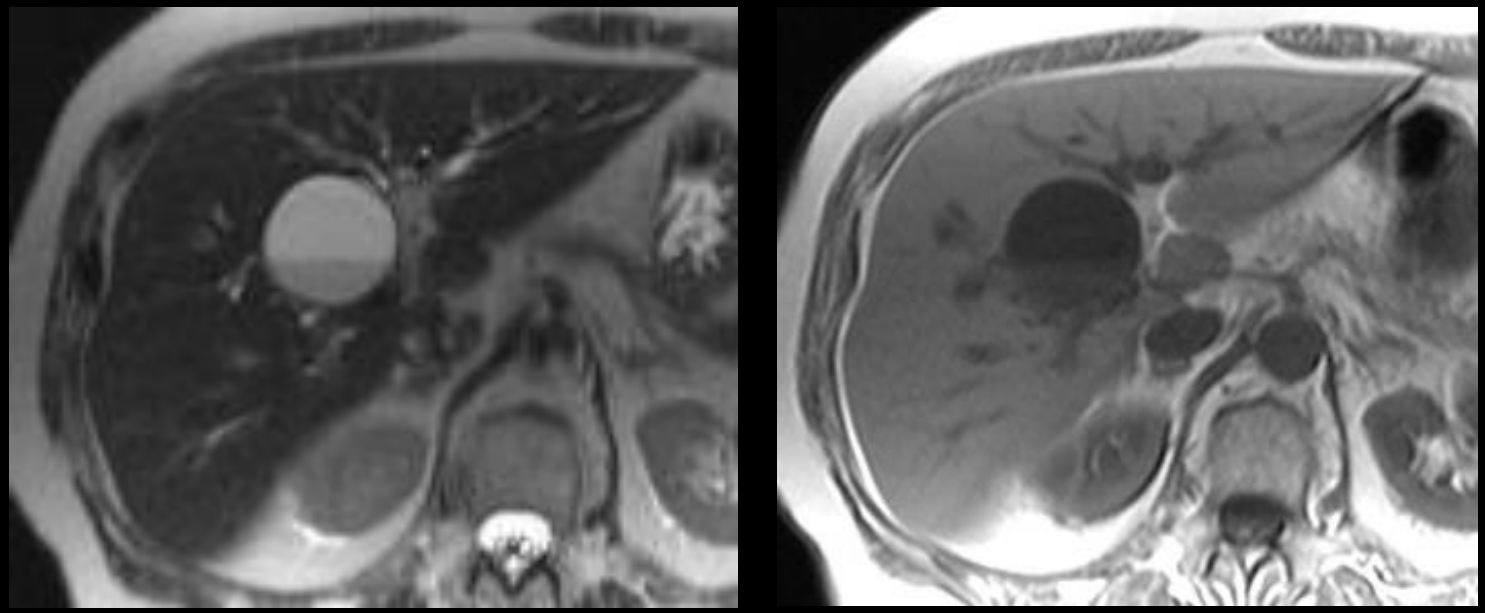

Fig 4: HASTE and In Ph GRE T1 MRI of a CHFC arising from a typical location within segment 4 of the liver. Note the fluid filled appearance, which can often mimic a simple cyst with haemorrhage.

CHFC is a rare lesion that predominately affects women. Histologically it is similar to a bronchogenic cyst and has no connection with the biliary tree. It is often an incidental finding as patients are typically asymptomatic. Very infrequently it can undergo malignant transformation by squamous metaplasia of its epithelial lining. It usually appears as a simple cyst on CT imaging, but can have fluid filled levels on MRI due to its proteinaceous content. With contrast there is no enhancement on either CT and MRI.

3) Imaging Findings: Ductal-plate malformations
A cause of liver cysts

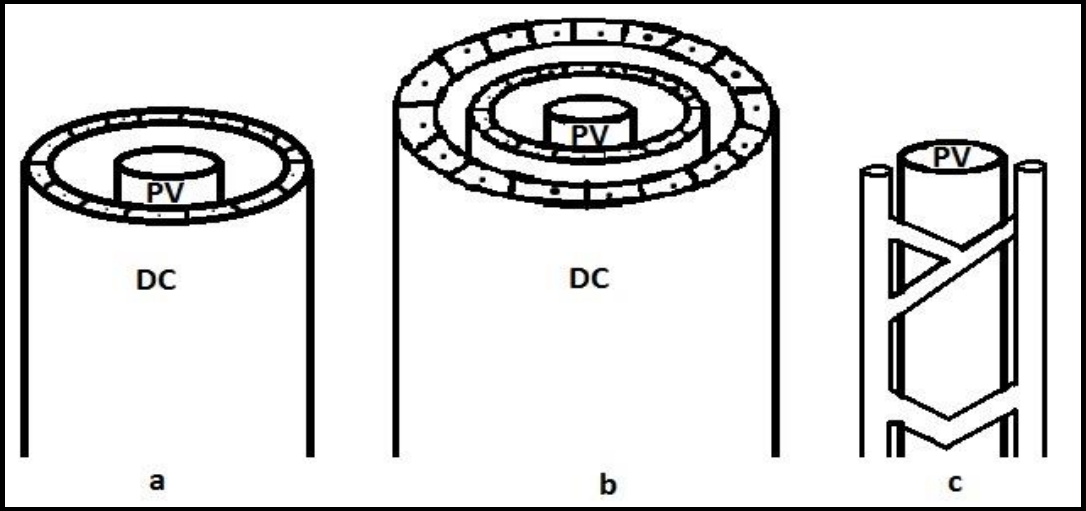

Fig 5: Normal ductal plate formation. A single layer of ductal cells surround a portal vein (a). It is followed by the formation of a second layer creating slit-like primitive bile duct lumen (b). This undergoes reabsorption to form the fine bile ducts surrounding the portal vein (c).
Adapted from Brancatelli G et al. Radiographics 2005 25, 659-670.

Ductal plate malformations occur when there is insufficient reabsorption of the ductal plates during embryogenesis, which go on to form the biliary tree (see fig 5). This leads to a large spectrum of liver disorders / cysts depending on the affected interlobar bile duct. Congenital hepatic fibrosis, bile duct hamartoma and ADPLD arise from malformation of the small to medium interlobar bile ducts. Whereas Caroli disease and choledochal cysts arise from the malformation of the larger ducts. The next six slides will explore the cystic manifestations of this disorder in more detail.

3) Imaging Findings: Ductal-plate malformations
Bile Duct Hamartoma

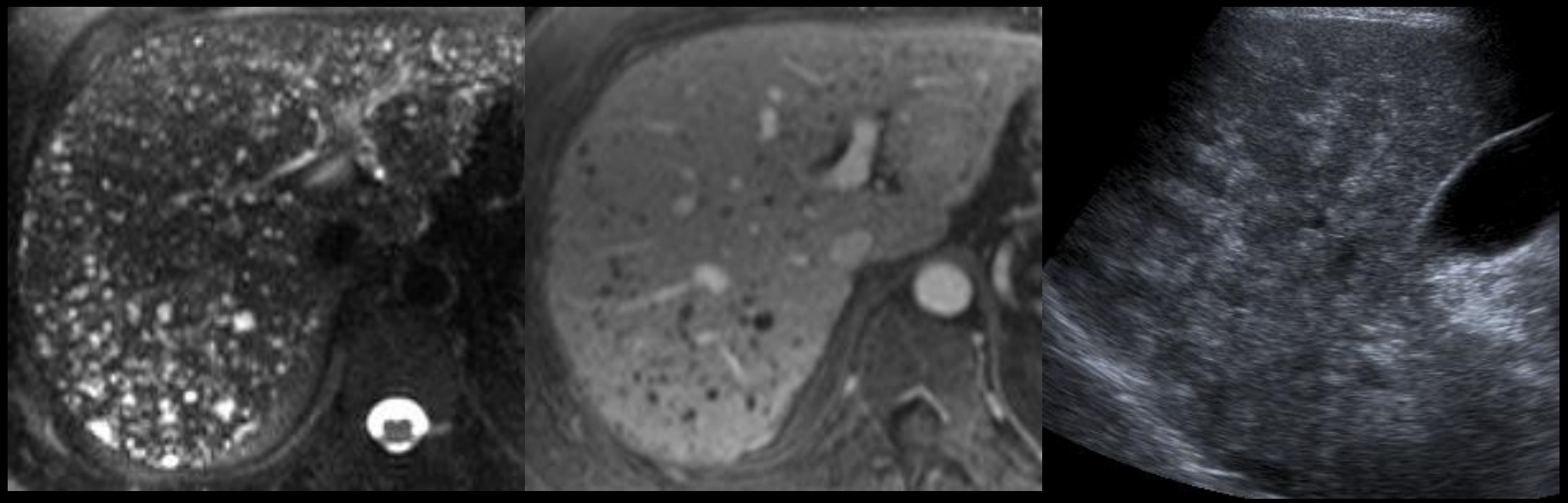

Fig 6: T2 FS, PV Ph MRI and US scan of the liver containing innumerable hamartoma giving a "Starry Sky" appearance. Note their irregular contour and hyperintense appearance on the T2 sequence.

Bile duct hamartoma, or von Meyenburg complex, are composed of dilated duct like structures lined by biliary epithelium and accompanied by a variable amount of fibrous stroma. CT imaging shows multiple hypoattenuating, almost cyst like, nodules throughout both lobes of the liver. They can be distinguished from simple liver cysts due to their size (<1.5cm) and irregular contour. On MRI imaging the nodules are strongly hyperintense on T2 and hypointense on T1 sequences with no connection to the biliary tree. The hamartoma appear more solid / echogenic on ultrasound with associated comet tail artefact.

3) Imaging Findings: Ductal-plate malformations
Polycystic liver disease (ADPLD)

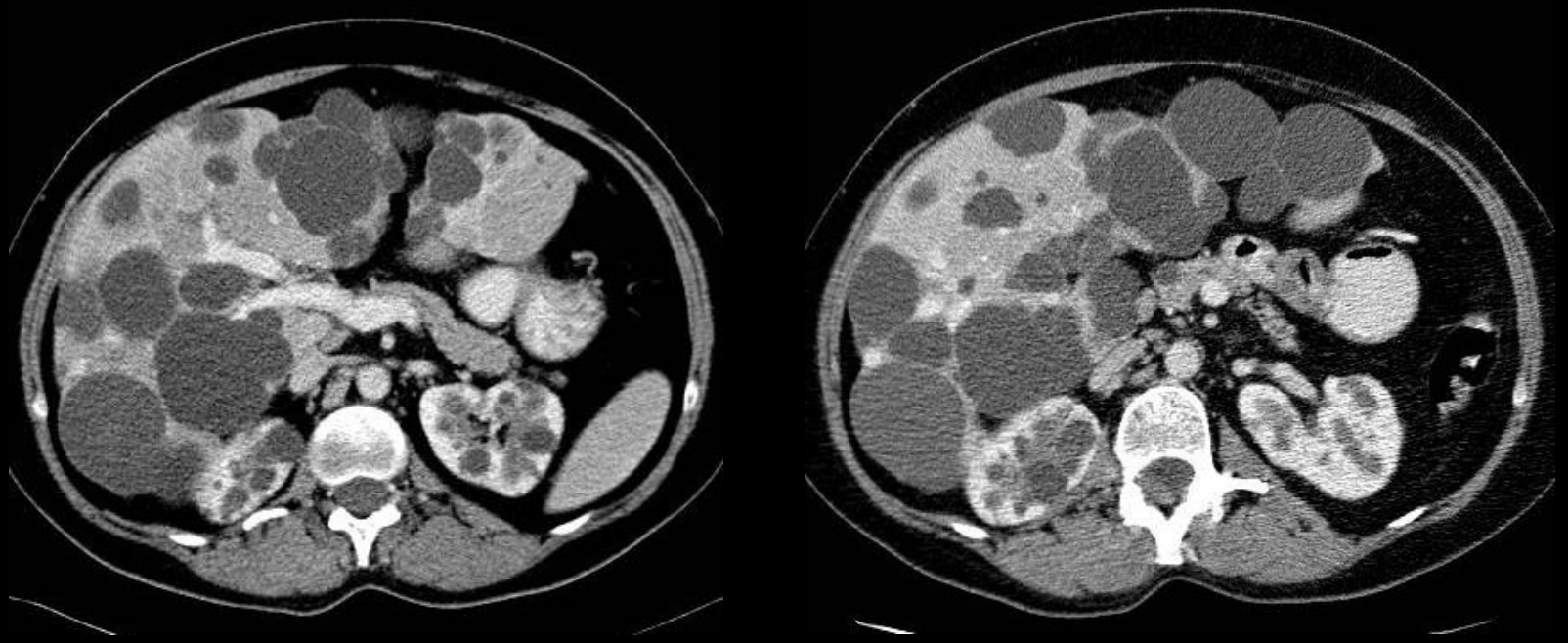

Fig 7: PV Ph CT scan of the liver and kidneys containing numerous simple cysts.

ADPLD results from the progressive dilatation of the abnormal ducts in biliary hamartoma, which loose continuity with the biliary tree. Hepatic disease is present in up to 40% of patient's with ADPKD, but may also occur in the absence of renal cysts. Patients can experience pain if the liver capsule is stretched, and in advanced disease, can develop hepatomegaly, liver failure or Budd Chari syndrome. Appearances are similar to simple biliary cysts with homogenous low attenuation on CT, and low T1 and high T2 signal on MRI imaging without enhancement. Cysts complications such as haemorrhage or infection can occur.

3) Imaging Findings: Ductal-plate malformations
Caroli Disease

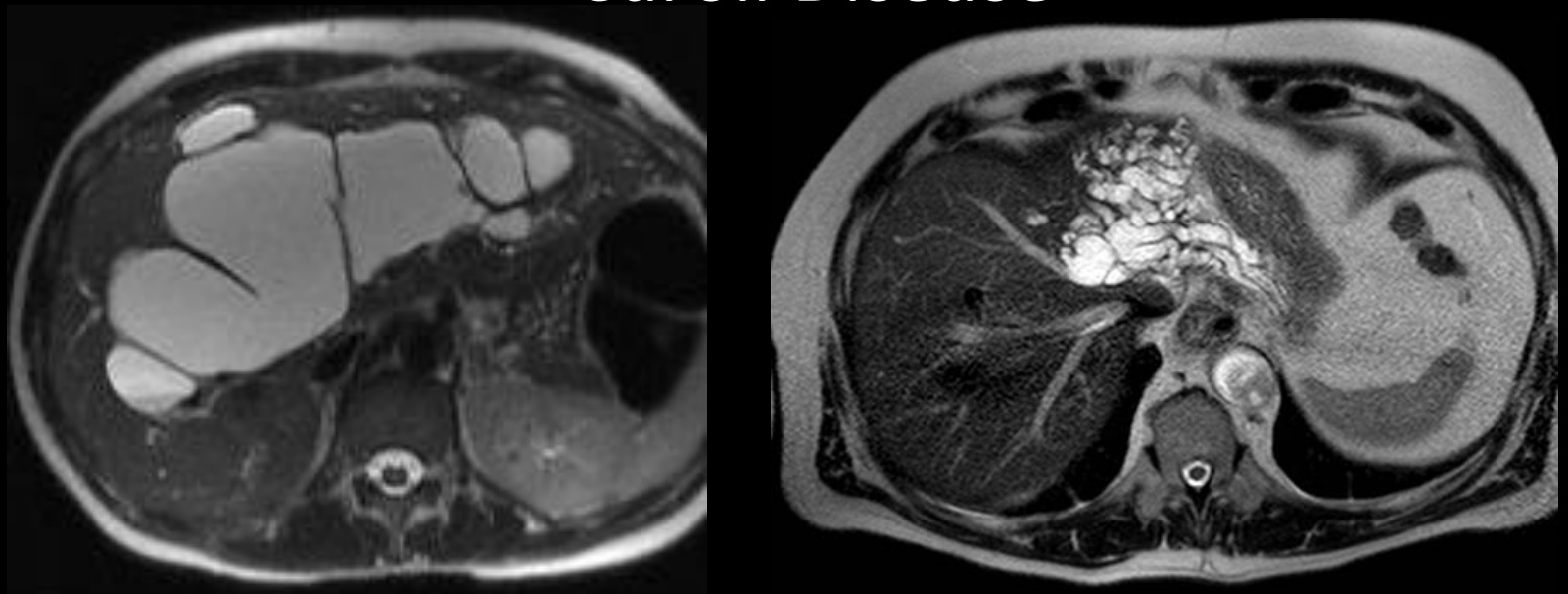

Fig 8: HASTE MRI of the liver of two cases of Caroli disease. The first image shows gross saccular dilatations of the biliary tree replacing large portions of both liver lobes. Whereas the second image shows less pronounced duct dilated but more focal involvement and atrophy of the left lobe.

Caroli disease is an autosomal recessive condition that manifests in childhood or adolescence. It exists in two forms. Caroli disease "proper" and Caroli "syndrome". The former results from malformation of the ductal plates of the larger intra hepatic ducts. It is characterised by focal or diffuse saccular dilatation of the intra-hepatic ducts. It can be associated with intrahepatic stone formation, cholangitis and abscess formation. Whereas Caroli syndrome arises from abnormalities of the more peripheral ducts, which results in peri portal fibrosis and cirrhosis.

3) Imaging Findings: Ductal-plate malformations
Caroli Disease

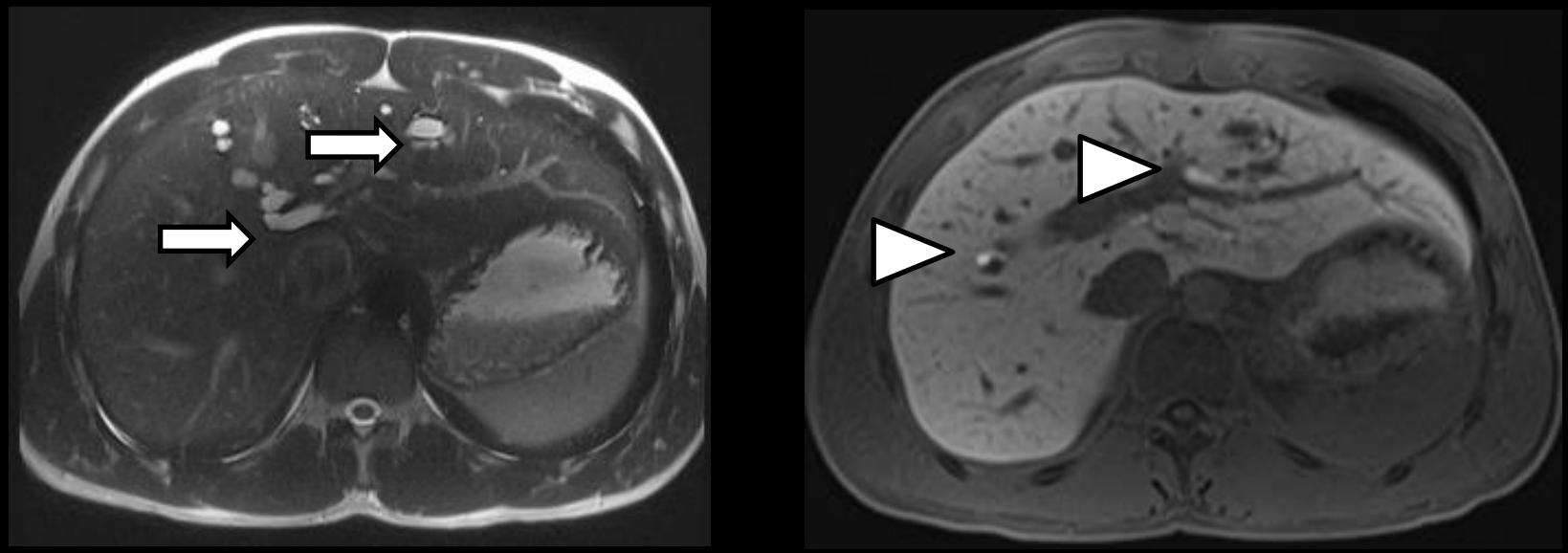

Fig 9: HASTE and HPB Ph MRI of the liver with Caroli disease. The images show focal saccular dilatation of the biliary tree (straight arrows), which accumulates hepatobiliary-specific contrast during the HPB phase (arrow heads) confirming its communication with the biliary tree.

Caroli disease proper is typically hypo-attenuating on CT imaging and have homogeneous low T1 and high T2 signal on MRI imaging. With the addition of a hepatobiliary-specific contrast agent the saccular dilatations can accumulate contrast confirming its communication with the biliary tree (see fig 9). A finding that is very suggestive of Caroli disease is the “central dot sign”, which represents contrast enhancement of a portal vein radical within the saccular dilatation. If the disease is localised, the treatment of choice is surgical resection.

3) Imaging Findings: Ductal-plate malformations
Choledochal Cysts

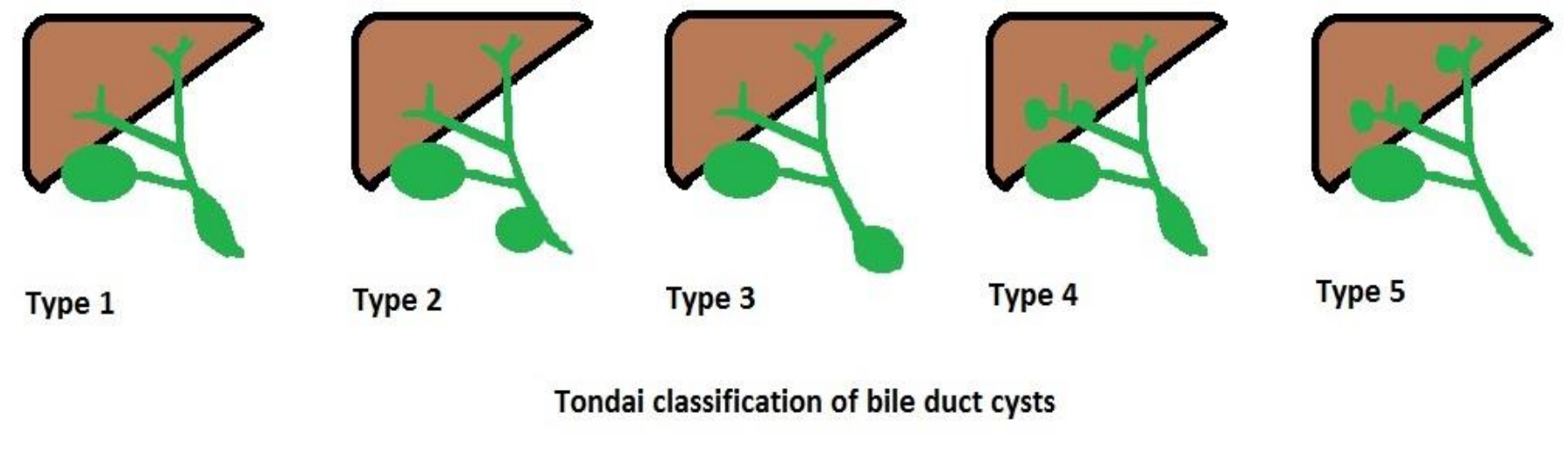

Fig 10: Tondai Classification: Type 1 dilatation of the extrahepatic duct. Type 2 diverticulum of the extrahepatic duct. Type 3 dilatation of extrahepatic duct within duodenal wall. Type 4 dilatation of intra and extra hepatic ducts. Type 5 multiple dilatations/cysts of intrahepatic ducts only.

Choledochal cysts represent cystic or fusiform dilatation of the biliary tree. They are extremely rare and and can present with abdominal pain and jaundice. The exact aetiology of choledochal cysts remains unclear. Many believe it is part of the ductal plate malformations due to its association with other intra- and extra hepatic duct disorders. Alternatively it could be secondary to reflux of pancreatic enzymes into the biliary tree from a congenitally abnormal pancreaticobiliary junction. Choledochal cysts are divided into five types based on the Todani classification (see fig 10).

3) Imaging Findings: Ductal-plate malformations
Choledochal Cysts

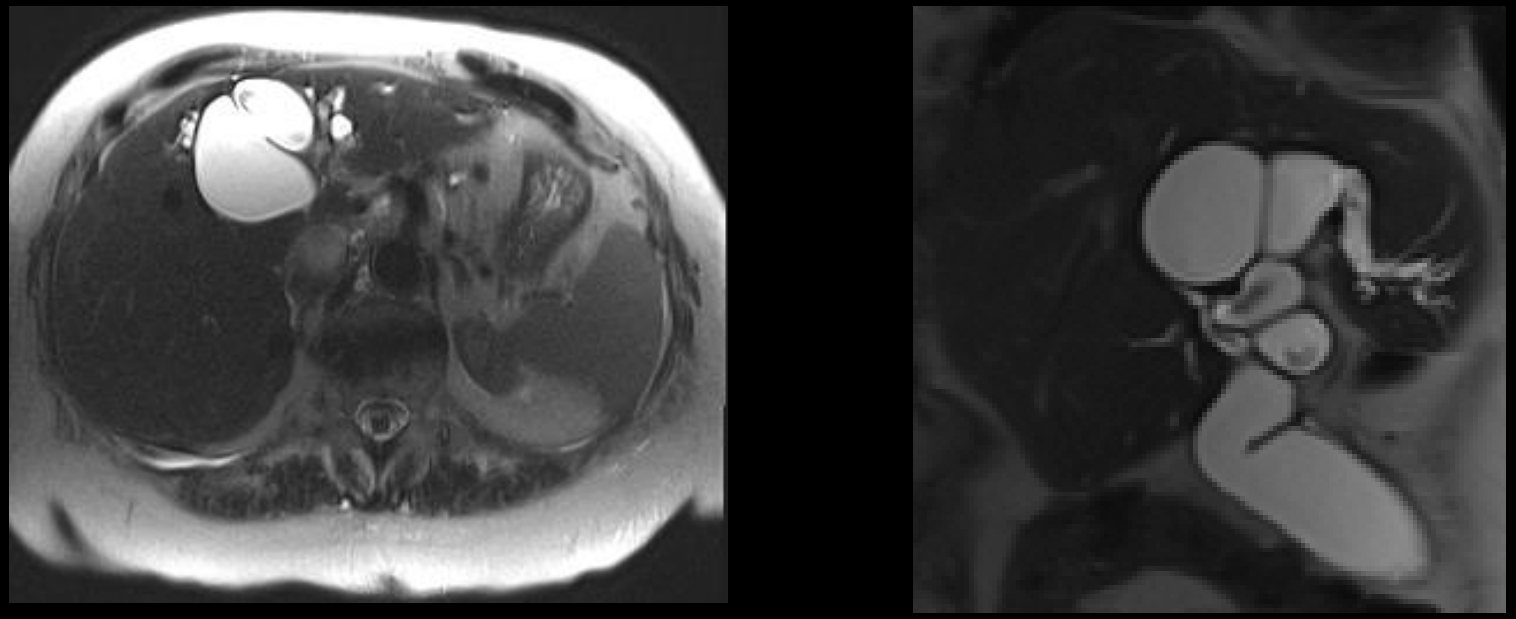

Fig 11: Axial and coronal HASTE MR of the liver and biliary tree demonstrating a Type 4 choledochal cyst of the CBD and intra-hepatic biliary tree.

On imaging, choledochal cysts appear as cystic or fusiform dilatation of the intra or extra hepatic biliary tree. They vary in size, with larger cysts containing up to 5-10 litres of bile. Cysts are best demonstrated on MRCP and have homogenous high T2 signal unless it has been complicated by stone formation. Due to an increased risk of cancer, choledochal cysts are usually surgically removed.

3) Imaging Findings: Cystic Metastases
Necrosis and cystic degeneration

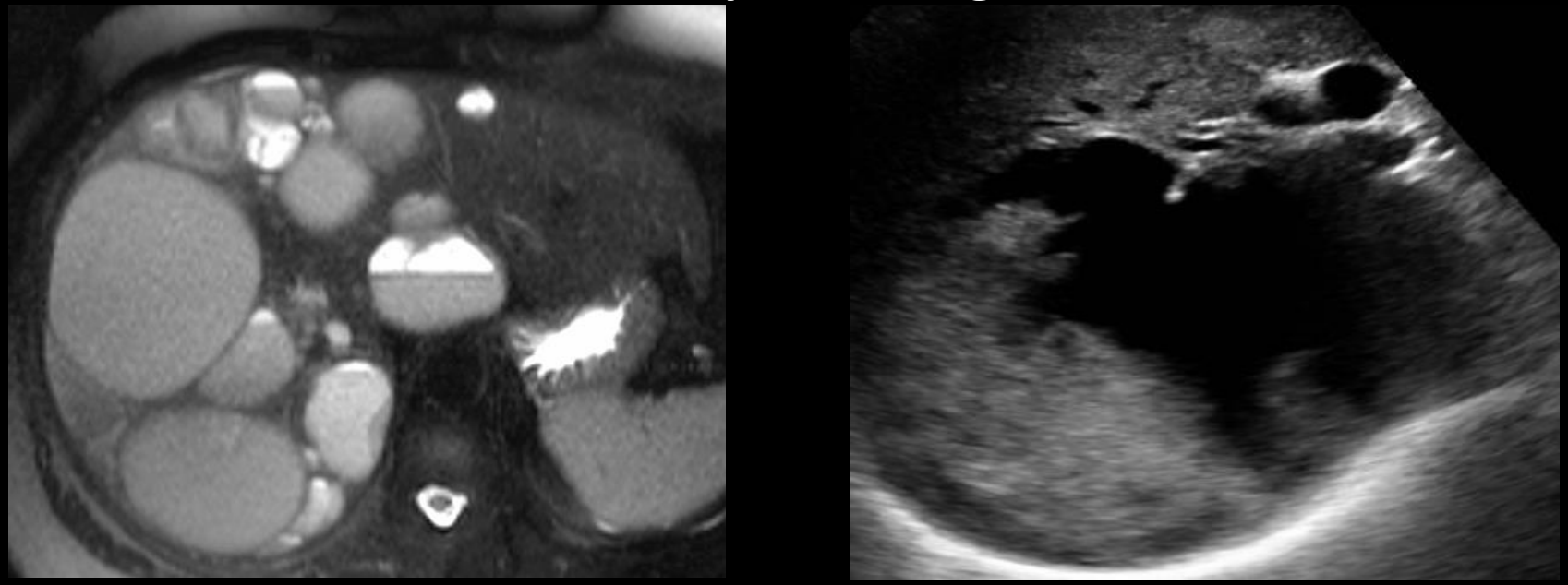

Fig 12: HASTE FS MR and US of a liver containing neuroendocrine metastases with internal cystic change due to necrosis and haemorrhage giving a fluid level appearance.

Most liver metastases are solid, but some can have partial or a complete cystic appearance. There are two main pathological processes that can cause this manifestation. The first is due to necrosis and cystic degeneration of a rapidly growing tumour such as melanoma, sarcoma or a neuroendocrine tumour (see fig 12). These lesions quickly outgrow their blood supply and can produce fluid – fluid levels when accompanied by haemorrhage.

3) Imaging Findings: Cystic Metastases
Mucinous adenocarcinoma

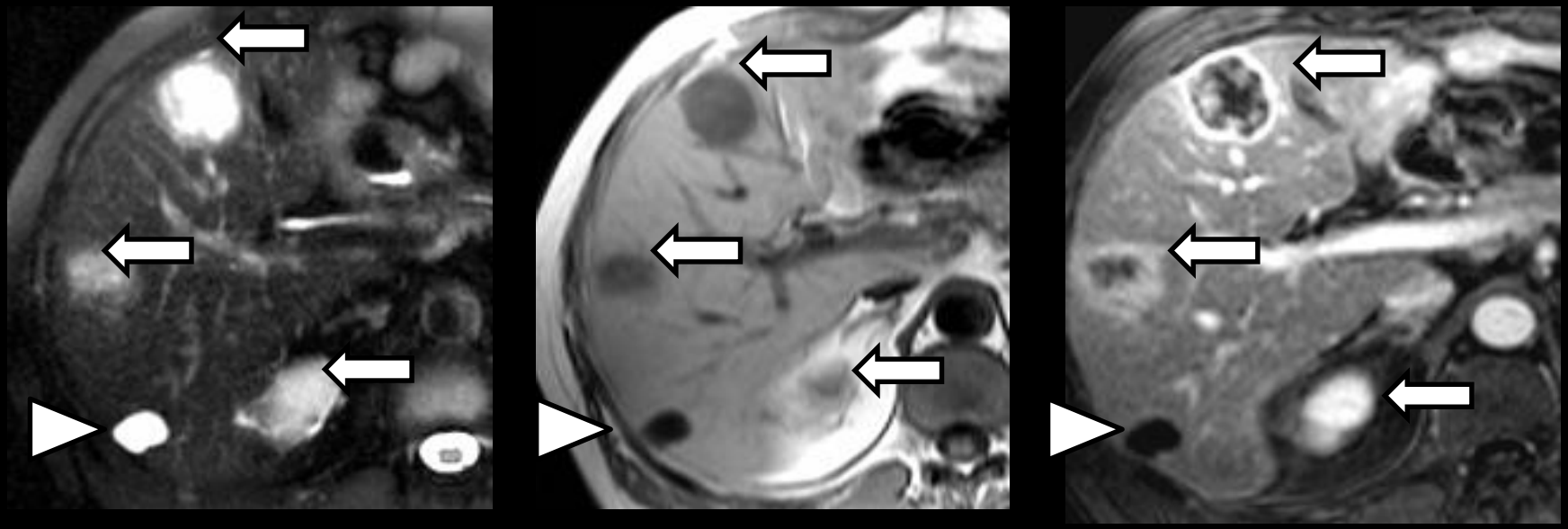

Fig 13: HASTE FS, GRE T1 and PV Ph MRI of a liver containing colorectal cystic metastasis (straight arrows) and a solitary simple cysts (arrow heads). Note the thick irregular enhancing walls of the metastases.

The second pathological process resulting in cystic liver metastases is the spread of a mucinous tumour. This is typically found in colonic (see fig 13) and ovarian adenocarcinoma. The latter can be differentiated from colonic malignancy by its peritoneal rather than haematogenous spread, which results in serosal and not intra parenchymal deposits. On all types of imaging, lesions can be single or multiple with complex features such as thick, irregular, enhancing walls; thick or nodular septations or internal debris.

3) Imaging Findings: Cystadenoma / Cystadenocarcinoma

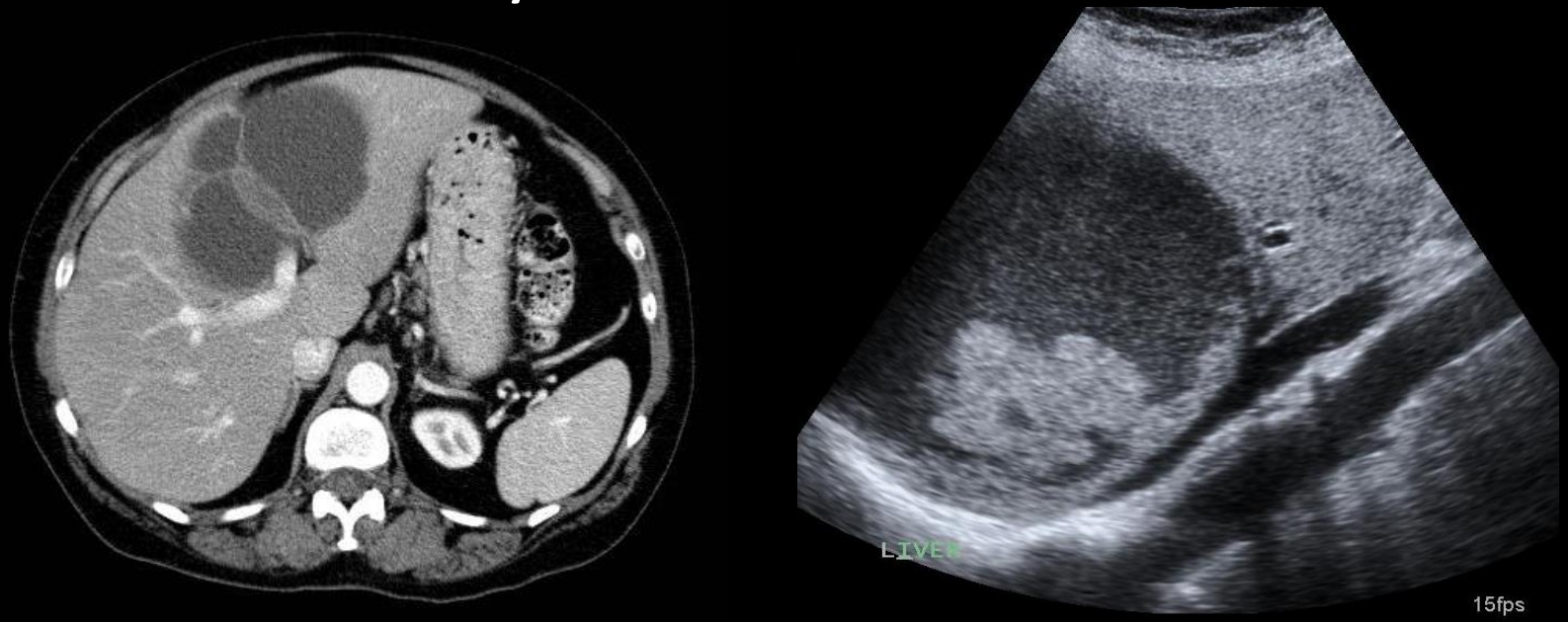

Fig 14: PV Ph CT scan and US scan of the liver demonstrating the internal septae and polypoid appearances of a cystadenoma.

A rare tumour predominately of middle aged women. They mainly arise from the intra-hepatic ducts with a predilection for the right lobe (55%). Symptoms are rare. Fluid within the lesion can be gelatinous, purulent or haemorrhagic. On CT imaging appearances are of a solitary lesion with a thick fibrous capsule, mural nodules and internal septae. A polypoid or pedunculated appearance are more commonly associated with cystadenocarcinoma. MRI features are similar, although signal characteristics vary according to the content of the lesion.

3) Imaging Findings: Undifferentiated Embryonal Sarcoma (UES)

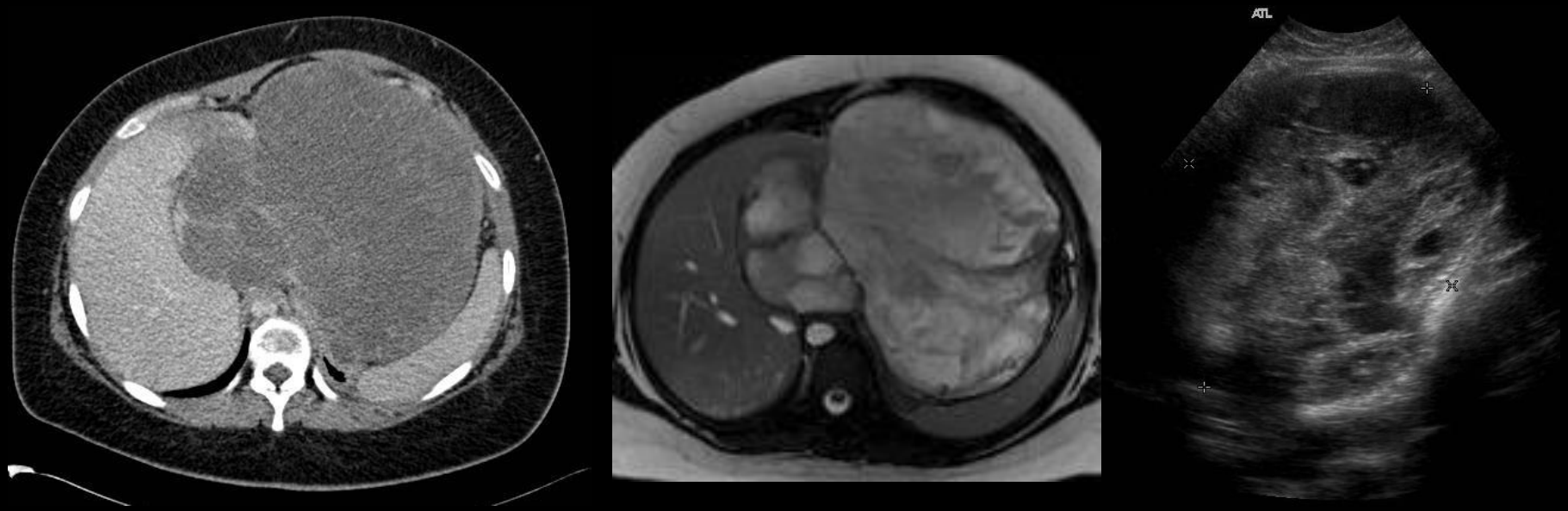

Fig 15: PV Ph CT scan, FISP balanced FFE MRI and US scan of an UES. Note how the lesion appears cystic on CT and MRI, but solid on US imaging. A key characteristic of UES.

UES is a rare tumour that mainly affects adolescent and young adults. Patients present with pain, fever and weight loss. On imaging it is typically a solitary lesion with well defined borders. Although it looks predominately solid at gross pathology, CT and MRI images usually demonstrate a cystic appearance due to high water content of the myxoid stroma. US however can show the solid nature of the tumour. This discordance between modalities can often help with the diagnosis. On contrast-enhanced CT and MRI, heterogeneous enhancement occurs of the peripheral solid components.

3) Imaging Findings: Abscess

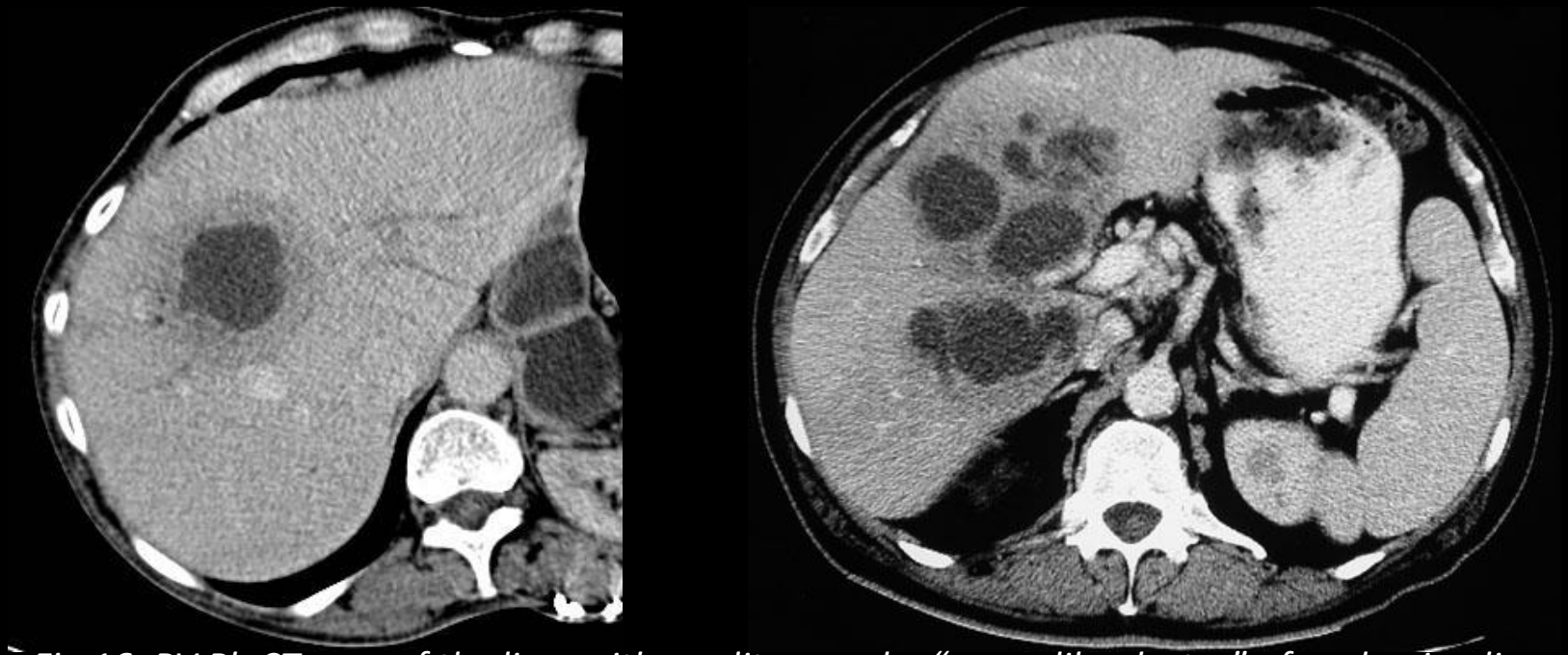

Fig 16: PV Ph CT scan of the liver with a solitary and a "grape like cluster" of coalescing liver abscesses.

Abscesses can be classified as pyogenic, amoebic or fungal. Pyogenic abscesses are thick walled with homogenous low attenuation on CT imaging and homogenous low T1 and high T2 signal on MRI imaging. With IV contrast, there is peripheral rim enhancement due to increased capillary permeability leading to a "double target sign". Peri lesional oedema can also be seen in 50% of abscesses. Amoebic and fungal abscess have similar characteristics to pyogenic abscess requiring correlation with clinical history and serology.

3) Imaging Findings: Hydatid Cyst

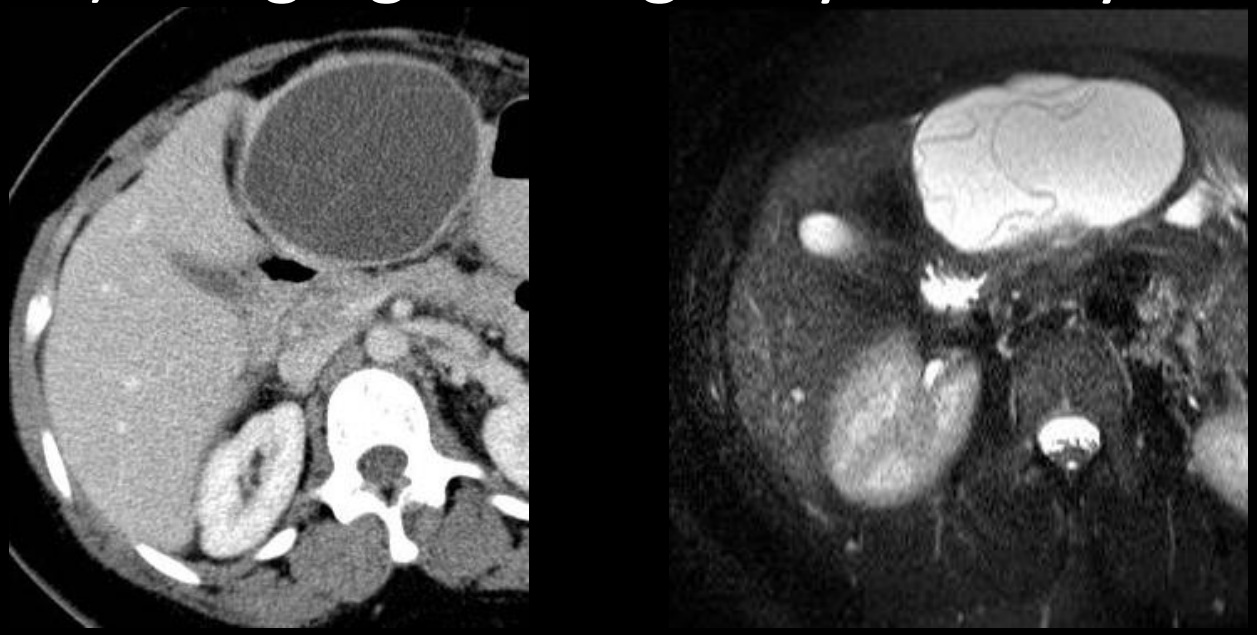

Fig 17: PV Ph CT scan and HASTE FS MRI of the liver containing a hydatid cyst in the left lobe. MRI is better able to demonstrate the internal architecture, in this particular case, a detached endocyst giving a "water lily sign". This signifies a transitional stage of the cysts.

Hepatic echinococcosis is caused by the ingestion of tape worm eggs that invade the liver via the portal system to form hepatic cysts. Patients present with abdominal pain and jaundice. Cysts are comprised of three layers. The pericyst, ectocyst and endocyst with daughter cysts forming from the endocyst. Pertinent clinical and laboratory features include the presence of an eosinophilia, positive serology and recent travel to an endemic area such a South America and Central Asia.

3) Imaging Findings: Hydatid Cyst

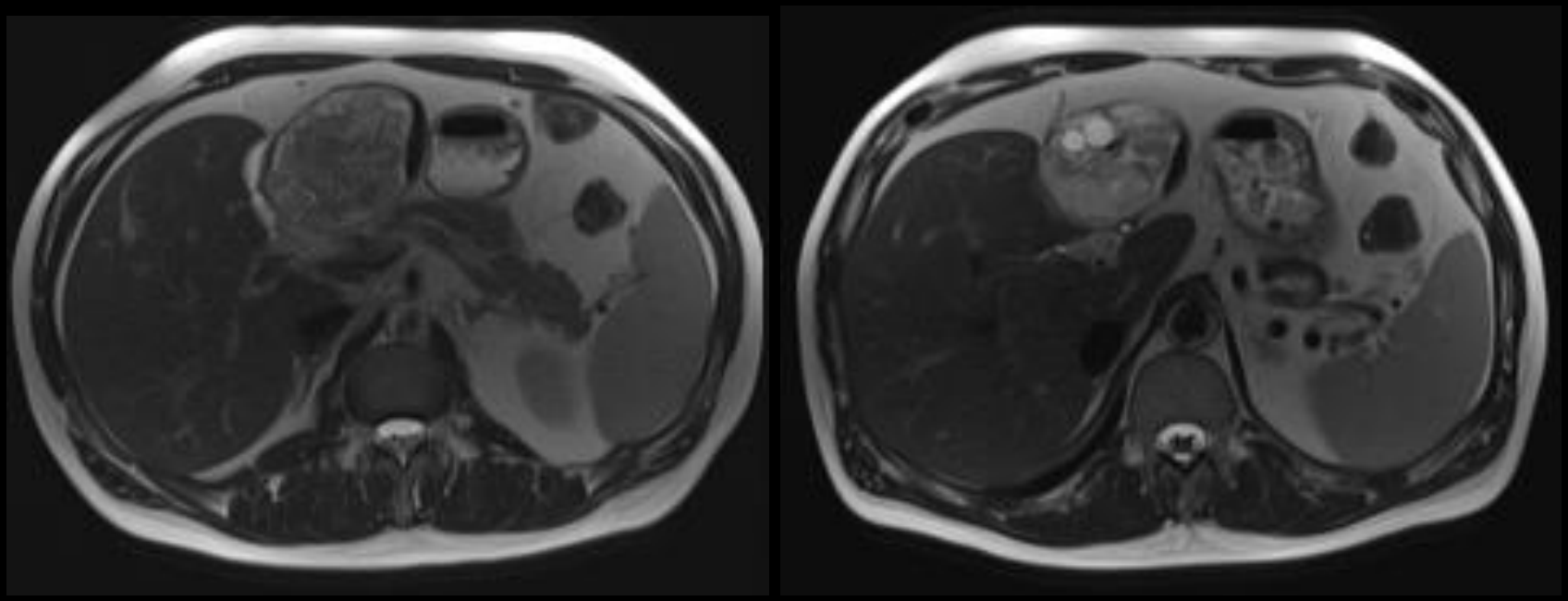

Fig 18: HASTE MRI of the liver containing a hydatid cyst in the left lobe. Unlike the previous example it contains numerous septations and daughter cysts suggesting a more active stage of the cyst.

On CT cysts are hypoattenuating and can be associated with peripheral calcification and daughter cysts. The hydatid matrix on MRI is hypointense on T1 and hyperintense on T2. Daughter cysts have a lower T2 signal relative to the main cyst. Complication of hydatid cysts includes bile duct compression and rupture of the cyst into the biliary system causing cholangitis. Patients can be treated conservatively with albendazole or undergo surgical resection / drainage.

3) Imaging Findings: Haematoma

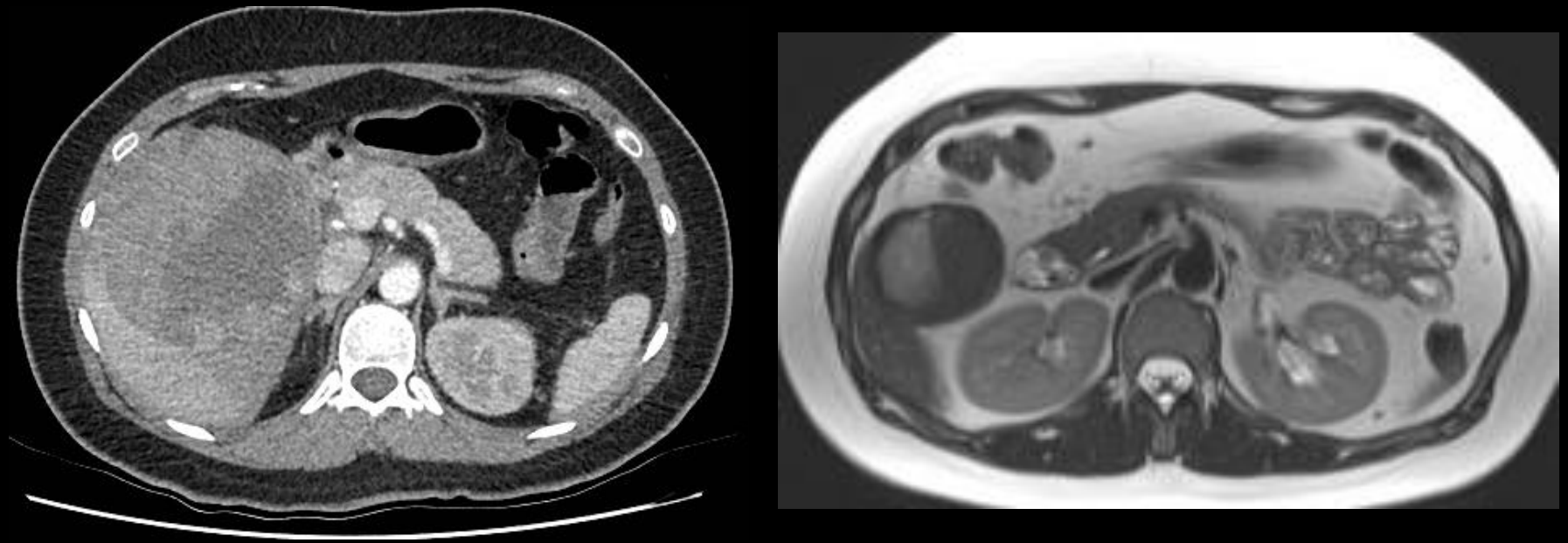

Fig 19: PV Ph CT scan and follow-up HASTE MRI of a liver hematoma within the right lobe caused by an adenoma. The haematoma has started to mature on CT and could be mistaken as a complex cyst.

Intrahepatic haematoma is associated with trauma and surgery, but can also be attributed to neoplasms such as adenomas. Patients present with symptoms of blood loss and peritoneal irritation. Appearances can be variable on CT depending on the age of the blood (see fig 19). In an acute or sub acute setting haematoma has a higher attenuation to water. Whereas in chronic cases hematoma has an attenuation close to water. The MRI signal depends on the amount of methaemoglobin. In the subacute cases it typically has high T1 and isointense T2 signal.

3) Imaging Findings: Biloma

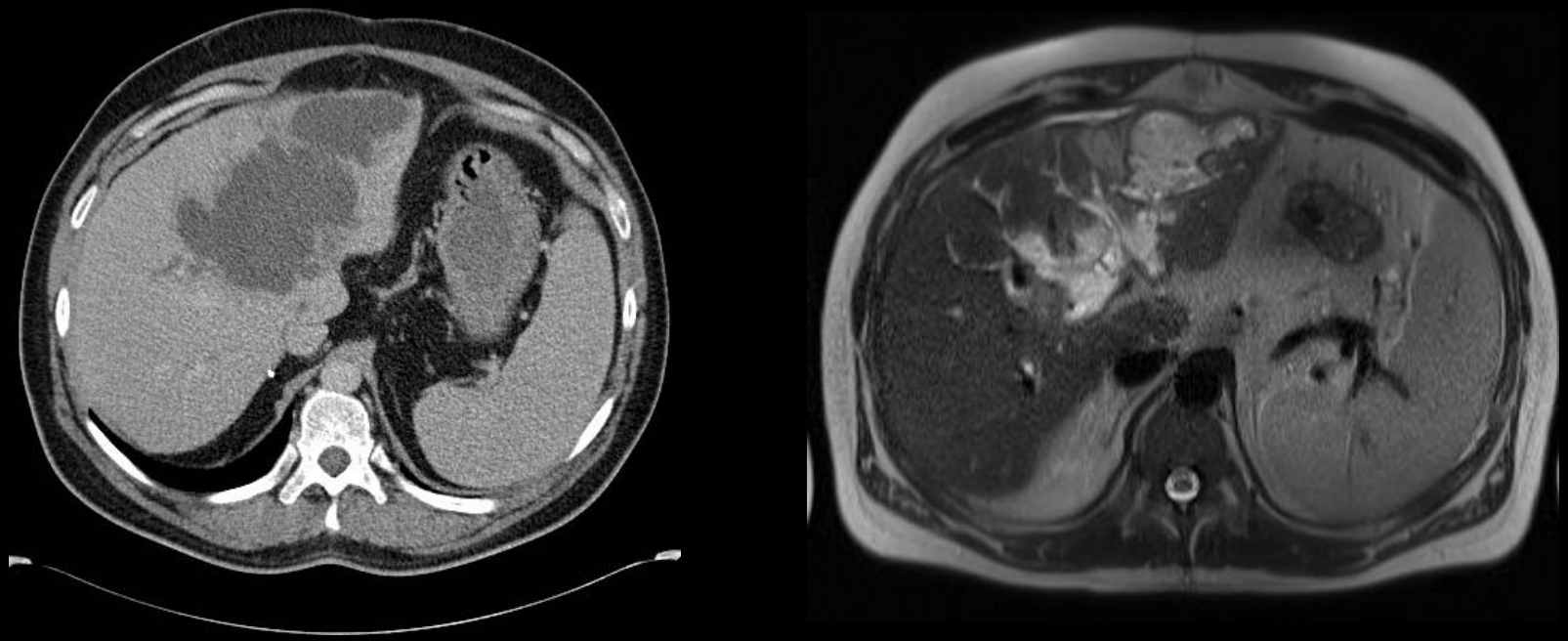

Fig 20: PV Ph CT scan and HASTE MRI of a large biloma of the left lobe secondary to ischemia after an orthotopic liver transplantation. Note the irregular contours and lack of a true capsule.

Biloma form from the disruption of the biliary tree. It can be spontaneous, secondary to trauma or hepatobiliary surgery. The latter causes stenosis/thrombosis of the hepatic artery, which forms the main blood supply to the biliary tree. Leakage within the liver parenchyma induces an inflammatory response that can form a pseudo-capsule. On CT and MRI imaging, a biloma is well defined or slightly irregular cystic area without septations or calcifications, or a true capsule. The collection can also accumulate hepatocellular contrast agent on delayed imaging due to active leakage of bile.

3) Imaging Findings: Peri Biliary Cysts

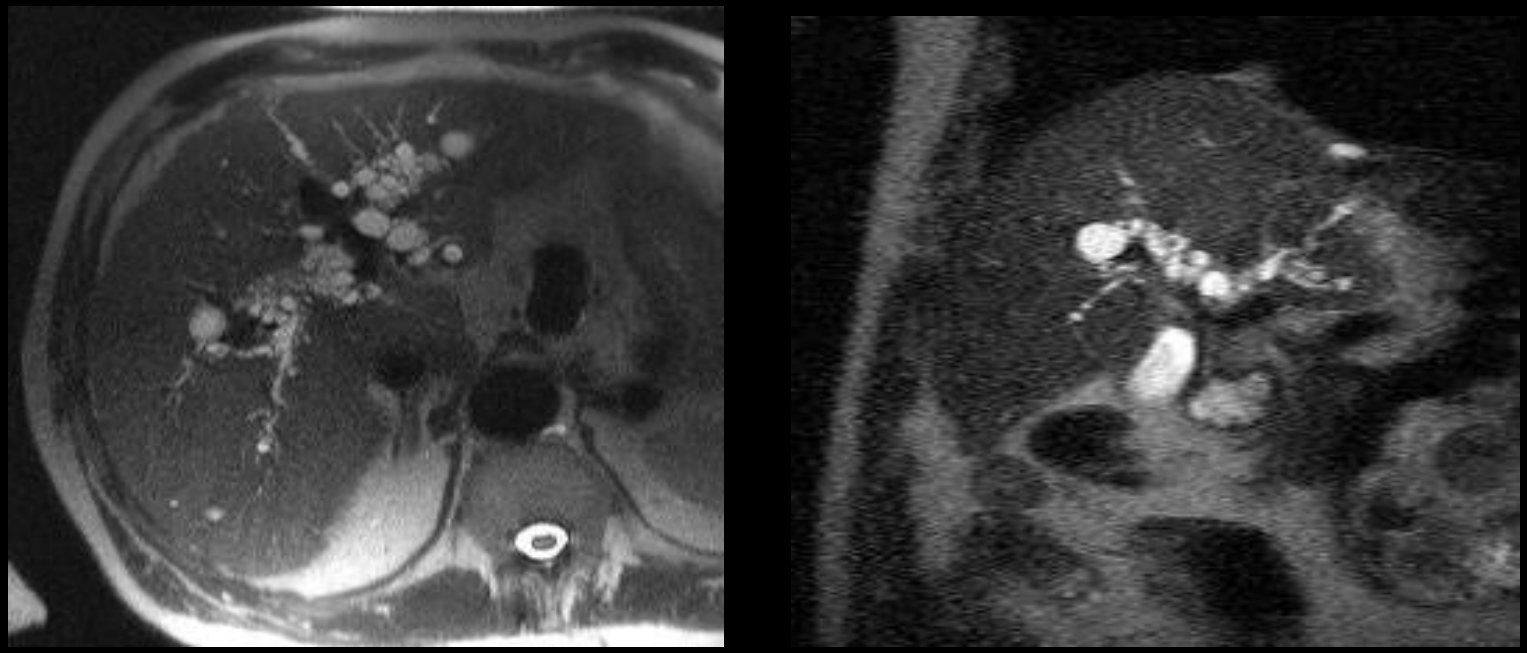

Fig 21: HASTE MRI of the liver containing multiple peri biliary cysts. Their distribution along the portal vessels gives them a "string of beads" appearance.

Peri biliary cysts are found in patients with chronic liver disease, especially in alcohol related disease. They arise from dilated peri biliary glands located along portal veins. The cysts do not communicate with the biliary tree and favour the left lobe. They have variable size and morphology and create a "string of beads" appearance due to their perivascular distribution. Peri biliary cysts have a similar appearance to simple cysts with low attenuation on CT, low signal intensity on T1 and high signal intensity on T2 MRI sequences with no enhancement.

4) Conclusion

- Diagnosis of liver cysts can sometimes be challenging
- Familiarisation with the presence or absence of complex features can be helpful with their diagnosis
- It is hoped this will empower radiologists to make more accurate diagnoses, thus reducing the need for liver biopsy and patient anxiety

5) References

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