An uncommon presentation for an uncommon disease

Dr Alessandro Antonello, Dr Raimat Salami, Dr Saeideh Toupchiani, Miss Bethany Tucker,

Dr Tassos Grammatikopoulous, Dr Annamaria Deganello

King's College Hospital

Clinical Background:

- A 12-year-old male patient presented at his local hospital following multiple episodes of haematemesis and melaena. This was associated with abdominal pain and an episode of collapse. He began experiencing these symptoms after eating at a fast-food restaurant.
- No significant past medical or family history
- On examination, he was pale and lethargic:

A- Airway patent

B- RR 25- 30, no recession, chest clear

C- HR 144, BP systolic 70, cold peripheries to mid arm and mid leg, prolonged CRT,

D- Abdomen SNT, able to speak though weak, can give name, age, address, orientated, GCS 14 (opens eyes to command), No palpable lymphadenopathy

E- Cold to touch, no rash

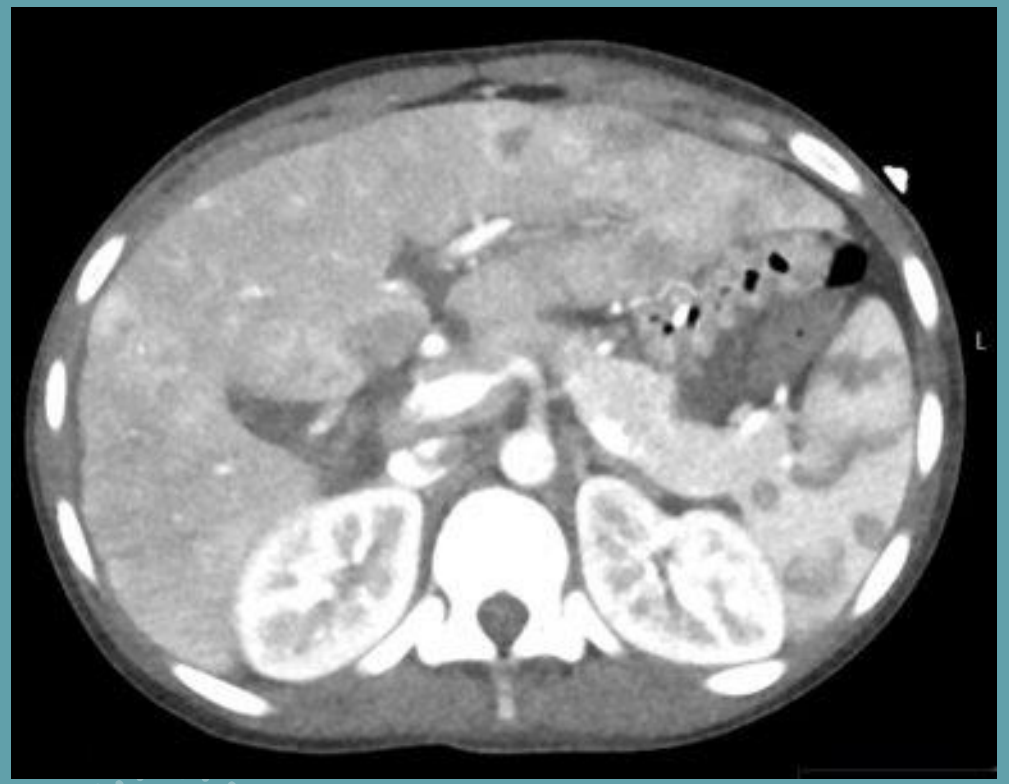
G- BM 12.8

- He was found to be anaemic with elevated inflammatory markers and deranged liver function tests. Autoantibody screen was normal. Blood gas was acidotic with a high lactate.

Next Steps:

- Inflammatory changes of the mesenteries and small amount of pelvic free fluid were found on initial ultrasound
- Normal initial OGD
- Normal initial colonoscopy
- Abnormal appearance of the liver on diagnostic laparoscopy with "white plaque like capsular lesions"

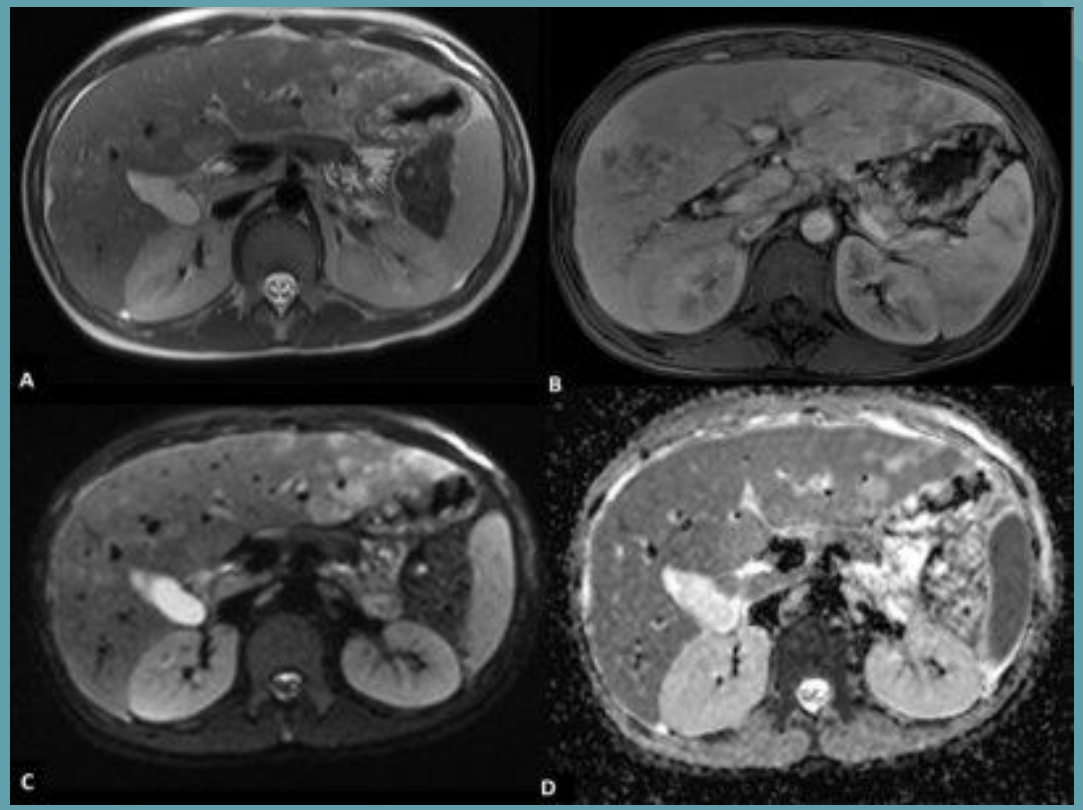
CT Abdomen and Pelvis:



CT appearances of the liver (late arterial phase scan). Multiple ill defined low attenuating lesions with peripheral arterialisation were demonstrated throughout the hepatic parenchyma.

MR Liver and Histology:

- MR appearances of the liver (A: T2 weighted images, B: postcontrast images – gadovist, arterial phase, C: DWI D: ADC map): Multiple ill defined high T2 signal lesions were demonstrated throughout the hepatic parenchyma. These demonstrated shine through on DWI/ADC but no restricted diffusion. There was also peripherally increased arterialisation.

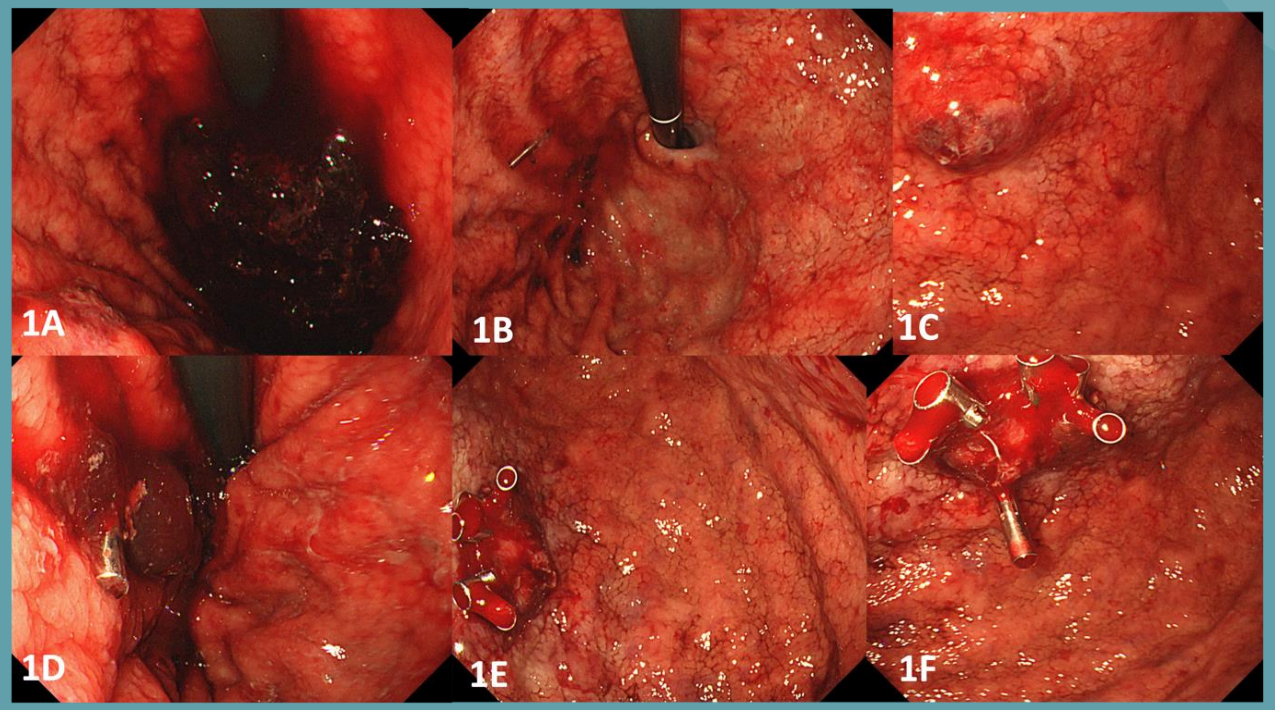


Transfer and management at King's

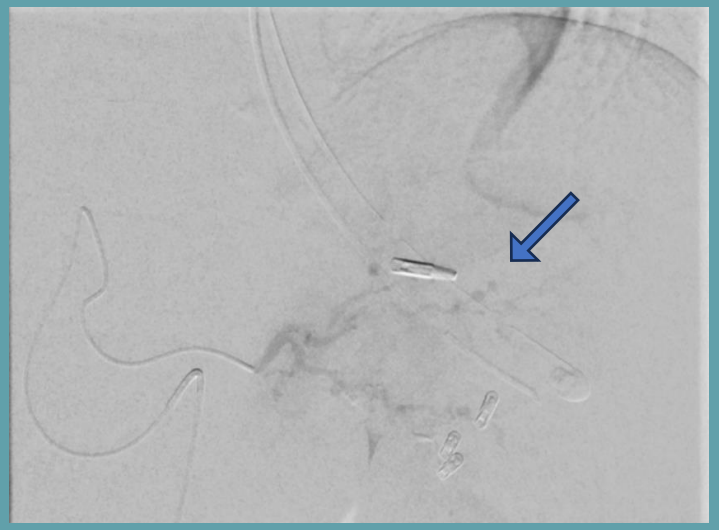
- Repeat CT: numerous liver nodules against a background of varied, nodular parenchyma, accompanied by small splenic and gastric varices and moderate ascites. Thoracic and abdominal lymphadenopathy.
- Liver histology from the lesions demonstrated non-specific inflammatory changes.
- Due to recurrent UGI bleed the patient underwent two additional interventional endoscopies involving endoclips, adrenaline injection, haemospray, Sengstaken-Blakemore tube placement, and eventually embolisation of the left gastric and branches of the left hepatic artery.
- Vasculopathic changes were observed during the embolisations, and the inflammatory markers remained raised throughout.

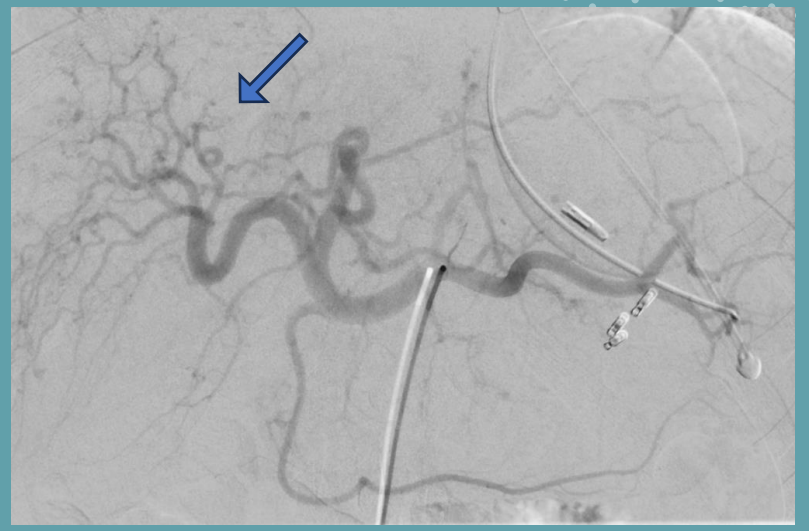
Endoscopic Findings

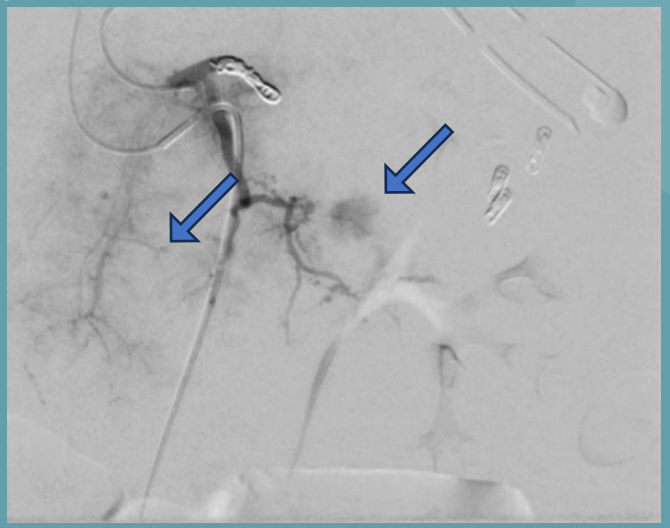
Figures 1A-E: Upper endoscopy revealed a thrombus, a bleeding ulcer and a haemangioma-like lesion which required endoclips



Angiographic Findings



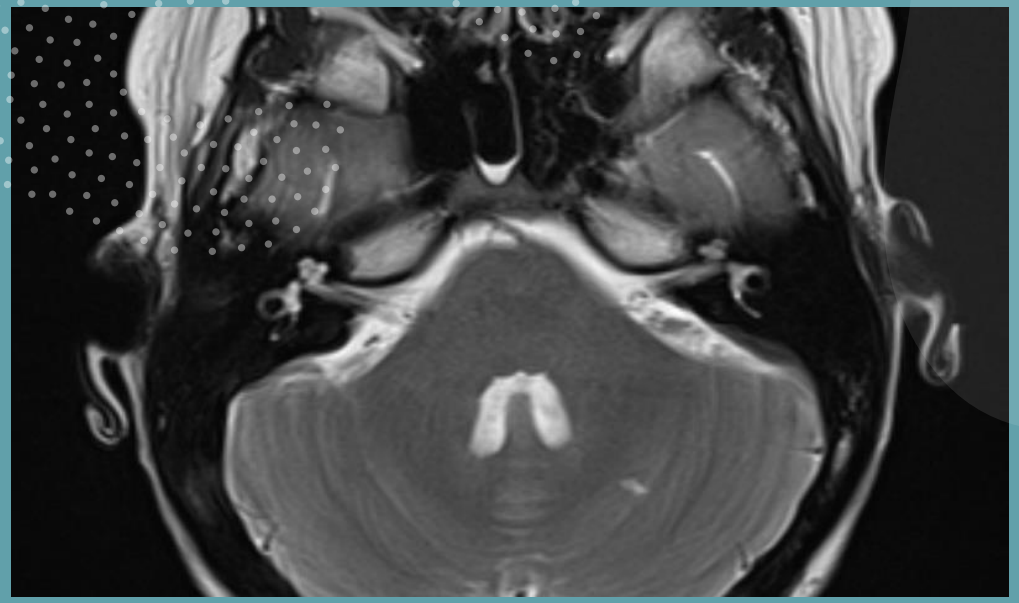


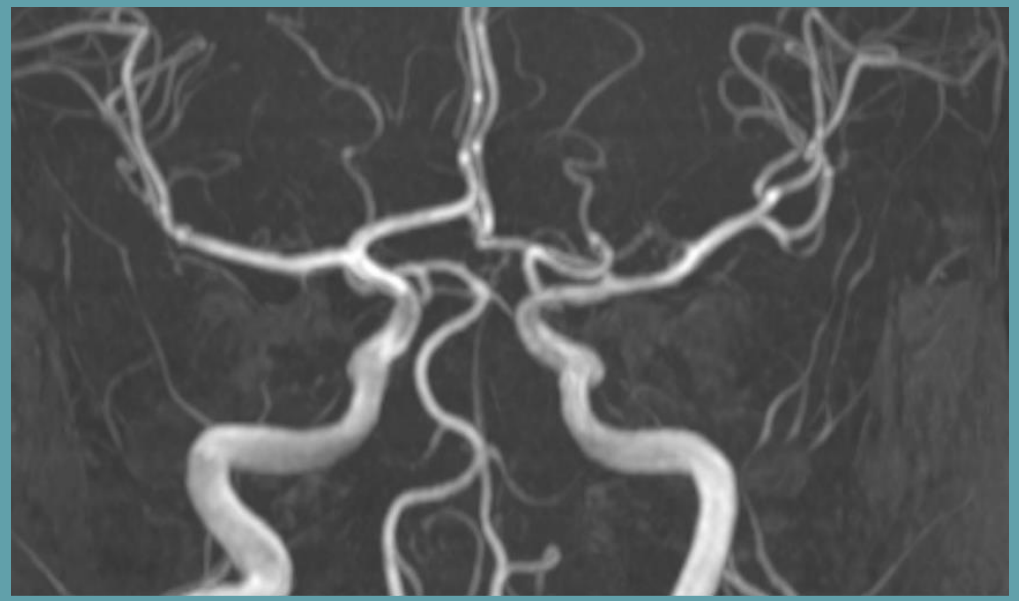


Conventional angiography demonstrated the presence of vessel abnormalities with extensive beading. Multiple foci of contrast extravasation were also demonstrated and treated with PVA particles and coils

Neuroradiology

Due to confusion and headaches an MRI brain and MRA were performed: the presence of small mature cerebellar infarct, a small anterior communicating artery aneurysm and a degree of beading of the anterior cerebral arteries were demonstrated.





Paediatric Vasculitis

- Based on the clinical and imaging findings a diagnosis of vasculitis was made
- Treatment with corticosteroids was initiated, leading to the termination of gastrointestinal bleeding episodes after 14 days. Subsequently, a biweekly intravenous cyclophosphamide regimen was administered during the prednisolone tapering period for three months, with no recurrence of bleeding episodes to date.
- In children, vasculitis is an uncommon contributor to gastrointestinal bleeding. This case highlights the significance of including vasculitis in the differential diagnosis, especially when confronted with persistent upper gastrointestinal bleeding that does not respond to conventional treatments.