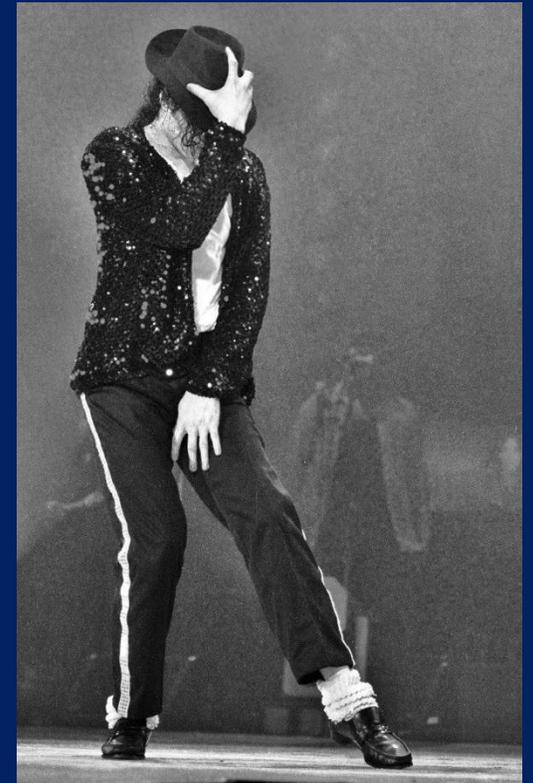


A case of solitary hepatic epithelioid haemangioendothelioma (HEHE)

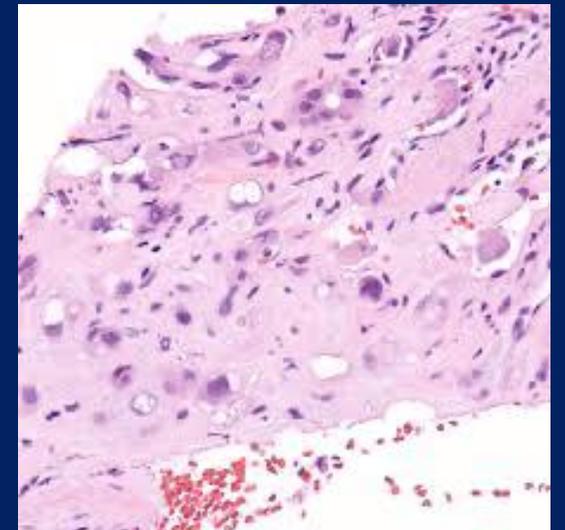
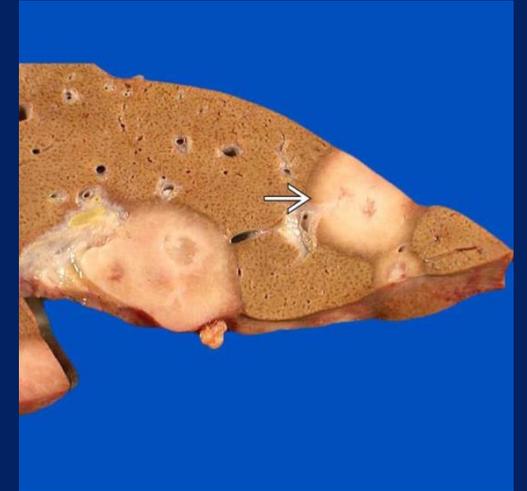
Dr James Mawdsley, Radiology ST3 – UHBW, Severn Deanery

Dr Hedvig Karteszi, Consultant Radiologist – UHBW, Severn Deanery



Pathology/aetiology

- Extremely rare, slowly progressing, low-grade malignant vascular tumour
 - Lungs, lymph nodes, peritoneum, bone, spleen, and diaphragm are the most common sites of extrahepatic involvement
- Possible association with *Bartonella* infection [1]
- Metastatic in 40% of cases - lung, bone, spleen



Presentation

- Median age of occurrence – 50 y/o (range - 7 – 86)
- Female:male – 3:2^[1]
- Up to 40% asymptomatic at diagnosis
 - Pain, cough, lethargy, weight loss

Classic imaging features

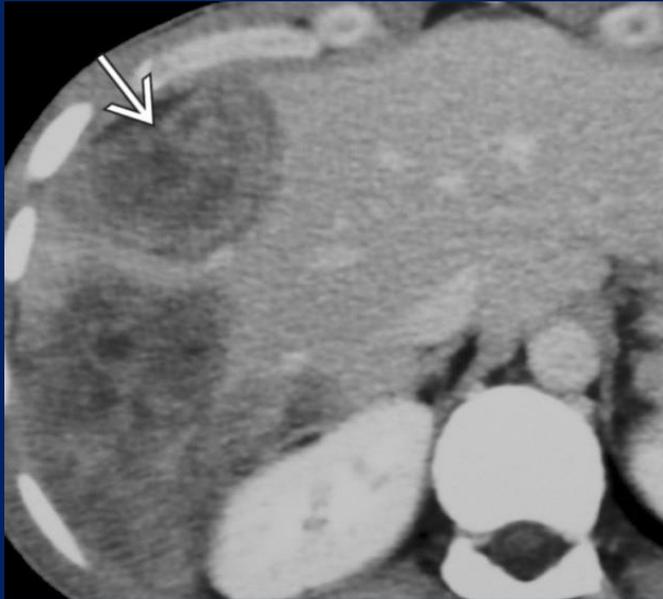


Fig.1

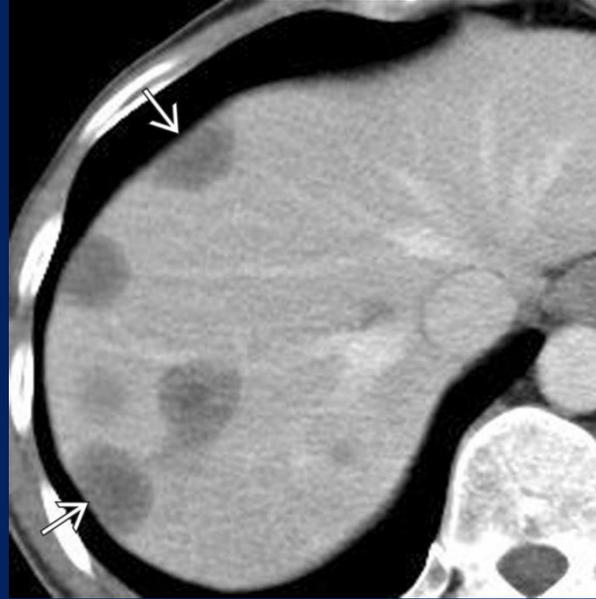


Fig.2

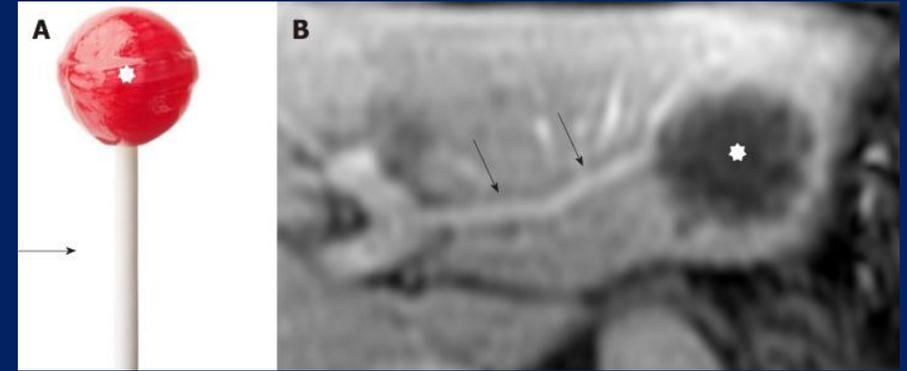


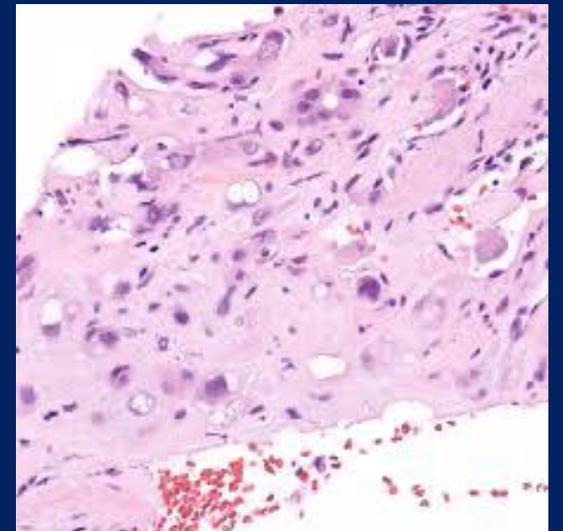
Fig.3

- Peripheral hepatic nodules, often coalescent (fig,1)
- Capsular retraction (fig 2)
- Lollipop sign – a hepatic or PV terminating at or just within the periphery of some of the lesions (fig.3)
- Target-like enhancement pattern (fig.1)
- Mild/moderate peripheral diffusion restriction

Solitary lesions are rare!

Histopathological diagnosis

- Positive stain for CD34, factor VIII, and CD31; express Fli-1 protein
- CAMTA1 expression is useful for distinguishing EHE from other vascular tumours ^[3]



Differential diagnosis

- Cholangiocarcinoma
 - Heterogeneous mass with capsular retraction
 - Intrahepatic bile duct dilation
 - Satellite lesions possible

- Focal confluent fibrosis
 - Common in advanced cirrhosis

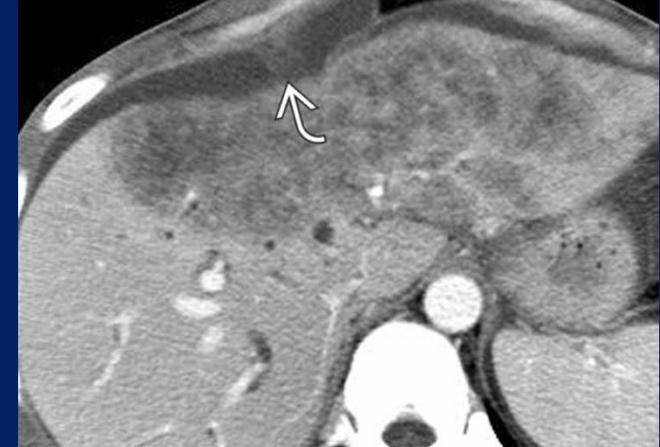


Fig 1. cholangiocarcinoma

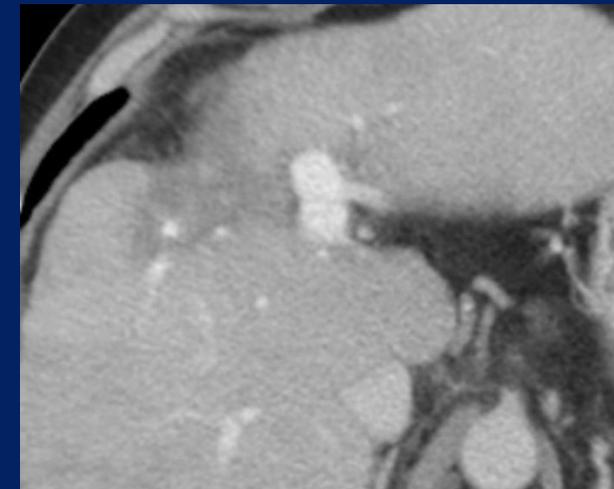


Fig 2. focal confluent fibrosis

Management

- Surgical resection remains the preferred modality for patients presenting with localized disease confined to a single organ ^[2]
- Liver transplantation can be considered in unresectable disease
- Chemotherapy
- Targeted beam radiotherapy
- Immunotherapy - ongoing trials of trametinib

Prognosis

- 60% presenting with metastatic disease at diagnosis
- 5-year survival between 59 – 86%

Solitary HEHE - case synopsis

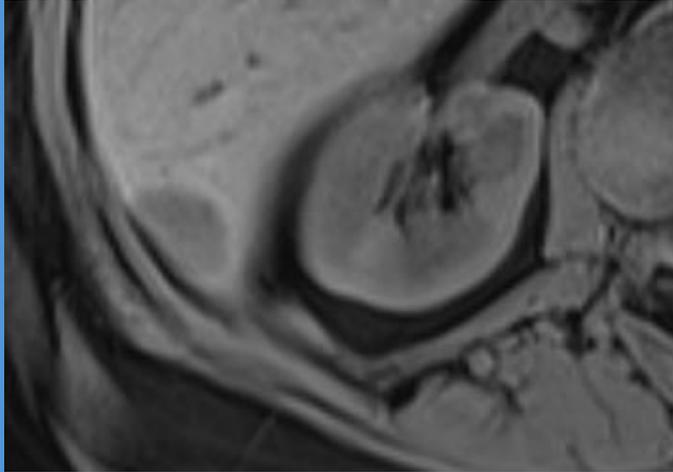
Background

- 79F – white British.
- No significant past medical or surgical history.
- Performance status 2, Frailty score 3.

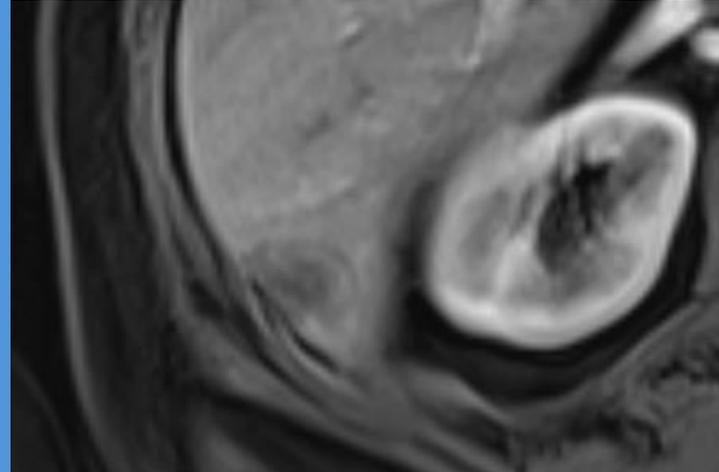
Case history

- *November 24* – incidental liver lesion on CT colonography.
- *January 25* – solitary 3cm lesion in segment 6. Slight capsular retraction.
 - Differential diagnosis - intrahepatic cholangiocarcinoma vs metastasis.
- *March 25* – PET-CT to assess for distant primary.
 - Mild uptake in liver lesion.
 - Focal uptake within neck/oesophagus – OGD + MRI neck – NAD.

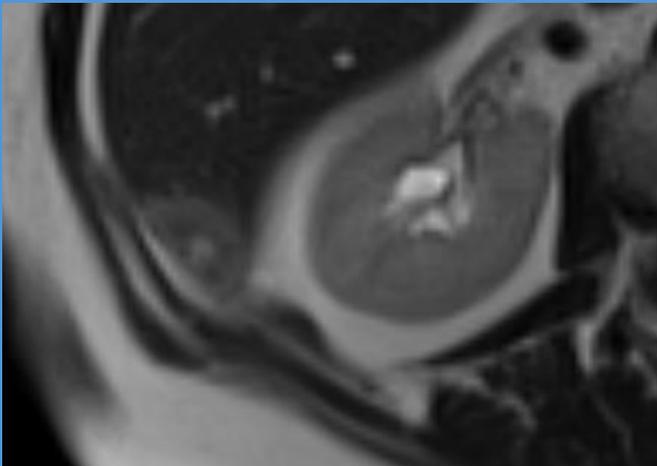
Solitary HEHE - case synopsis



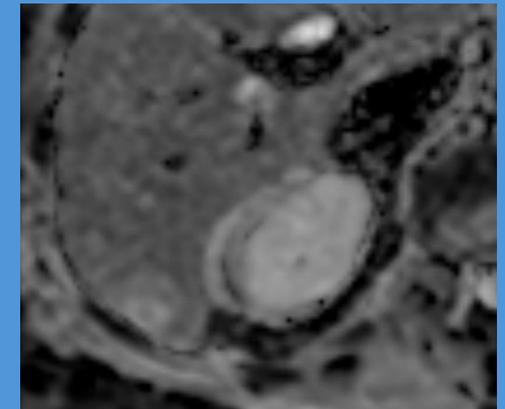
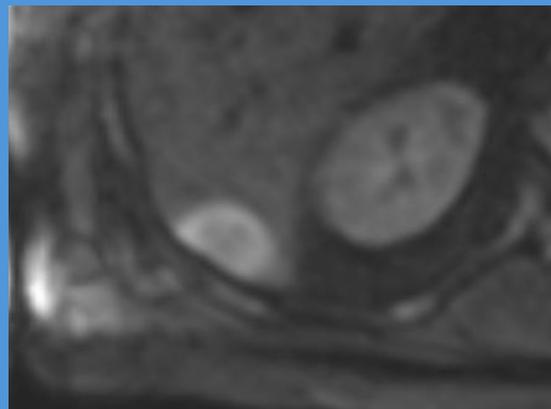
T1: hypointense lesion with slight capsular retraction



T1 C+ (Gd): target-type enhancement with central low intensity



T2: moderate T2 high signal with bright central core



DWI: demonstrating mild/moderate peripheral diffusion restriction

Solitary HEHE - case synopsis

- *Surgical management*
 - Wedge excision. Good post-operative recovery.
- *Histopathology*
 - Epithelioid haemangioendothelioma.
 - +ve for vascular markers ERG and CD34, weakly +ve for CAMTA1.
- *Follow up*
 - Oncology did not recommend adjuvant treatment.
 - Follow up cross-sectional imaging in 6 months.

Case discussion

- Rare – incidence 1 in 1,000,000
- Solitary tumours uncommon, making up 13% of HEHE's ^[3]
- Diagnostically challenging as single, solitary tumours may appear non-specific and mimic other, more common pathology
- Decision to perform wedge resection made on imaging - no tissue diagnosis prior to surgery
- Diagnosis made on post-operative histopathological analysis

References

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3. Kubo N, Harimoto N, Araki K, Hagiwara K, Yamanaka T, Ishii N, Tsukagoshi M, Igarashi T, Watanabe A, Miyazaki M, Yokoo H, Kuwano H, Shirabe K. The Feature of Solitary Small Nodular Type of Hepatic Epithelioid Hemangioendothelioma. *Case Rep Gastroenterol*. 2018 Aug 21;12(2):402-410. doi: 10.1159/000490524. PMID: 30186092; PMCID: PMC6120404.
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