Median arcuate ligament syndrome and implications in celiac axis embolization

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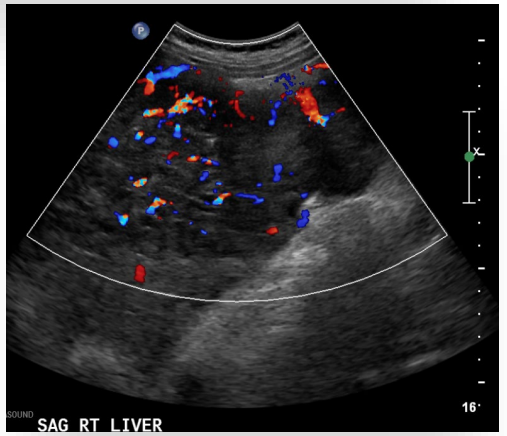
Case History

- 65 year old male
- Initially presented to general surgery with several month history of generalized malaise, weight loss, and decreased appetite.
- PMHx: hypertension
- PSHx: appendectomy
- All: None

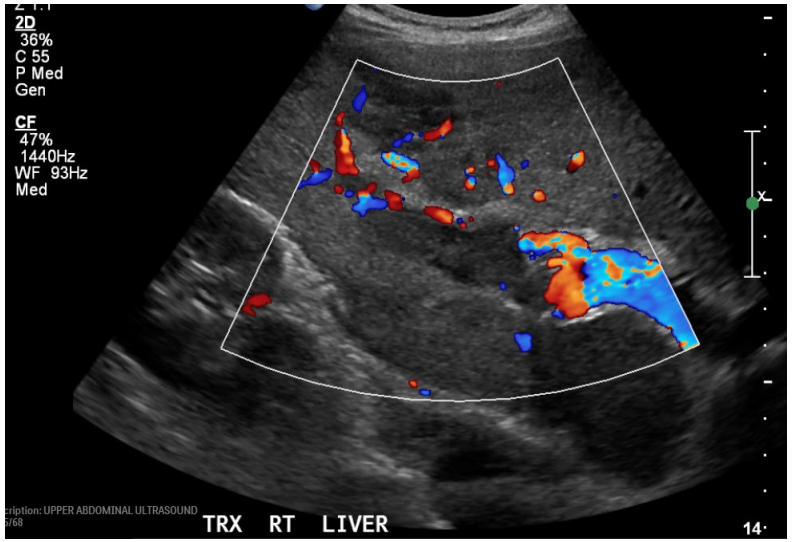
Ultrasound

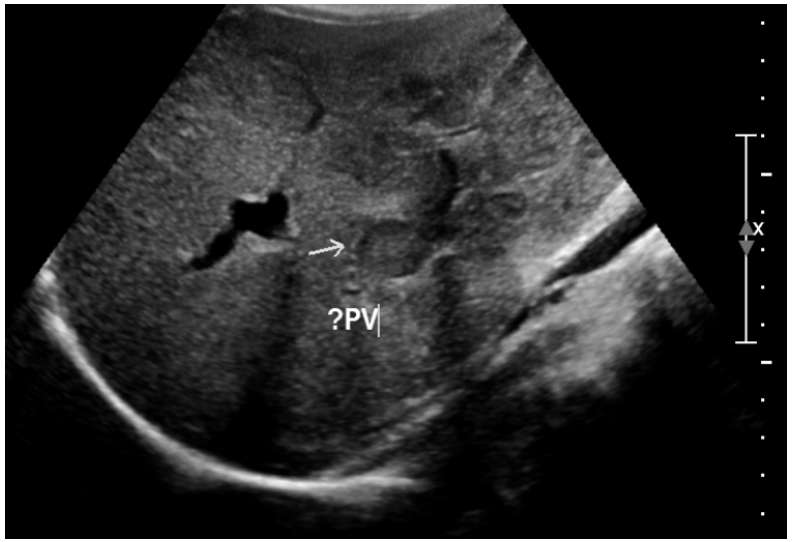
- Large vascular mass in the right lobe of the liver measuring 18 x 8 x 10 cm.
- Hepatosplenomegaly.
- No evidence of cirrhosis.
- Mild ascites.

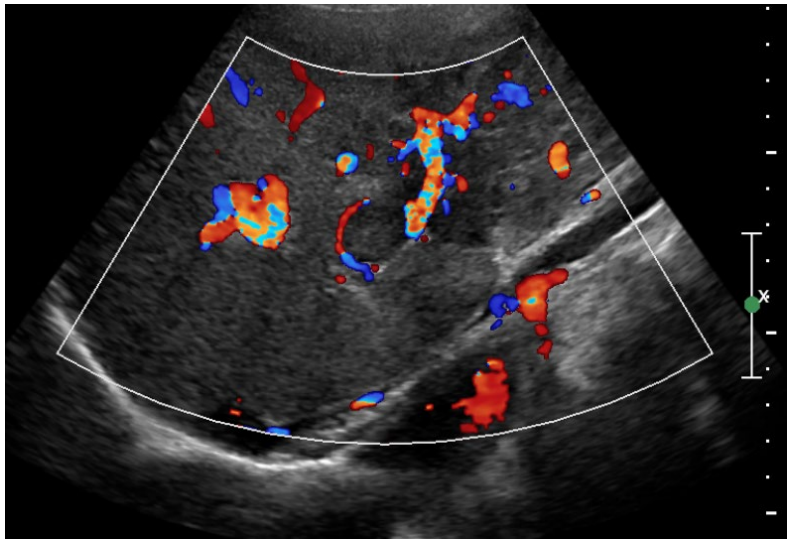


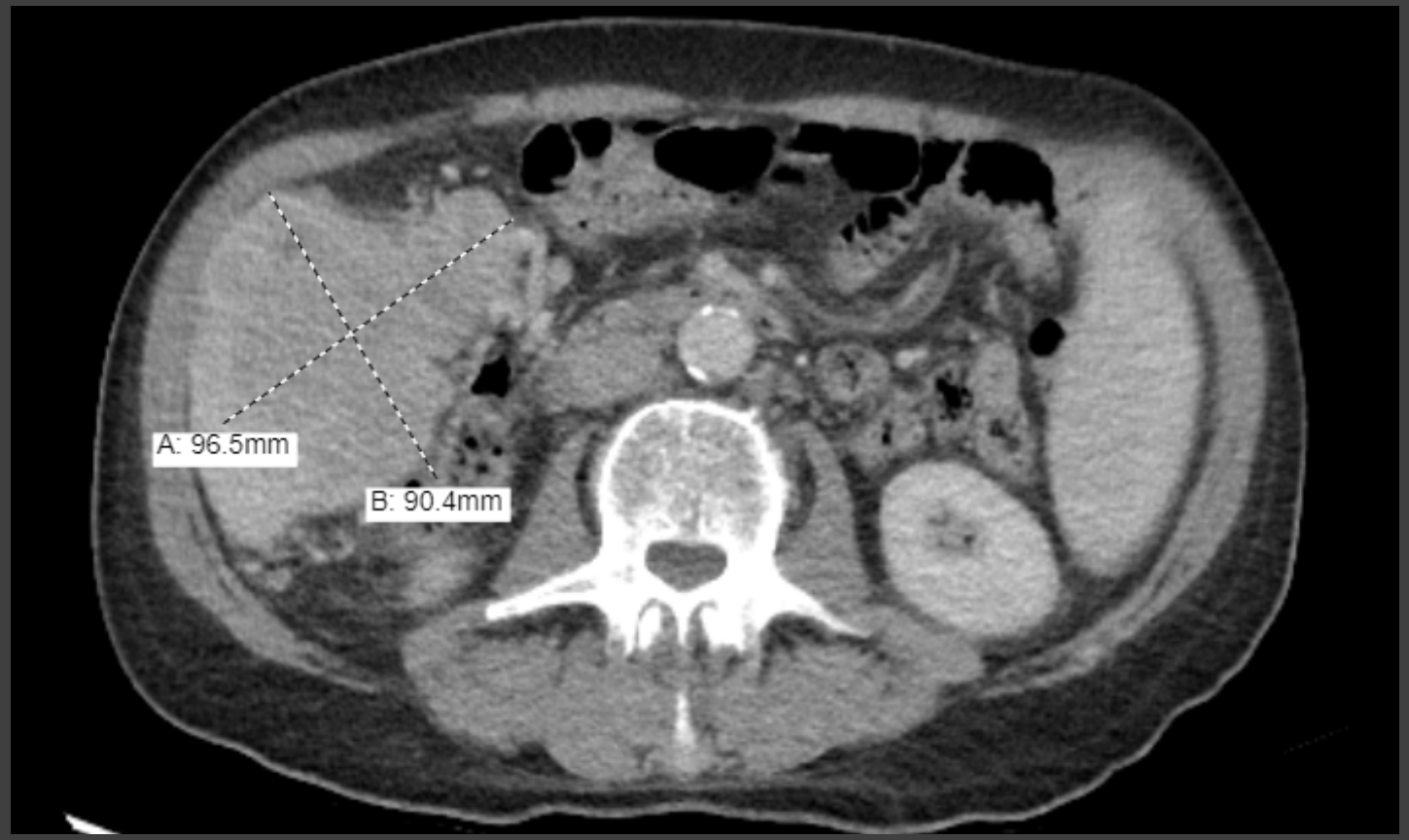
Ultrasound

Suspected thrombus of the right posterior branch of the portal vein.



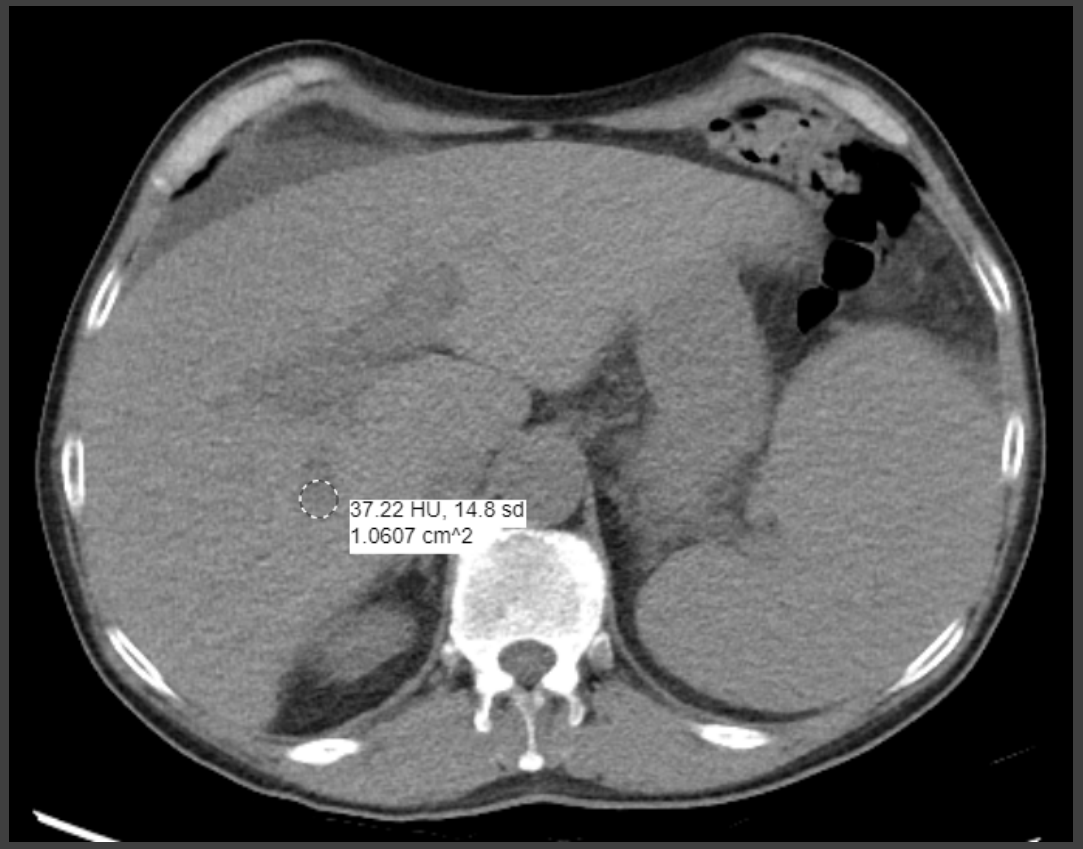


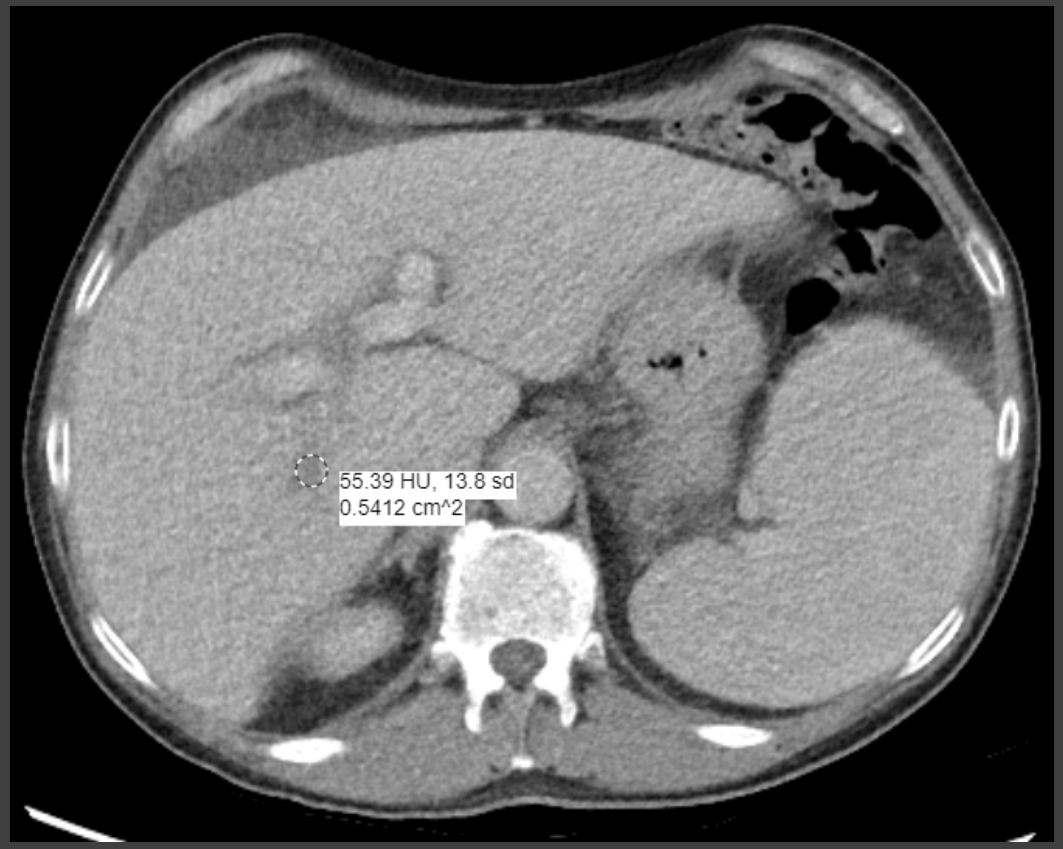




Multiphasic CT Abdomen/Pelvis

- CT findings corresponds to US findings.
- Lobulated exophytic mass from the lower pole of the right lobe, 9.7 x 9 x 15.4 cm.
- Mild heterogenous enhancement on arterial phase, with washout on subsequent PV and delayed phases.

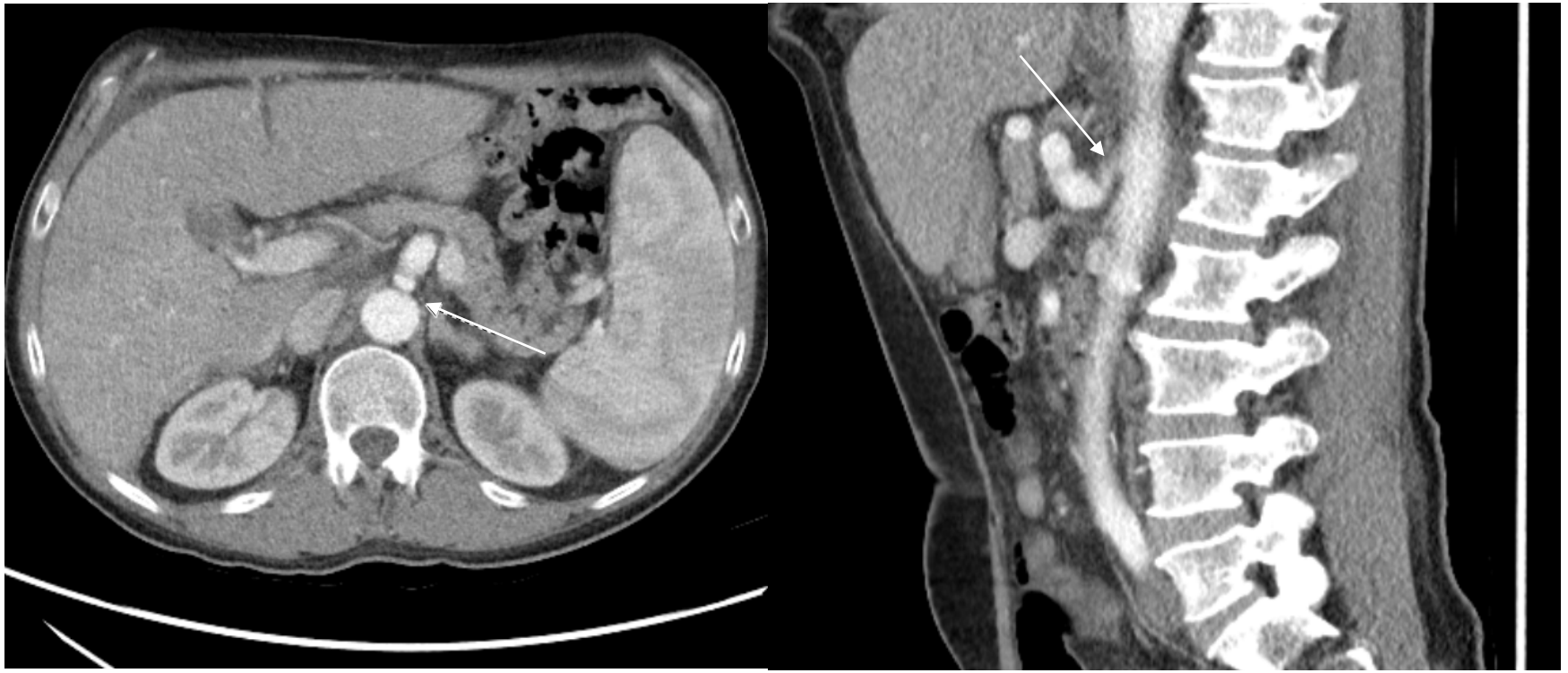




Multiphasic CT Abdomen/Pelvis

- Vascular findings:
  - 1. Occlusion of the posterior branch of the right portal vein, with enhancement, suggestive of tumor thrombus.

Multiphasic CT Abdomen/Pelvis



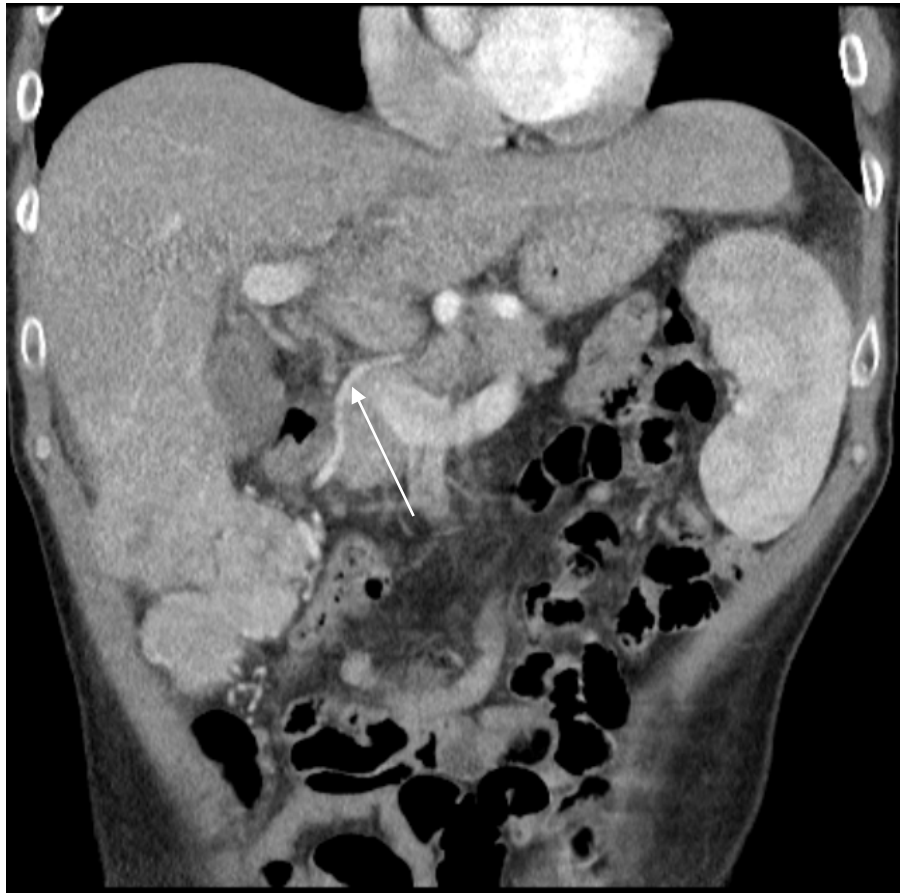
- Vascular findings:
  - 2. J-shaped celiac origin with post stenotic dilatation of 1.5 cm, suspicious for median arcuate ligament configuration of the celiac axis.

Multiphasic CT Abdomen/Pelvis

- Vascular findings:
  - 3. Collateral vessels along the inferior and posterior margin of the right hepatic lobe, possibly feeding the hepatic mass.

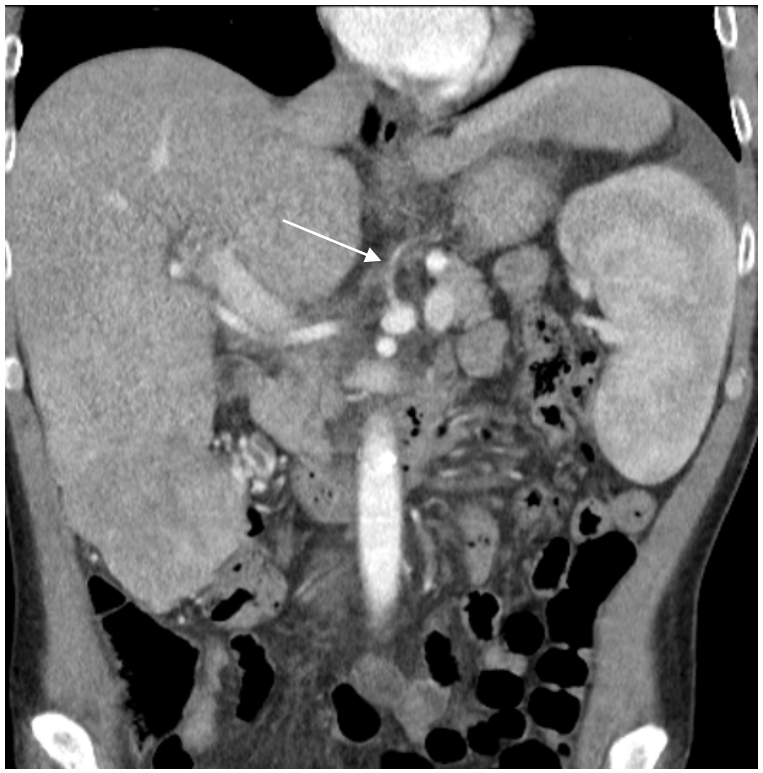
Multiphasic CT Abdomen/Pelvis

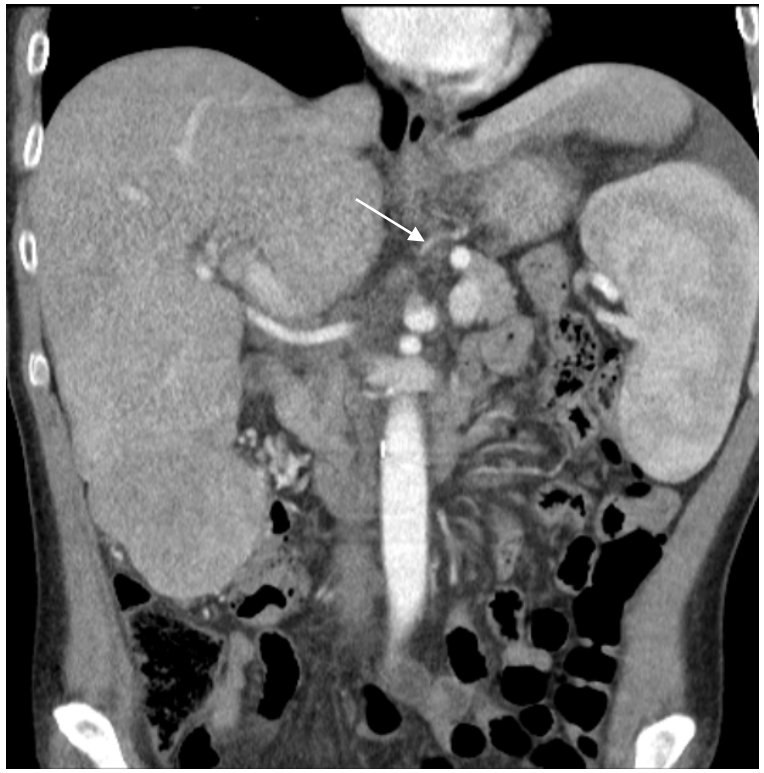
- Vascular findings:
  - 4. Some of these collateral vessels appear to be branches of the GDA, which arises directly from the celiac axis.

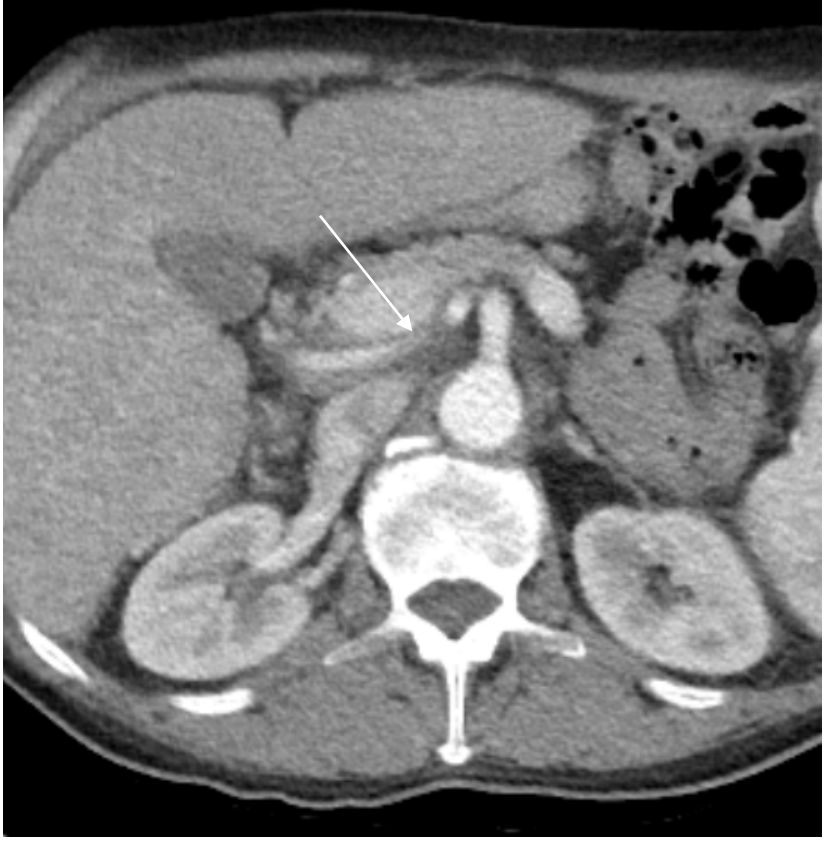


Multiphasic CT Abdomen/Pelvis

- Vascular findings:
  - 5. Replaced left hepatic artery, originating from the left gastric artery.







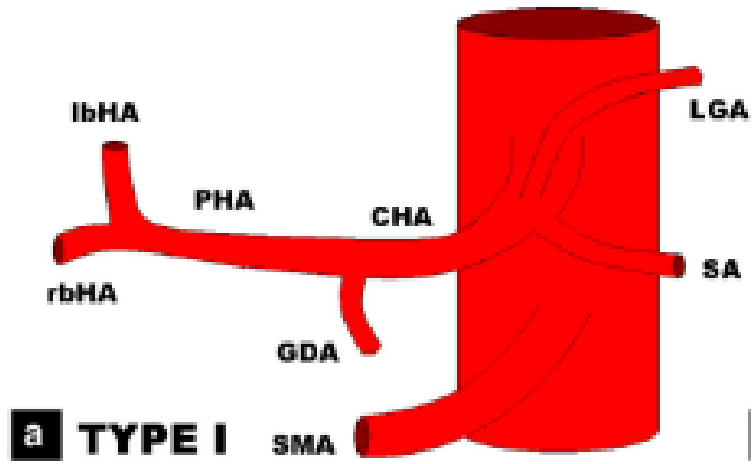
Multiphasic CT Abdomen/Pelvis

- Vascular findings:
  - 6. Replaced right hepatic artery, arising from the SMA.

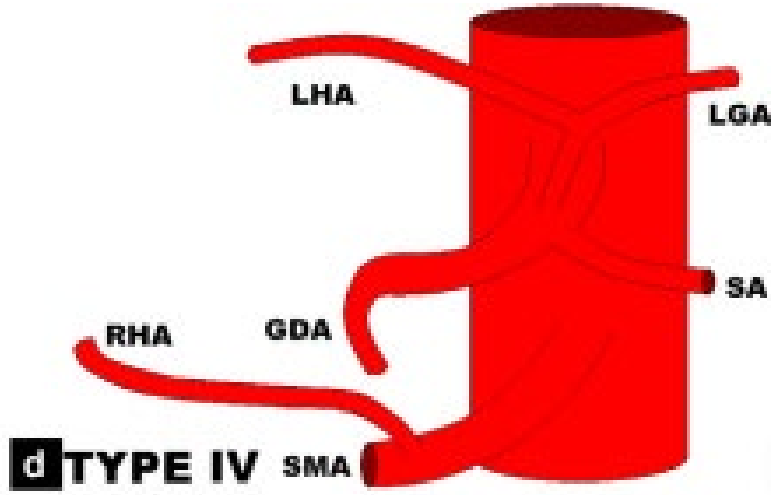
Anatomic variations of the celiac axis and hepatic arteries

- Several different classifications of hepatic artery variants exists but Michel's classification has been most often used and provides an anatomic approach [1].
- In 1955, Michel describes 10 types of hepatic artery variations.
- Our patient's anatomy is compatible with Type IV hepatic artery variant:
  - L hepatic artery from L gastric artery
  - R hepatic artery from SMA
  - GDA directly arise from celiac axis (no common hepatic artery).
- Rare variation in 1% of cases.

Conventional anatomy



Patient anatomy



[2]

Pathology

- US-guided needle core biopsy of the right liver lobe mass.
- Diagnosis:
  - Most likely primary high grade poorly differentiated hepatocellular carcinoma.
  - Hep C positive.

Metastatic Workup

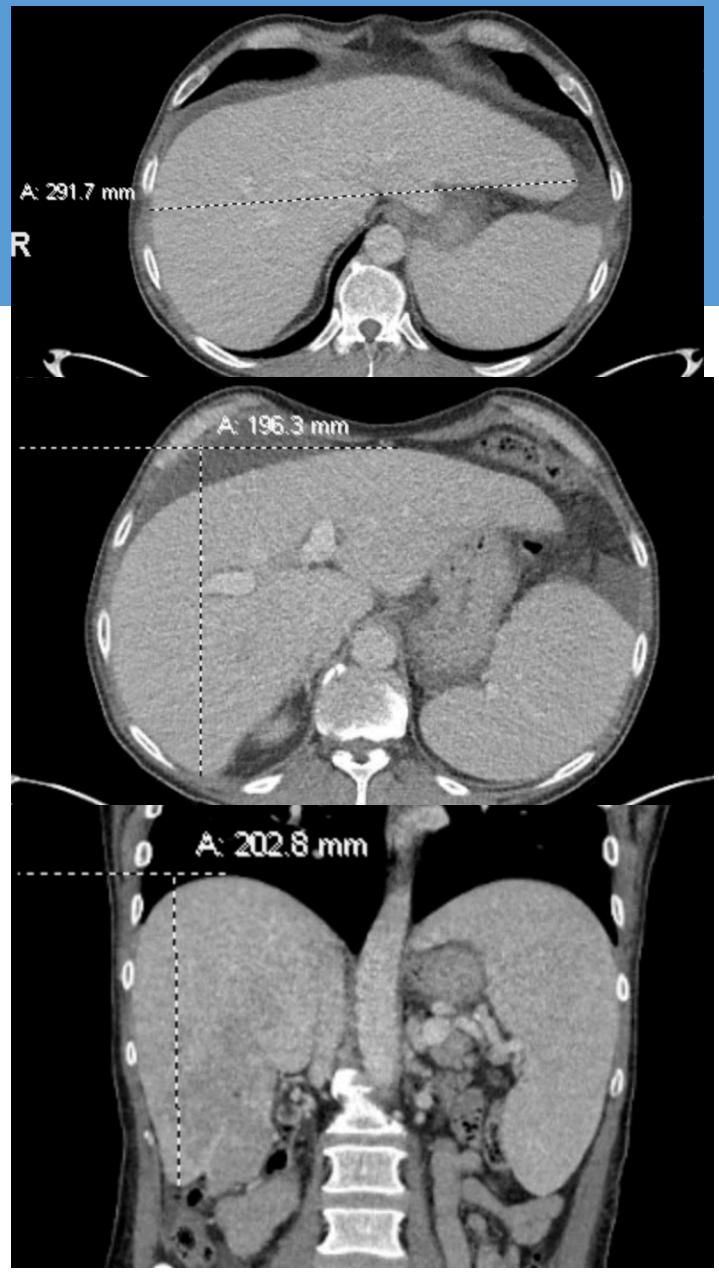
- No evidence of metastatic disease on CT neck, chest, abdomen and pelvis.
- Focal FDG uptake involving bowel loops of the RUQ/hepatic flexure.
  - Direct visualization and polypectomy with colonoscopy was negative for malignant disease.
- Bone scan demonstrated no evidence of skeletal metastases.
- Contrast enhanced liver MRI added no further diagnostic information.

Treatment Option

Lab	Pre-procedure values
Creatinine (µmol/L)	90
INR	1
Albumin (g/L)	27
Bilirubin Total (µmol/L)	9
AST (U/L)	18
ALT (U/L)	6
ALP (U/L)	94
GGT (U/L)	38

Prognostic Scores	Pre-procedure values
MELD score	7 points
ALBI score	Grade 2
Child-Pugh score	7 points Class B
Liver volume	3602 mL

Volume = CC x LL x AP x 0.31

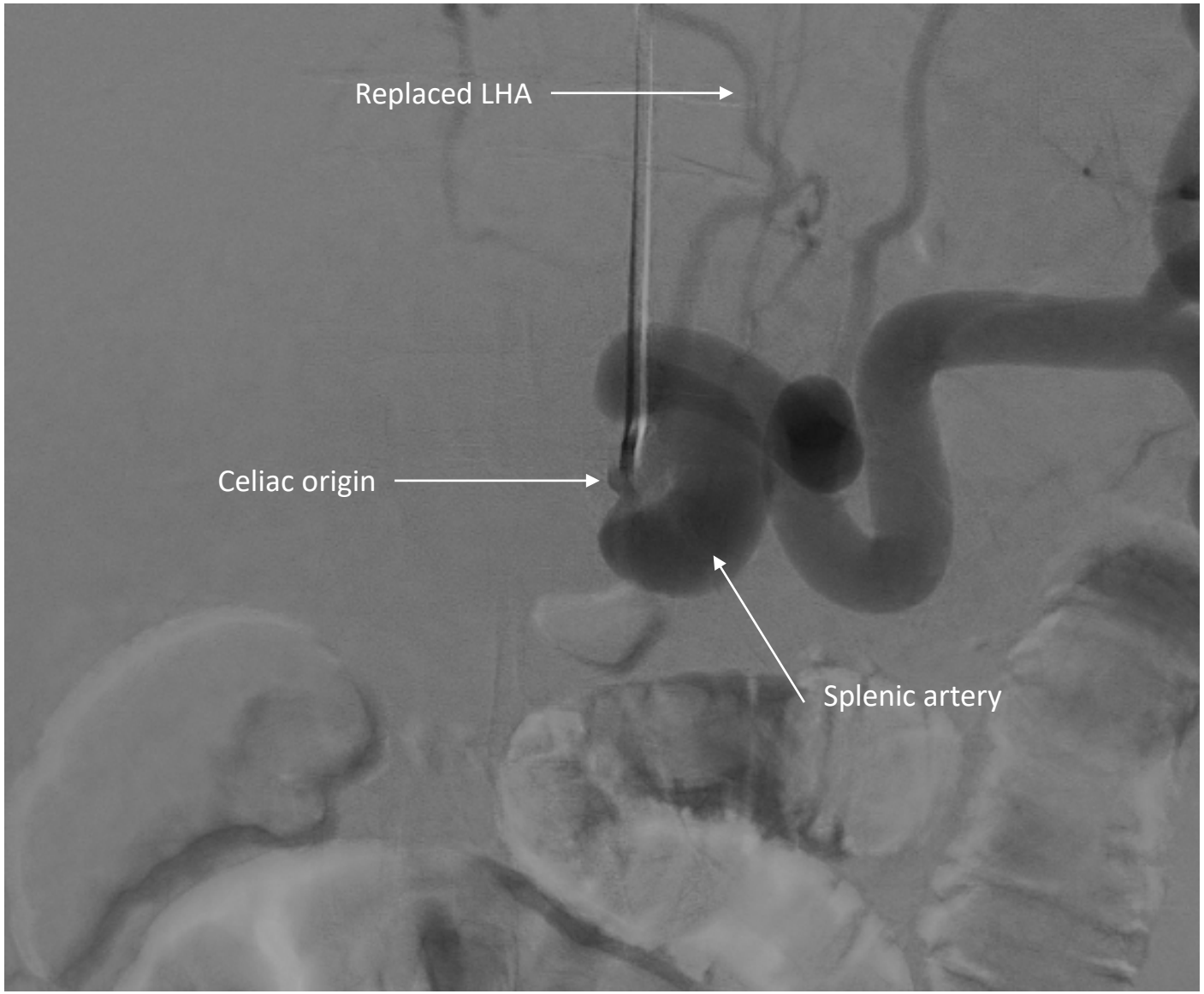


Treatment Option

- Treatment options were discussed at multidisciplinary rounds.
- Not a surgical candidate:
  - Tumor comes close to the bifurcation of the right and left portal vein, short of vein reconstruction.
  - May not be possible to get a clear margin.
- Systemic chemotherapy (sorafenib):
  - No systemic metastasis to make it clearly indicated as first step.
- Y-90 radioembolization:
  - Suboptimal due to tumor vascular supply – significant parasitized vessels from SMA.
- **TACE**
  - BCLC B/C: portal vein thrombus of a second order branch (right posterior branch), risk of hepatic infarction was less.
  - May benefit from TACE prior to systemic chemotherapy.
  - If no significant response → systemic chemotherapy.

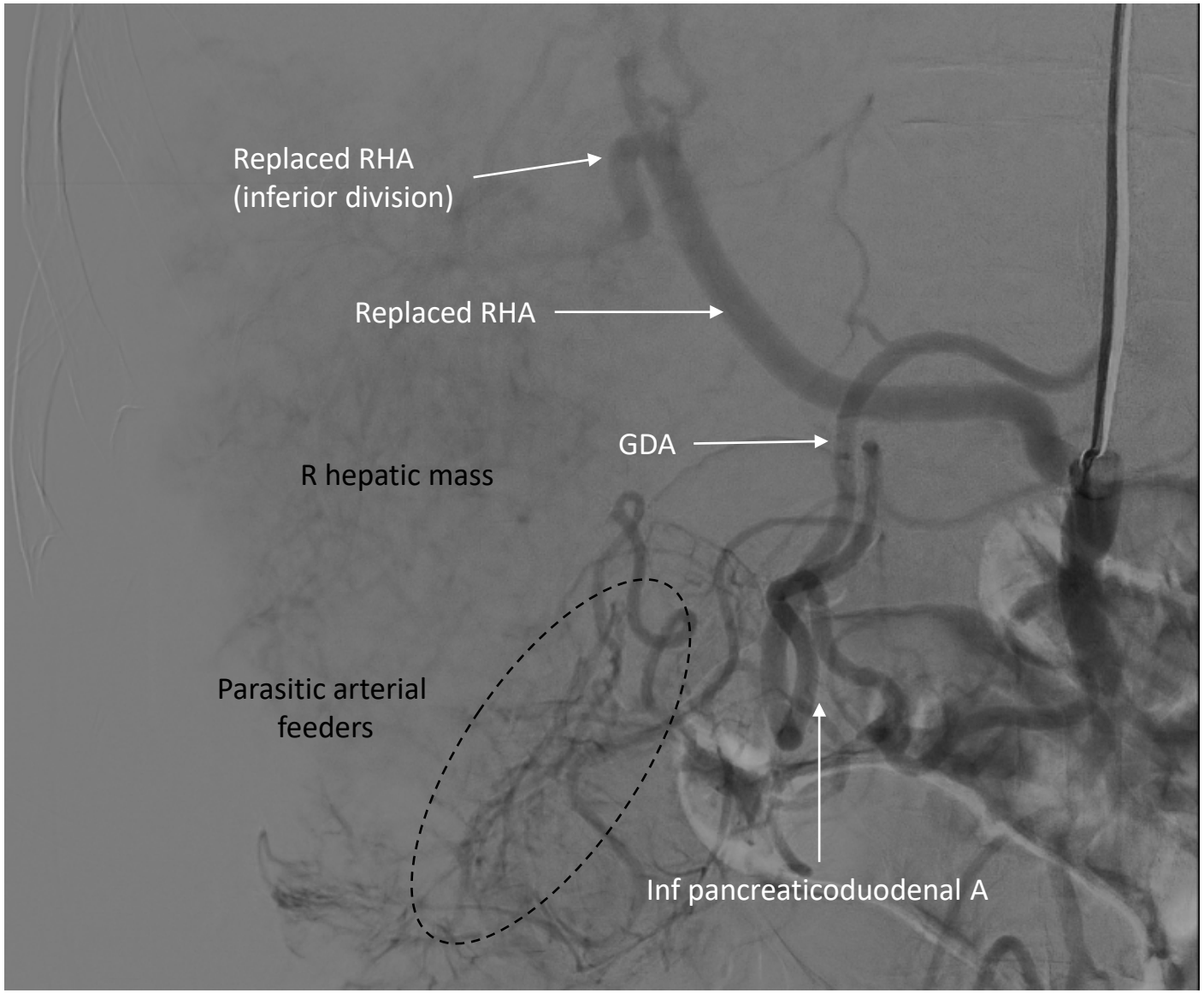
Angiography of celiac axis

- Left radial access.
- At the celiac axis, confirmation of hemodynamically significant stenosis secondary to median arcuate ligament configuration:
  - No flow seen to gastroduodenal artery (GDA).
  - Post-stenotic dilatation of the splenic artery.
- Small replaced left hepatic artery arising from the left gastric artery.
- The celiac artery was not selectively accessed due to high-grade stenosis.



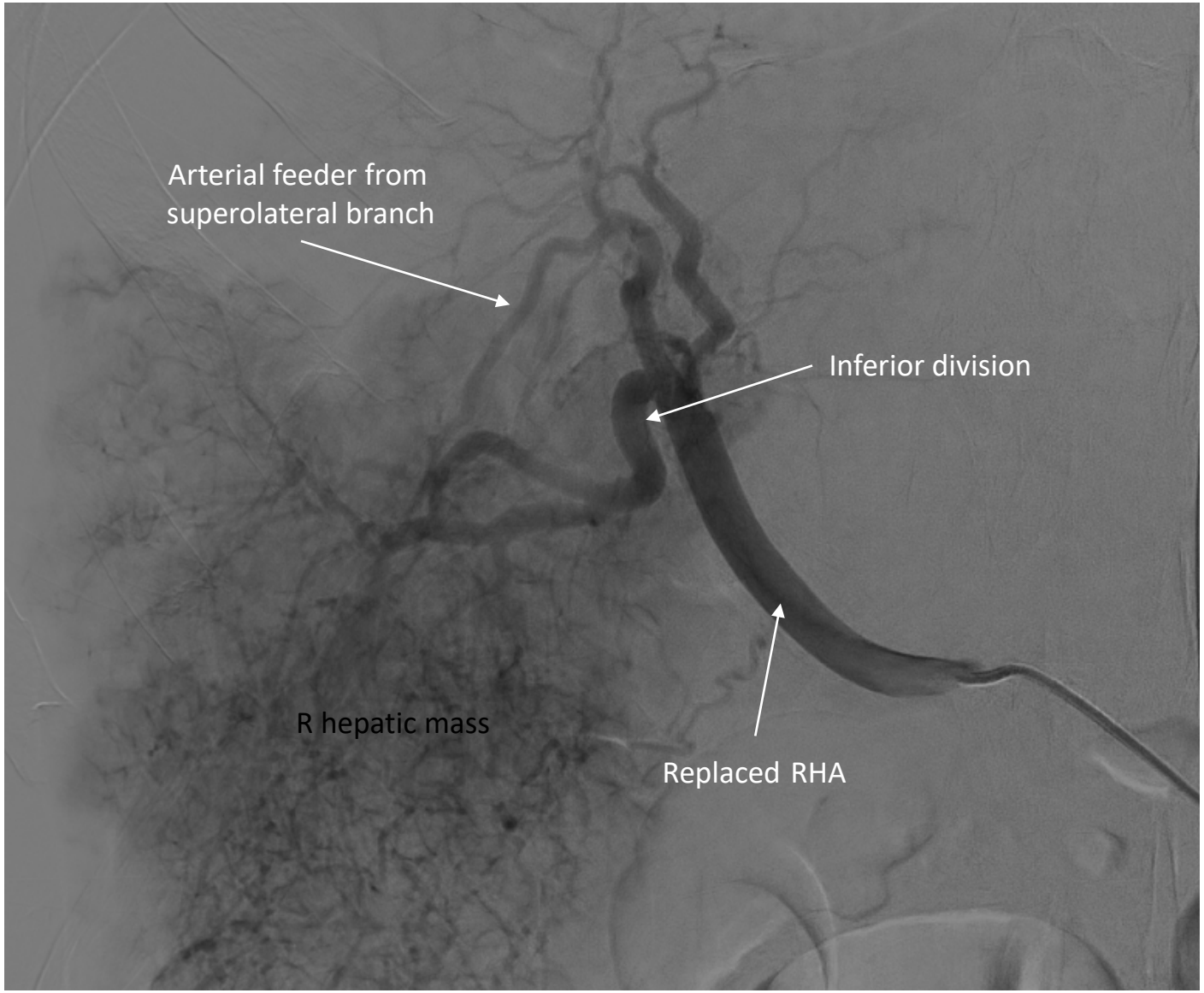
Angiography of SMA

- Confirmation of replaced right hepatic artery, with inferior division of the right hepatic artery supplying the majority of the intrahepatic tumor.
- Hypertrophic inferior pancreaticoduodenal artery, supplying the GDA.
- Multiple parasitic arterial feeders from inferior pancreaticoduodenal artery supplying the inferior and exophytic portion of the tumor.



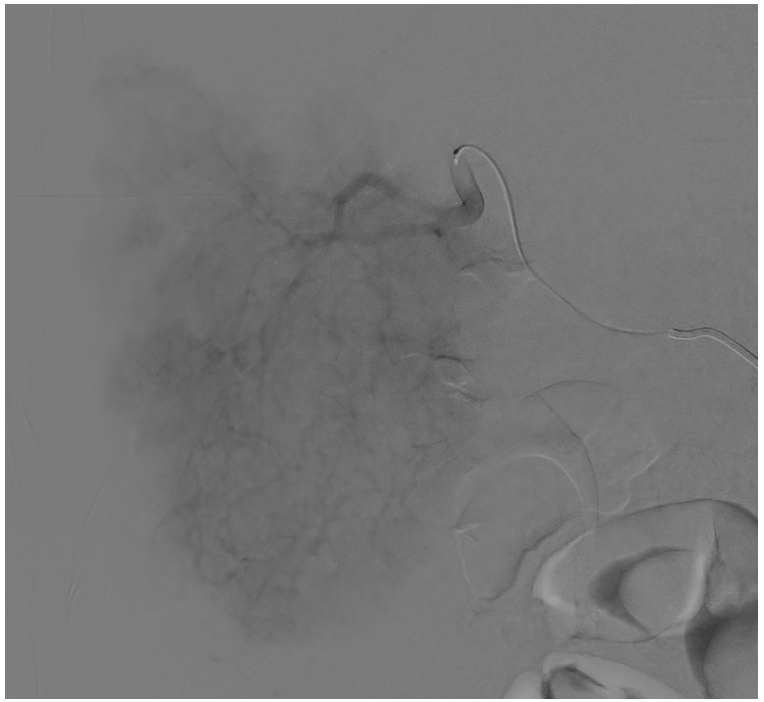
Angiography of replaced RHA

- Confirmation of replaced RHA as the primary arterial supply of the known HCC, particularly the inferior division.
- There was also a smaller additional arterial feeder from the superolateral branch of the replaced RHA.
- No definite arterial venous shunting.

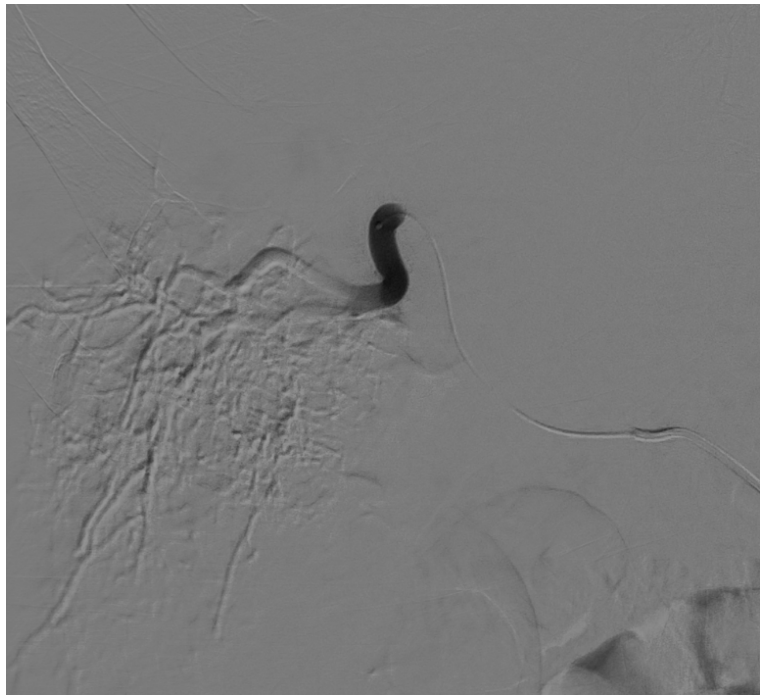


Embolization of the inferior branch of the replaced RHA

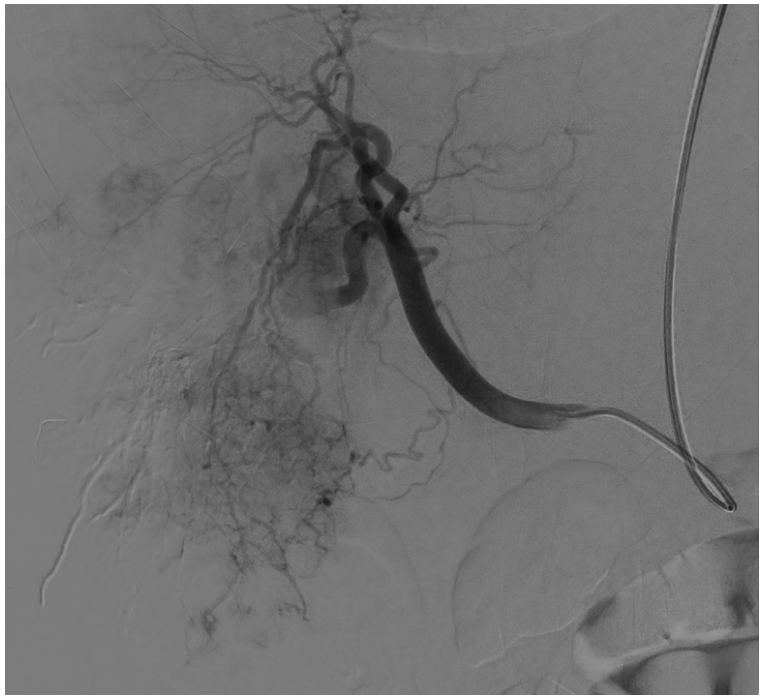
- Selection and angiography of the inferior division of the replaced RHA demonstrated extensive tumoral enhancement.
- Embolization to stasis performed using 75-150 µm DC beads loaded with 75 mg doxorubicin per vial, followed by 3 vials of 300-500 µm DC beads.



Inferior branch of replaced RHA angiography pre-embolization, demonstrating tumoral enhancement.



Inferior branch of replaced RHA digital subtraction angiography post-embolization, demonstrating stasis.



Replaced RHA angiography post inf branch embolization, demonstrating tumoral enhancement from superolateral arterial feeder.

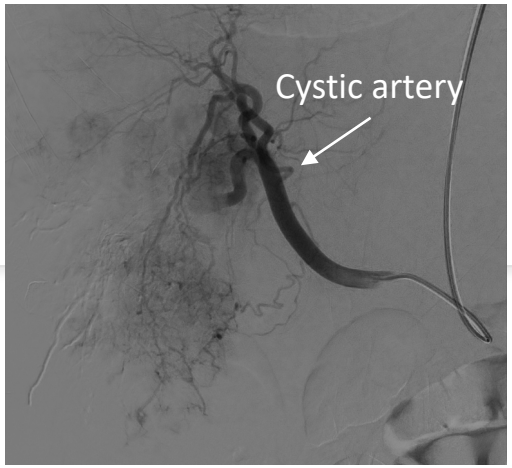
Embolization of suprolateral arterial feeder of the replaced RHA

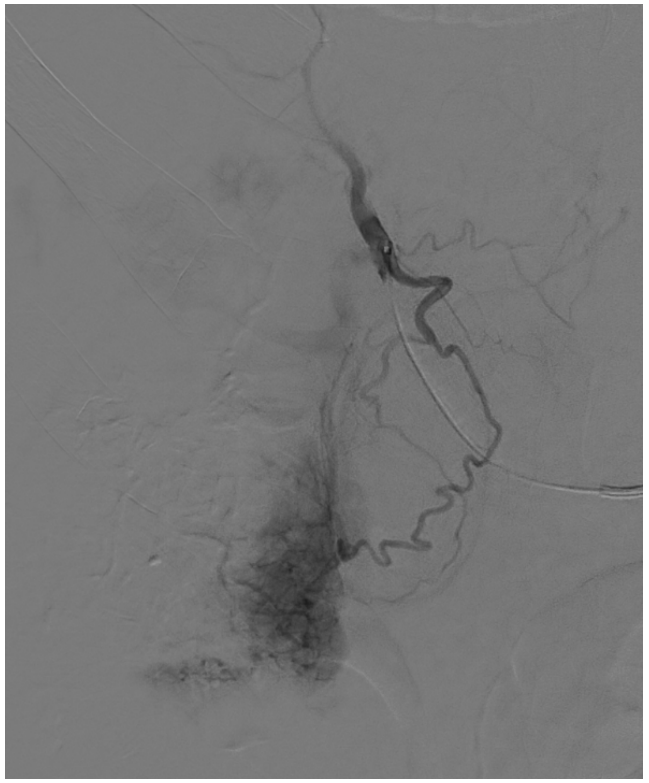
Superolateral branch of replaced RHA angiography pre-embolization, demonstrating tumoral enhancement.

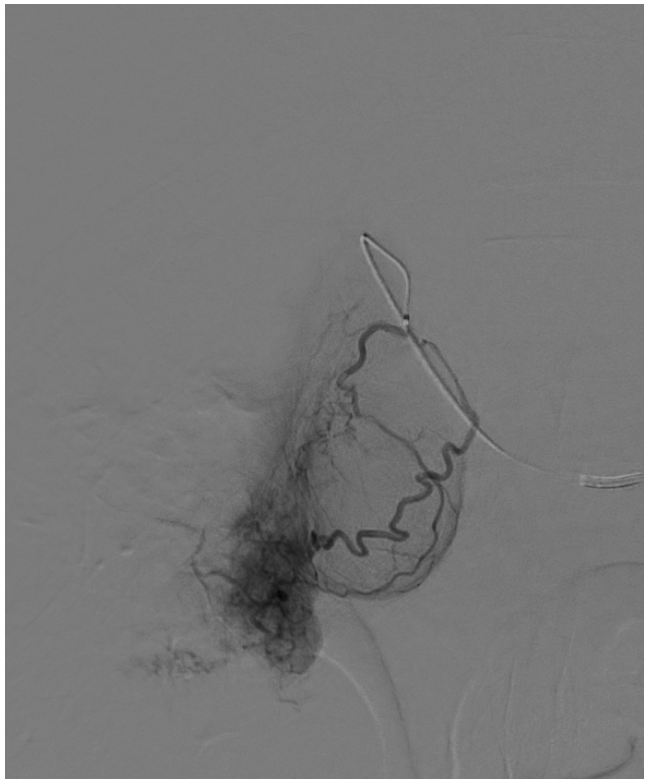
Superolateral branch of replaced RHA angiography post-embolization, demonstrating stasis.

- Selection and angiography of the superolateral branch of the replaced RHA demonstrated extensive tumoral enhancement.
- Embolization to stasis performed using 0.2 vials of 300-500 µm DC beads.

Cystic artery angiography

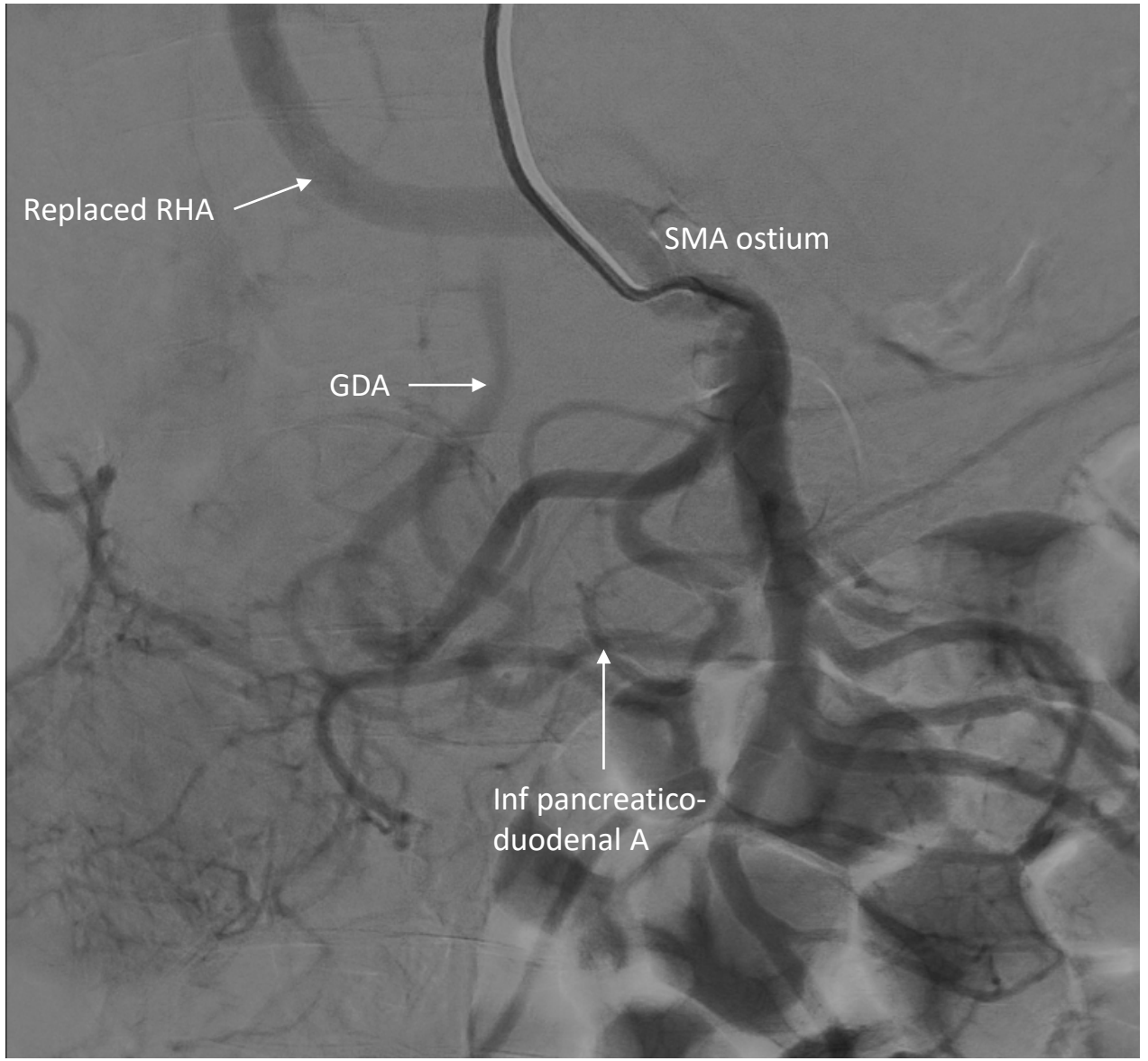






- Tumoral enhancement was seen from a distal branch of the cystic artery.
- However, distal embolization sparing the cystic artery was felt impossible.

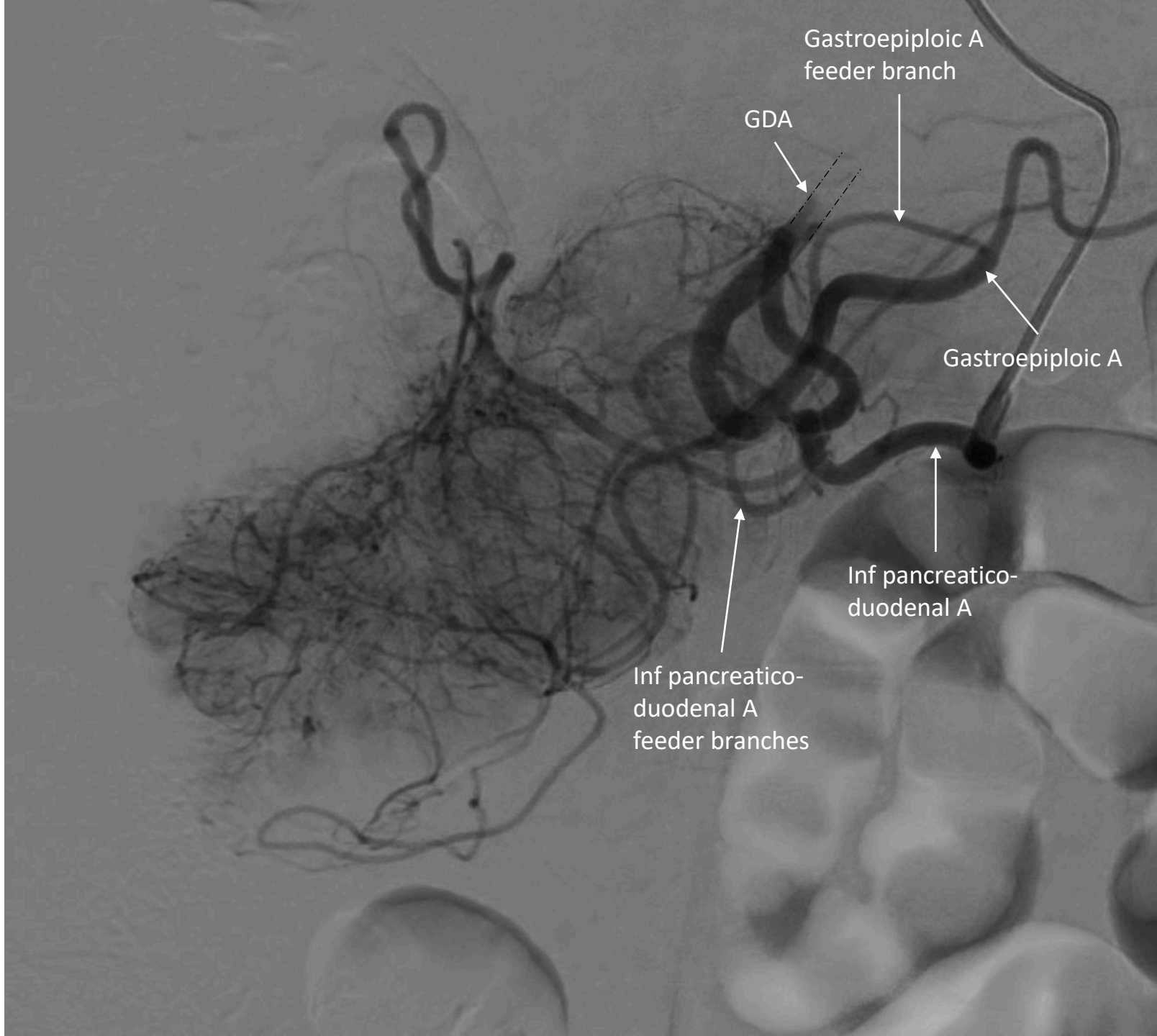
Attention was turned to the inferior pancreaticoduodenal arcade…



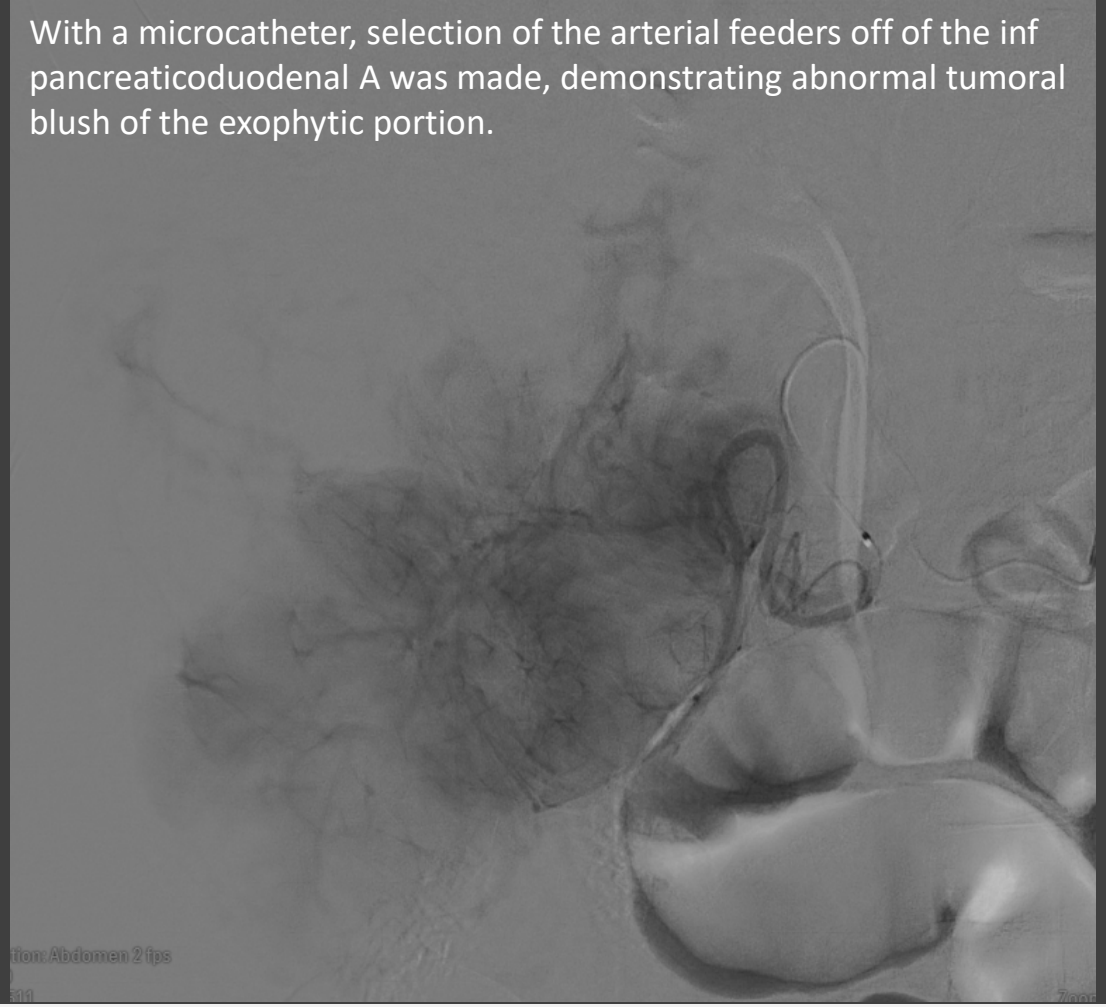
Angiography of the SMA axis.

Selection of the inferior pancreaticoduodenal artery was difficult but…

- Confirmed parasitic arterial supply via:
  - Multiple branches of the inferior pancreaticoduodenal artery.
  - Small branch from the gastroepiploic artery.
- Interestingly, when the inferior pcancreaticoduodenal artery was completely occluded by the catheter, a filling defect appeared within the GDA, which confirmed altered flow dynamics/reversal of flow originating from the celiac axis.



With a microcatheter, selection of the arterial feeders off of the inf pancreaticoduodenal A was made, demonstrating abnormal tumoral blush of the exophytic portion.



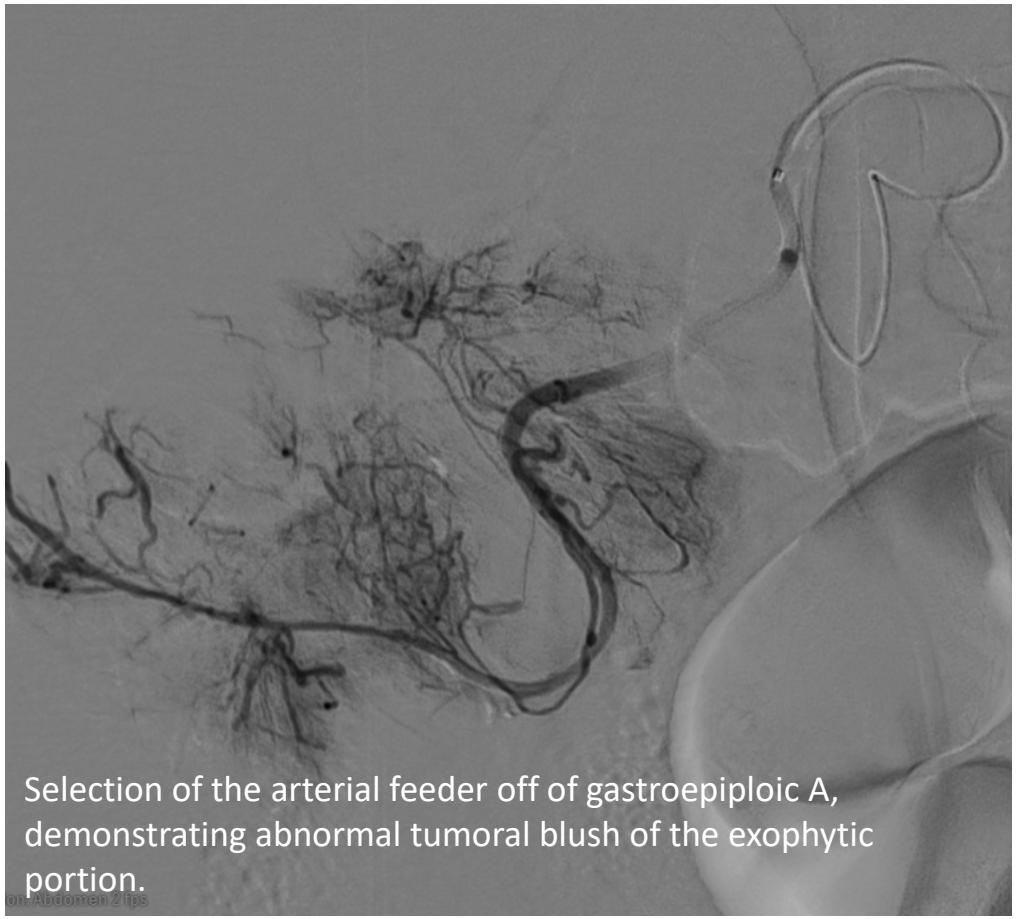
Embolization to stasis.

Embolization of inferior pancreaticoduodenal feeders

- Selection of inf pancreaticoduodenal feeders demonstrated tumoral blush, with no evidence of significant shunting or definite supply to the bowels.
- Embolization performed with 1 vial of 75-150 µm DC beads loaded with 75 mg of doxorubicin, followed by 1.5 vials of 300-500 µm DC beads to stasis.

Embolization of the gastroepiploic feeder

- A smaller branch arising off of the gastroepiploic artery was selected and embolization was performed using 0.3 vials of 300-500 µm DC beads to stasis.



Selection of the arterial feeder off of gastroepiploic A, demonstrating abnormal tumoral blush of the exophytic portion.

Embolization to stasis.

Post-embolization angiography

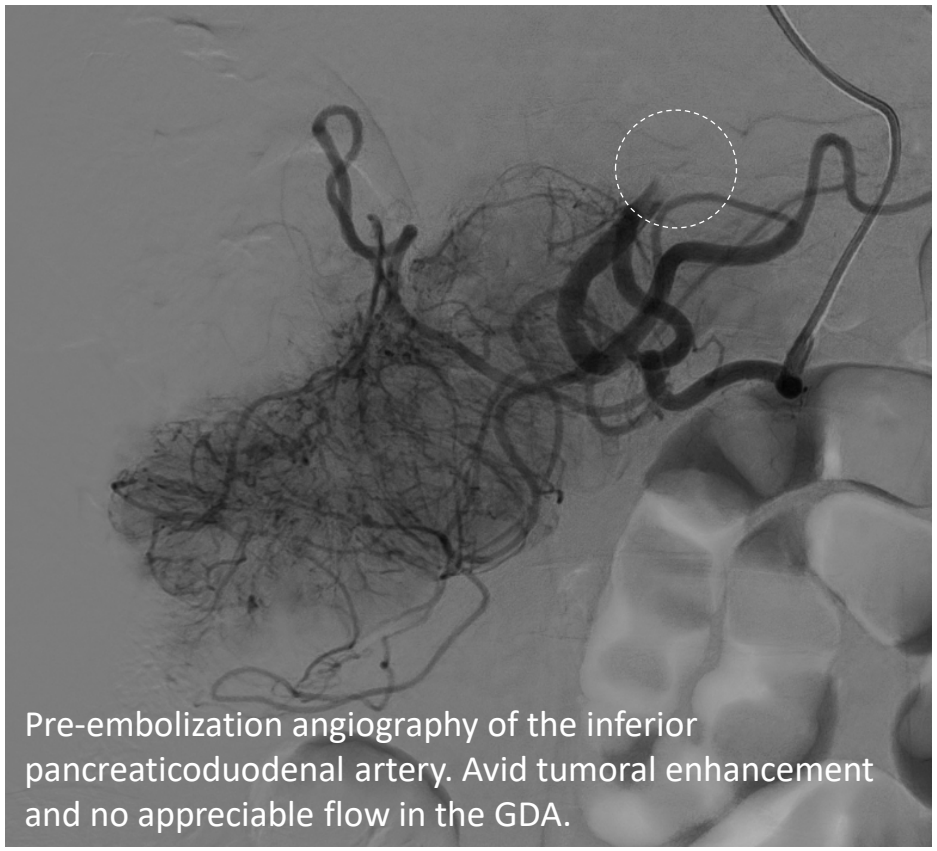
- Microcatheter was removed.
- Repeat angiography revealed no significant residual tumoral enhancement via the replaced right hepatic, inferior pancreaticoduodenal/gastroduodenal, and gastroepiploic arteries.

Post-embolization inferior pancreaticoduodenal angiography.

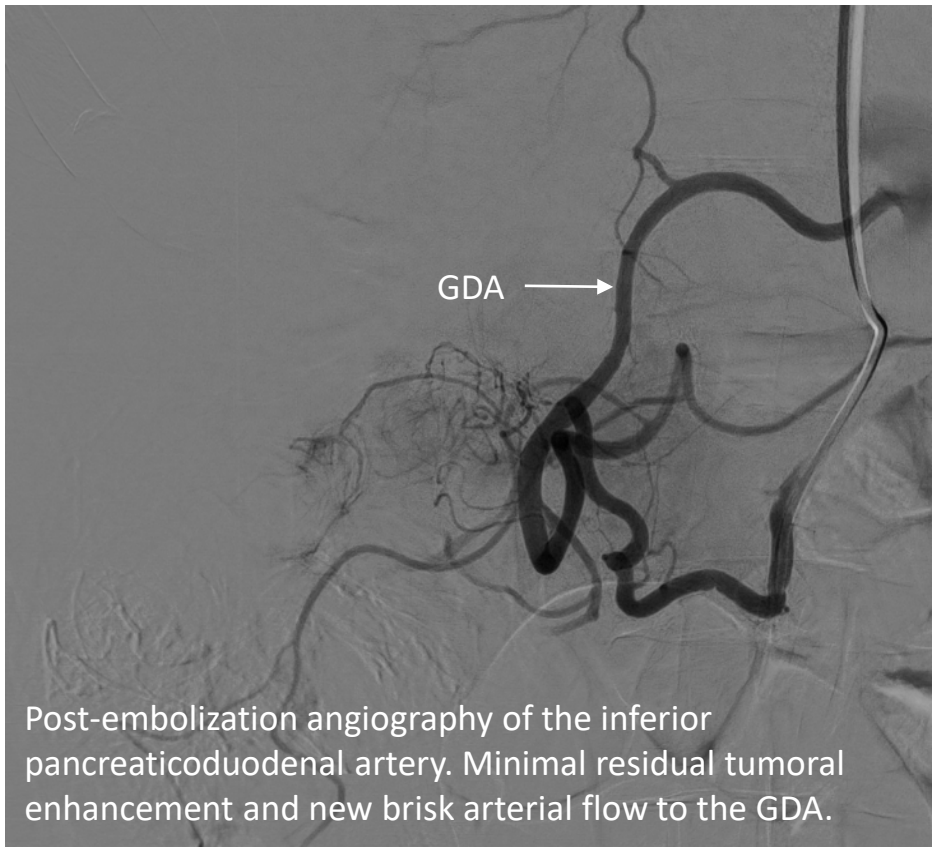
Post-embolization SMA angiography.

Post-embolization alteration in flow dynamics

- Secondary to altered flow dynamics with the parasitic arterial feeders off inferior pancreaticoduodenal/GDA and gastroepiploic arteries now occluded, there is increased brisk retrograde opacification of the GDA from the SMA, which ultimately retrograde into the celiac axis.



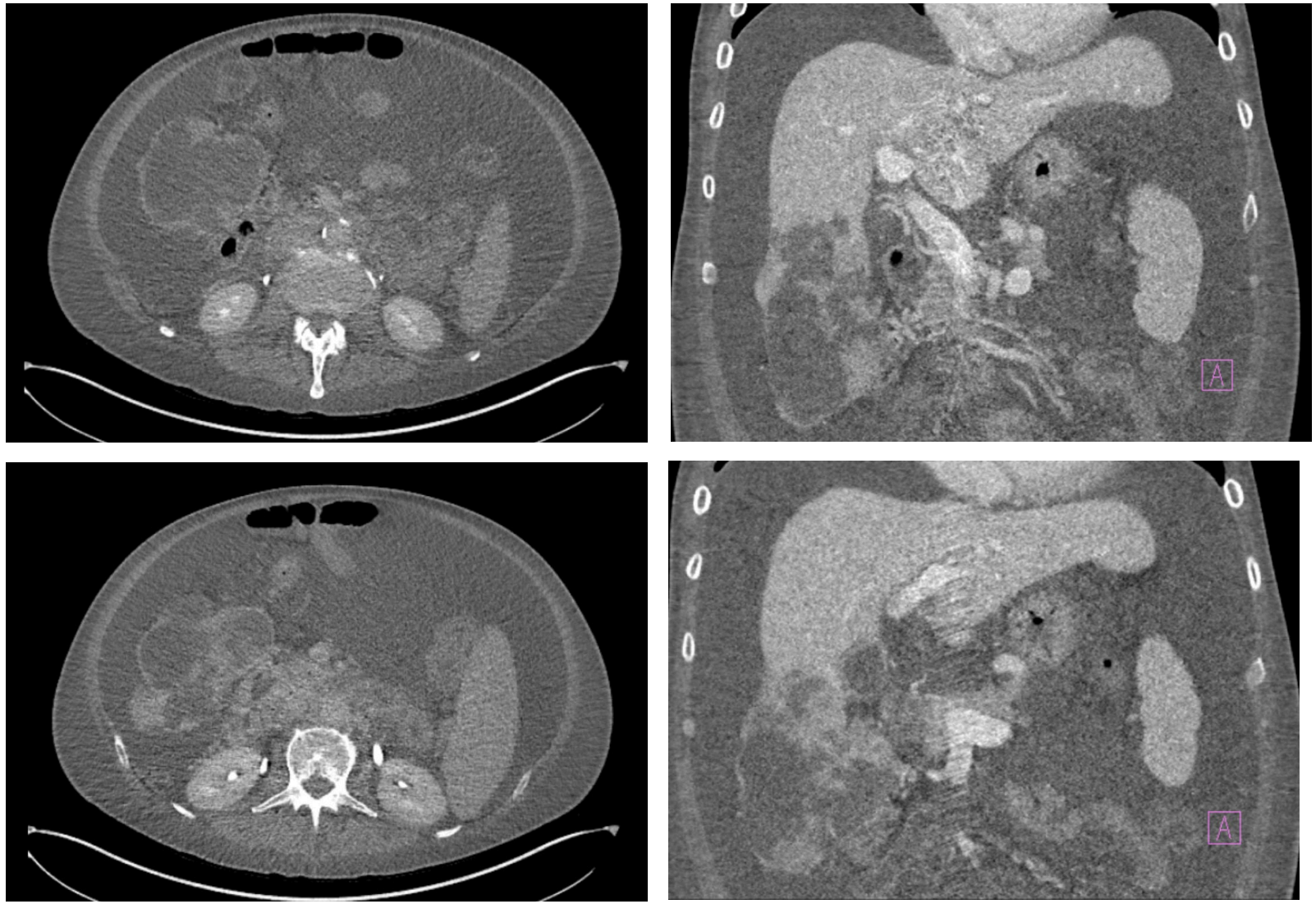
Pre-embolization angiography of the inferior pancreaticoduodenal artery. Avid tumoral enhancement and no appreciable flow in the GDA.



Post-embolization angiography of the inferior pancreaticoduodenal artery. Minimal residual tumoral enhancement and new brisk arterial flow to the GDA.

1-month post-embolization

- Significant tumoral necrosis achieved.
- New ascites.



Post-embolization course

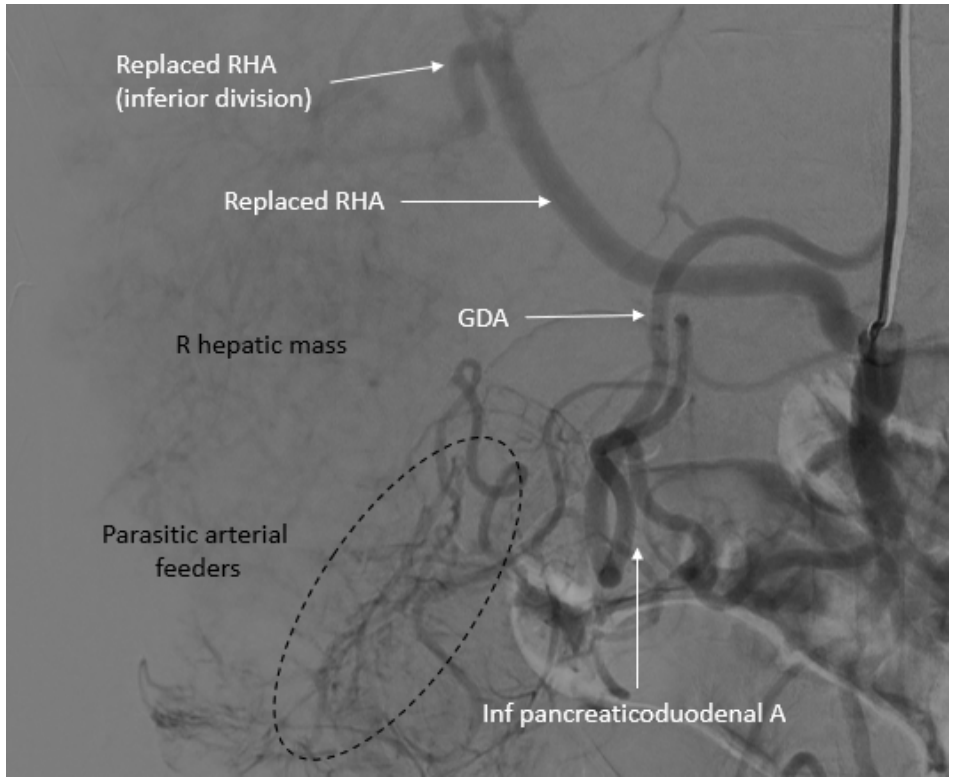
Prognostic Scores	Pre-procedure	Post-procedure
MELD score	7 points	6 points
ALBI score	Grade 2	Grade 2
Child-Pugh score	7 points Class B	8 points Class B
Liver volume	3602 mL	2160 mL

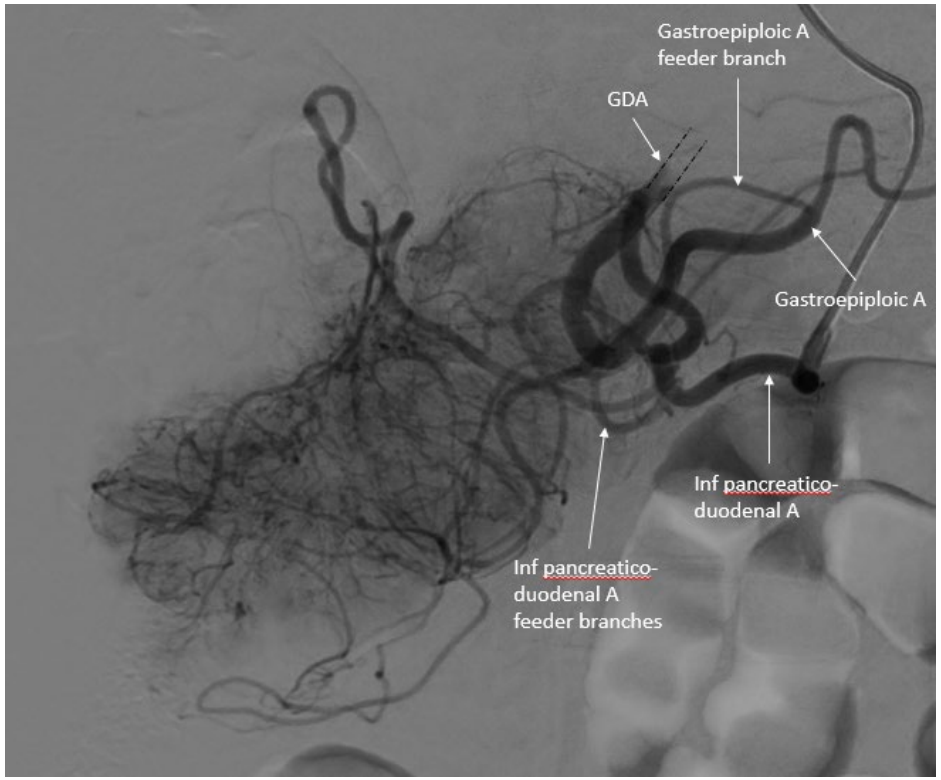
Lab	Pre-procedure	Immediate post	2 months post
Creatinine (µmol/L)	90	74	68
INR	1	1.1	1
Albumin (g/L)	27	25	29
Bilirubin Total (µmol/L)	9	7	8
AST (U/L)	18	24	16
ALT (U/L)	6	11	5
ALP (U/L)	94	114*	101
GGT (U/L)	38	-	-

- Up to 2-month post procedure, patient was not encephalitic, did not require re-admission, and LFTs remained stable.
- However, new ascites… cause?
  - ?Reactive ascites from significant tumoral necrosis
  - ?Decompensated liver failure from parenchymal atrophy and portal hypertension.
    - Although liver function was preserved pre-procedure, may have a decreased degree of liver function reserve (ie. large liver and spleen, dilated portal and splenic veins → ?portal HTN and at least hepatic fibrosis.
    - Serves to show that liver function status should be evaluated beyond the conventional Child-Pugh/MELD score classification, taking into account the compensated or decompensated status of the patient.

Practical Points

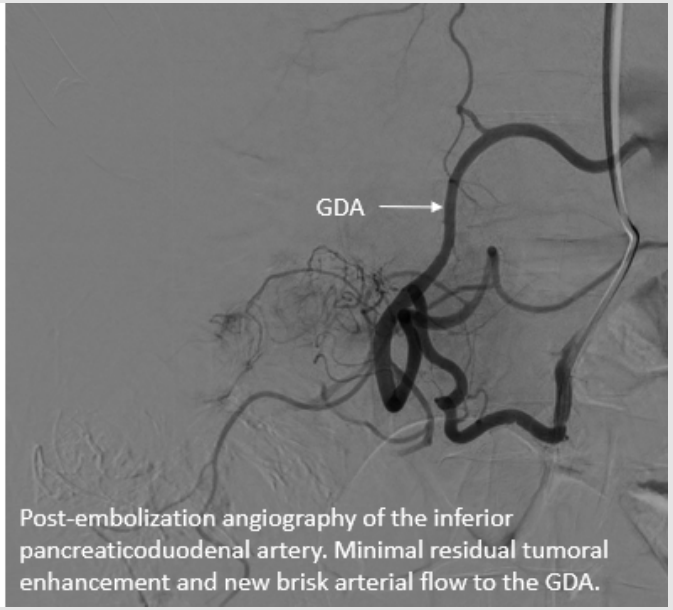
- Our case proves two important points:
- Point 1: Antegrade/retrograde flow dynamic changes depending on the position of the catheter and occlusion of the vessel by the catheter.





Practical Points

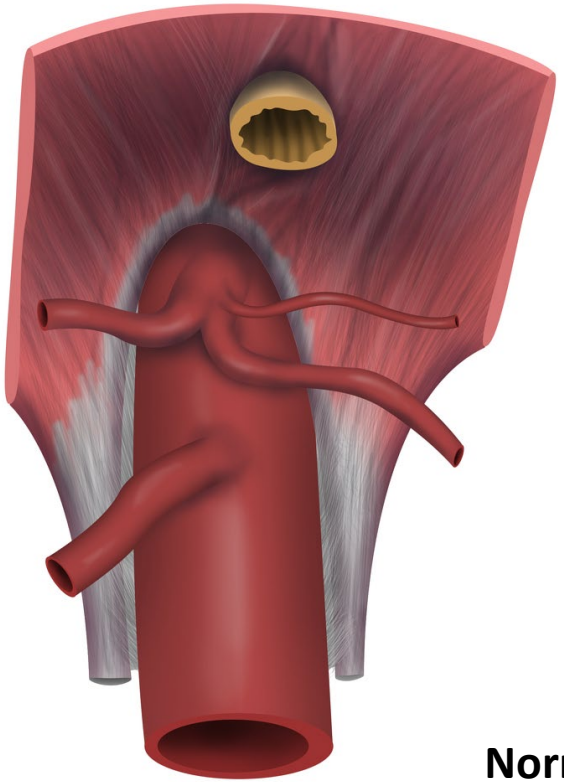
Pre-embolization angiography of the inferior pancreaticoduodenal artery. Avid tumoral enhancement and no appreciable flow in the GDA.



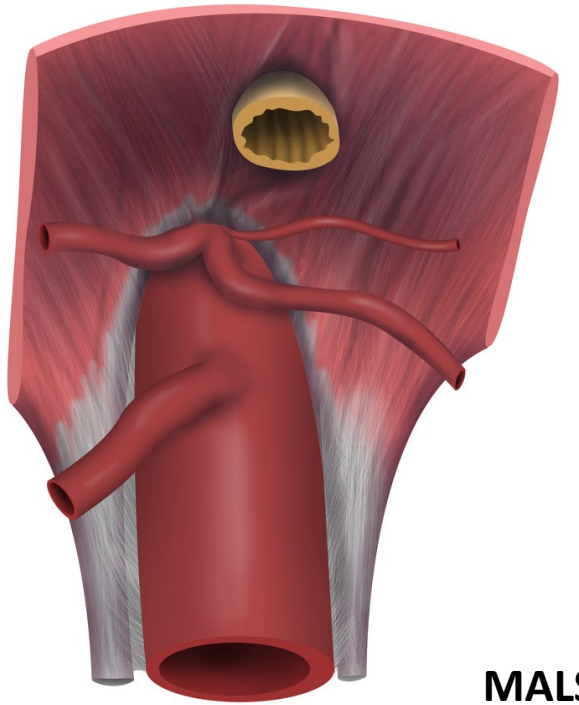
Post-embolization angiography of the inferior pancreaticoduodenal artery. Minimal residual tumoral enhancement and new brisk arterial flow to the GDA.

- Point 2: Preferential flow to/from the celiac axis changes dynamically depending on the degree of embolization of parasitized tumoral vessels.
- Therefore, embolization should be performed as selective as possible to decrease the risk of non-target embolization, which may behave in dynamic and unexpected ways.

Celiac artery stenosis



Normal



MALS

[3]

- Common condition caused by arteriosclerosis or compression by the median arcuate ligament, with an incidence ranging from 12.5-24%. [4, 5]
- Median arcuate ligament syndrome (MALS):
  - Common Ligament at the base of the diaphragm that unites the left and right diaphragmatic crura near T12 → forming a fibrous arch at the anterior aspect of the aortic hiatus.
  - When ligament is positioned more inferiorly relative to the celiac artery → celiac compression, most accentuated during end-expiration.
- Imaging features of celiac artery compression: [6]
  - "J" appearance of proximal celiac trunk on CT median sagittal sections in the arterial phase, due to focal narrowing and post-stenotic dilatation.
  - Dilatation of the pancreaticoduodenal arcade in absence of atherosclerosis.
- Often clinically asymptomatic due to the rich existing collateral circulation.

Collateral pathways in patients with celiac axis stenosis

Most frequently encountered collateral vessels from the SMA in patients with celiac axis stenosis are:

- Pancreaticoduodenal arcade (may develop as a single or double channel)
- Dorsal pancreatic artery
- SMA to R hepatic artery

In cases of hepatic artery variants, collateralization may come from the:

- Intrahepatic interlobar collateral vessels
- R gastric to L gastric arterial anastomoses
- L hepatic to L gastric arterial anastomoses
- Peribiliary arterial plexuses

[7]

Crossing a tight celiac axis

- Antegrade approach can often be successful.
  - Pedersoli et al. 2020: [8]
    - Recanalization in combination with celiac artery stent implantation in 39 (91%) of 43 patients was successful with an antegrade approach via the aorta.
    - Passage was only possible through a retrograde approach via the SMA and pancreaticoduodenal arcade in 4 (9%) patients.
  - Fioole et al. 2010: [9]
    - Occlusion/stenosis of the celiac trunk was successfully crossed endovascularly in 27 (90%) of 30 patients undergoing percutaneous transluminal angioplasty for chronic mesenteric ischemia.
- Access via the left brachial/radial artery may be more advantageous.
  - Given the acute angle between the celiac artery and the aorta. [10, 11]

Crossing a tight celiac axis

- Retrograde approach through the pancreaticoduodenal artery is a good alternative:
  - Kwon et al. 2002: [12]
    - The pancreaticoduodenal arcade was successful catheterized in 23 (64%) of 36 patients with celiac axis stenosis.
- Though limited by: [8]
  - Patency of the GDA and inf pancreaticoduodenal arcade.
  - Long lasting intervention and high x-ray exposure
    - Mean radiation dose in the retrograde group was 104,266 cGy/cm2 vs 42,256 cGy/cm2 in the antegrade group.
  - Increased amount of contrast used (172 ml vs 128 ml).
  - Increased necessity of analgesic medication. (50% vs 15%).
- And can rarely be complicated by: [13]
  - Non-target embolization to the spleen, when there is a reflux of embolic material with reversal of flow in the common hepatic artery.

Take-home points

Endovascular management in cases of celiac axis stenosis/occlusion is a highly challenging situation. Radial access is the preferred route of access.

Technical success heavily depends on anatomical knowledge ofthe arterial collateral supply, which can be identifies pre-intervention with spiral CT.

Antegrade access via the stenosed celiac axis is often amenable and success can mean improved procedural parameters for the patient (shorter procedure time, radiation exposure, and contrast use).

However, retrograde access via the pancreaticoduodenal arcade is a great alternative, with awareness of the implications in flow dynamics and technical downfalls.

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Thank you for listening!

Questions?