

# Hiding in Plain Hepatocyte: A Rare Case of Liver Disease

**Dr. Eleanor Baker, ST2 Radiology Trainee**

**Dr. Anisha Bhagwanani, Consultant GI Radiologist**

**Dr. Emily Tam, Consultant GI Radiologist**

**Wexham Park Hospital**

**Frimley Health NHS Foundation Trust**

# Clinical History

16-year-old female presenting with a 1-day history of right upper quadrant pain, anorexia and vomiting after eating raw seafood.

No significant past medical or surgical history. No regular medications. No oral contraceptive pill usage.

On clinical examination, hepatomegaly was noted, but the abdomen was otherwise soft and non-tender. Cardiorespiratory examinations and observations were normal.

# Initial Investigations

Test	On Admission	Day 1	Test	Result
Hb	107 (↓)	94 (↓)	APTT, PT	33.6 (-), 15.2 (-)
WCC	7.7 (-)	9.3 (-)	Ferritin	170 (-)
CRP	<0.6 (-)	5.7 (-)	Non-invasive liver screen auto-antibodies	All negative
ALT	93 (↑)	2,236 (↑↑)	Hep A IgG Ab	Detected
AST	200 (↑)		Hep B Surface Ag	Negative
Bilirubin	7 (-)	9 (-)	Hep C Ab	Negative
ALP	69 (-)	161 (-)	EBV and CMV	Negative
D-Dimer	6860 (↑↑)			
Creatinine	56 (-)			
K <sup>+</sup>	2.9 (↓)	4.4 (-)		

# Clinical Differential Diagnosis

## Budd-Chiari Syndrome

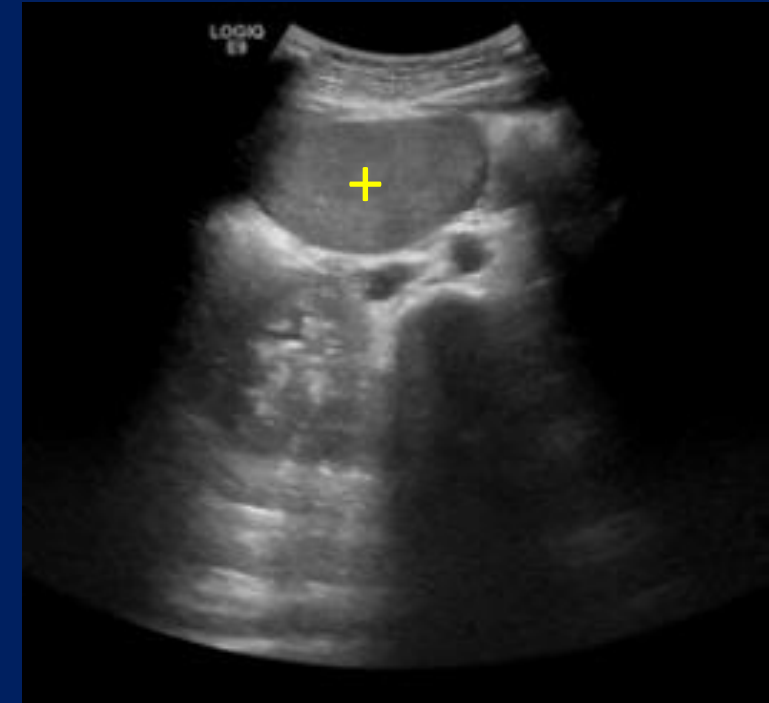
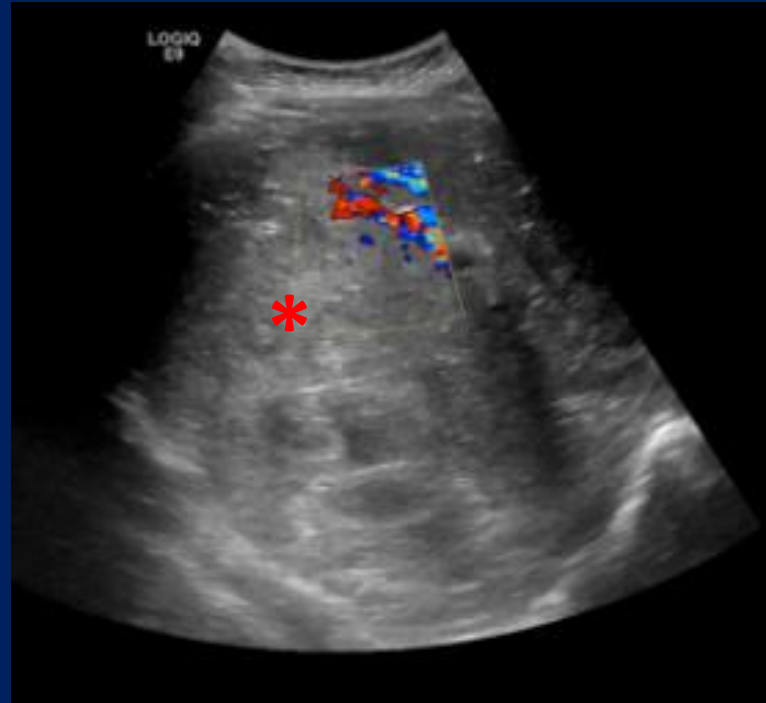
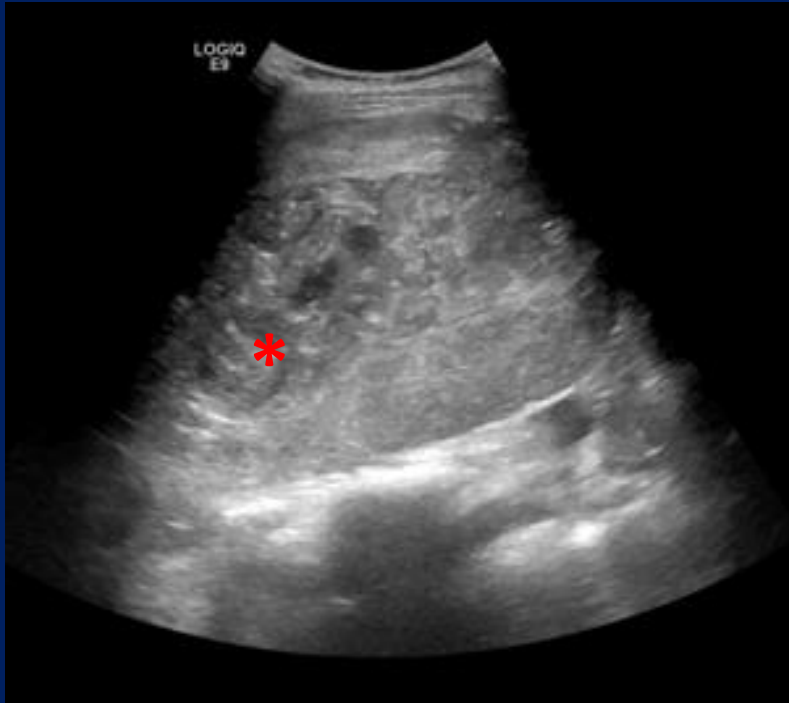
(RUQ pain, hepatomegaly and high d-dimer)

vs.

## Acute Viral Hepatitis

(RUQ pain, hepatomegaly, transaminitis, history of raw seafood ingestion, positive Hep A antibodies)

# Liver Ultrasound



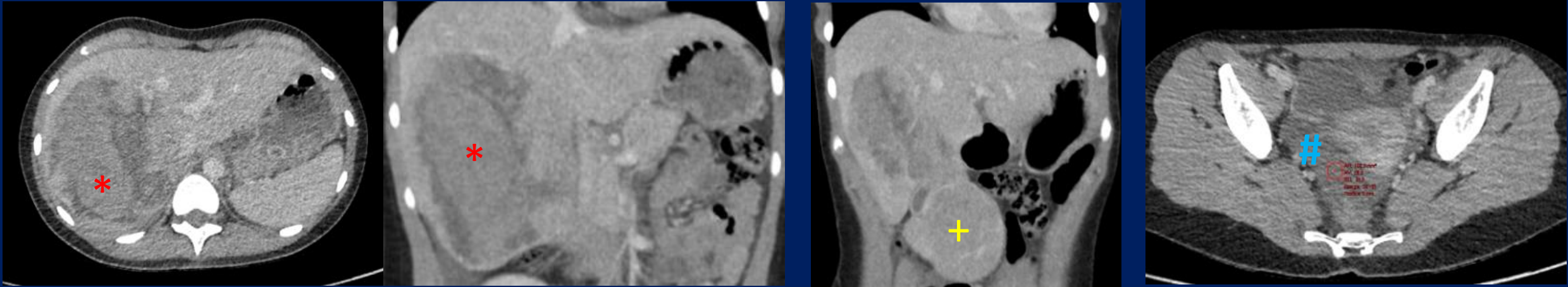
Heterogenous mass occupying the right lobe of the liver (\*)

Second, well-defined exophytic mass extending inferiorly from the left lobe of the liver (+)

Patent portal and hepatic veins

CT abdomen and pelvis recommended for further characterisation

# CT Abdomen and Pelvis



Axial and coronal images of contrast enhanced CTAP

- Large heterogenous right hepatic lesion
- Hyperdense components suspicious for intralesional haemorrhage (\*)
- Second, well-defined exophytic lesion arising from the inferior margin of segment III (+)
- Moderate volume intraperitoneal fluid with a mean density of approximately 30 HU (#) suggesting haemorrhagic ascites

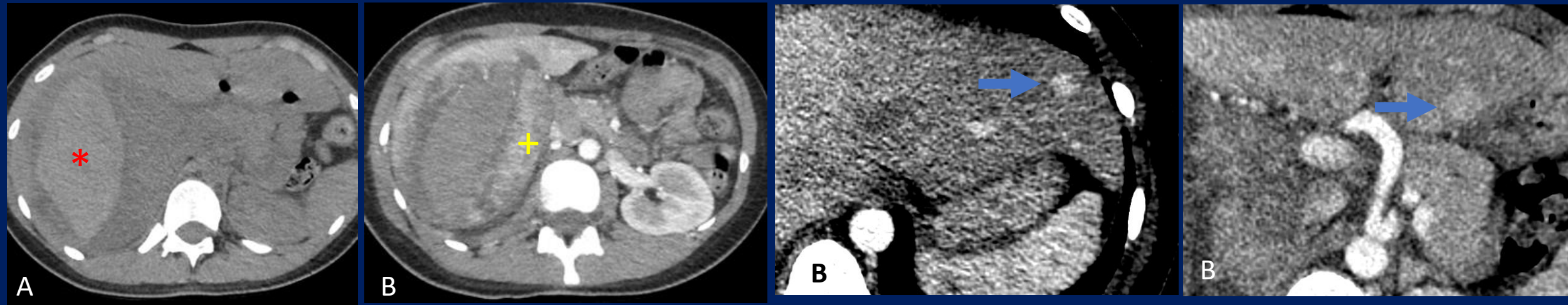
# Initial Management

The patient was clinically stable and therefore no emergency IR or surgical intervention was undertaken.

She was treated with 2 units of packed red cells, analgesia, anti-emetics and fluids.

Following discussion with a tertiary HPB referral centre, CT triple phase liver was suggested.

# CT Triple Phase Liver



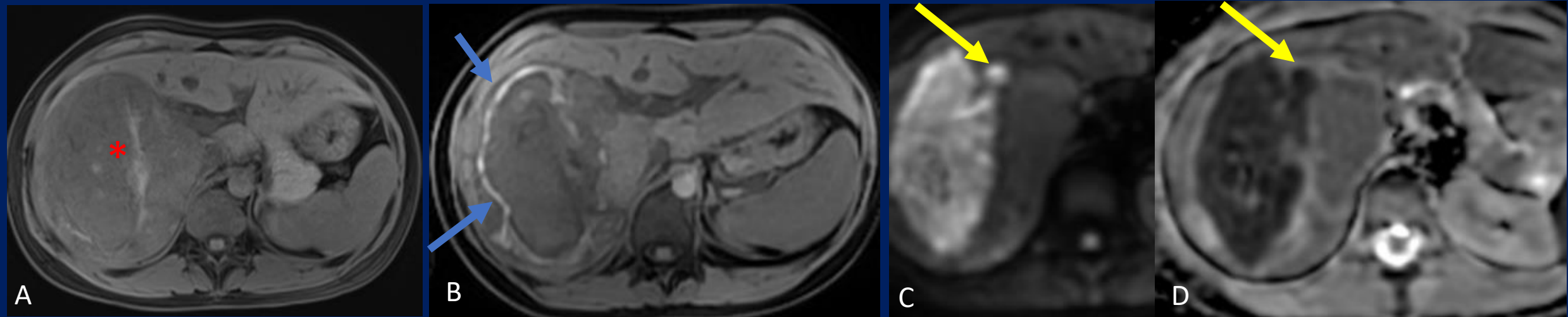
Non-contrast (A) and arterial phase contrast-enhanced (B) images

- Unchanged appearances of large right hepatic lesion with intralesional haemorrhage (\*) but no active contrast extravasation
- The lesion demonstrated peripheral nodular arterial enhancement (+)
- Multiple further small, arterially enhancing lesions identified within the background hepatic parenchyma (→)

MRI liver recommended for further characterisation



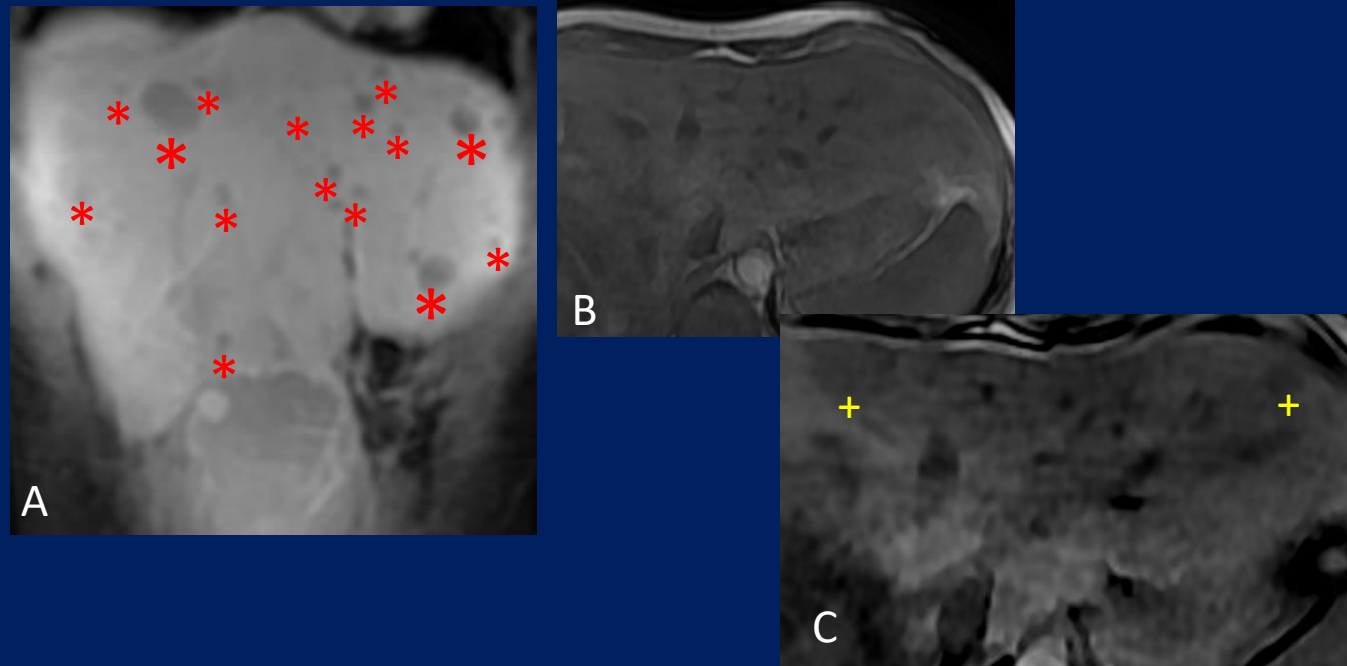
# MRI Liver



Axial T1 (A), T1 + arterial phase contrast (B), DWI (C) and ADC (D) images

- Solid, heterogenous right lobar mass with central non-uniform T1 signal indicating haemorrhage of variable age (\*)
- Peripheral arterial contrast enhancement ( → ) but no central filling, washout or uptake of hepatobiliary contrast
- Heterogenous diffusion restriction demonstrated on DWI and ADC imaging ( → )

# MRI Liver



Coronal T1 + contrast (A), axial in-phase (B) and out-of-phase (C) images

- More than 30 background liver lesions (\*) some of which demonstrate loss of signal on out-of-phase imaging (+) consistent with microscopic fat

## Radiological Differential Diagnosis:

Multiple Hepatic Adenomas

VS.

Multifocal Hepatocellular Carcinoma

VS.

Hepatic Metastases

# Further Management

The patient was transferred to a tertiary centre under the care of the HPB surgical team.

Ultrasound guided biopsy of the large right liver lesion was performed.

Histology demonstrated a well-differentiated hepatocellular lesion with large areas of intralesional necrosis. Features most in keeping with hepatic adenoma (subtype not apparent) but necrosis precluded further assessment.

# Further Management

MDT discussion resulted in recommendation for right hepatectomy.

Histology from right hepatectomy revealed HFN1 $\alpha$ -inactivated hepatocellular adenoma with focal FNH-like change on a background of multiple small hepatocellular adenomas revealing the final diagnosis of...

## **Hepatic Adenomatosis**

Genetic testing isolated a heterozygous pathogenic variant of the HFN1A gene, which is associated with an increased risk of maturity onset diabetes of the young, NAFLD and HCC.

Lifelong surveillance required.

# Hepatic Adenomatosis

The presence of numerous (>10) hepatic adenomas. A rare condition which is not usually associated with the classic risk factors of oral contraceptive or steroid use.

Mostly asymptomatic but may present with abdominal pain due to intraperitoneal haemorrhage.

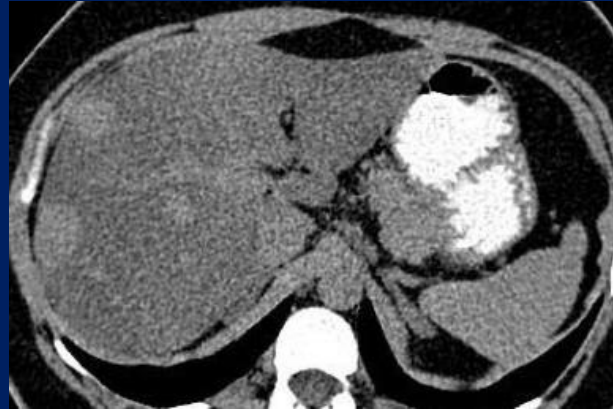
Adenomas are at risk of haemorrhage (60%) and malignant transformation to HCC (<10%).

Symptomatic adenomas may be surgically resected or ablated. Liver transplant can be considered for patients with widespread symptomatic disease. Surveillance for malignant transformation is required.

# Hepatic Adenomatosis

## CT:

- Non-contrast -> hepatic lesions with variable attenuation. Hyper- with haemorrhage, hypo- with intralesional fat
- Arterial and portal venous -> variable enhancement, arterial phases are usually the most sensitive



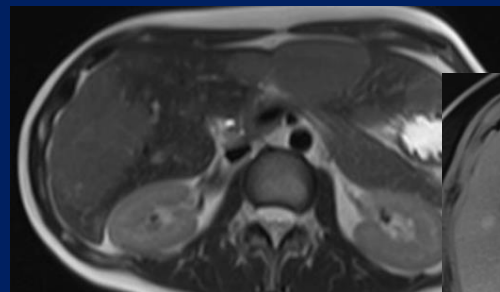
Non-contrast



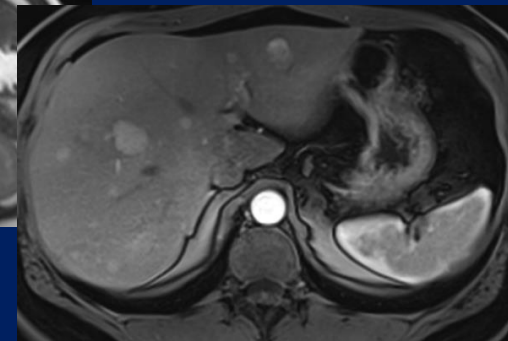
Arterial

## MRI:

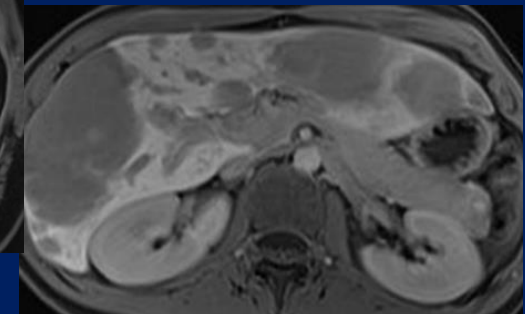
- T1 -> iso or hypointense to liver
- T2 -> hyperintense to liver
- Gadolinium -> arterial enhancement and variable portal venous/ delayed enhancement
- Hepatocellular contrast -> little to no enhancement



T2



Arterial



Hepatocellular contrast

# References

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