

AMSER Case of the Month

March 2023

57-year-old female with chronic epistaxis



Patrick Young, MS4
Midwestern University – Arizona College of Osteopathic Medicine

Jason Pierce, DO (PGY-5), Ruchi Yadav (Staff Radiologist)
Cleveland Clinic Foundation



Patient Presentation

- 57-year-old female presented with frequent episodes of epistaxis and melena. C/o fatigue, malaise, and increased SOB.
- One week ago, presented to the ED with HgB of 4.8, requiring 3 units of pRBC. Has needed 2-3 units of pRBC on a monthly basis, but required more transfusions recently.
- PMH: anemia 2/2 chronic GI bleed, COPD, indeterminate liver mass, multiple small intestinal angiodysplasias
- Exam: telangiectasias on lips, 2/6 systolic murmur auscultated in abdomen

Pertinent Labs

CBC

- WBC: 6.93
- HB: 7.6 (L)
- HCT: 25.2 (L)
- PLT: 338

BMP

- Na: 137
- K: 4.2
- Cl: 105
- CO2: 25
- BUN: 12
- Creatinine: 0.7
- Glucose: 93
- Calcium: 9.2

Coag Studies

- INR: 1.0
- aPTT: 26.3

What Imaging Should We Order?

ACR Appropriateness Criteria

Clinical Condition: Clinically Suspected Pulmonary Arteriovenous Malformation (PAVM)

Radiologic Procedure	Rating	Comments	RRL*
US echocardiography transthoracic with IV contrast	8		○
CTA chest with IV contrast	8	This procedure is often used following positive TTE.	☼☼☼
X-ray chest	7	This procedure is complementary to other examinations, such as TTE.	☼
US echocardiography transesophageal with IV contrast	6	This procedure is the reference standard for detecting right-to-left shunts but is more invasive than TTE.	○
MRA chest without and with IV contrast	6		○
CT chest without IV contrast	6		☼☼☼
Arteriography pulmonary	5	Although this procedure is appropriate for preinterventional planning, it is usually not appropriate as an initial test.	☼☼☼☼
US transcranial with IV contrast	5	This procedure is an alternative to TTE, although it is less widely available.	○
Pertechnetate albumin pulmonary scan	4		☼☼☼
MRA chest without IV contrast	3		○

This imaging modality was ordered by the IM hospitalist following a TTE

Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level



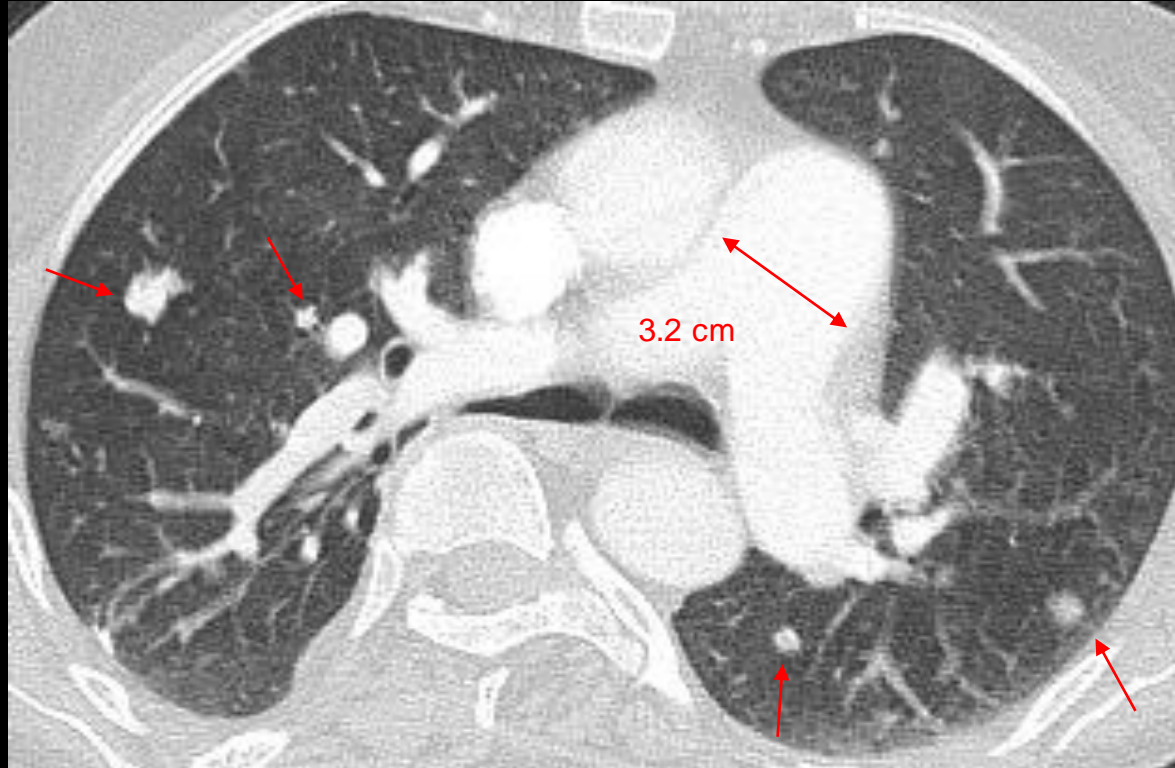
CTA Chest (unlabeled)



CTA Chest (labeled)

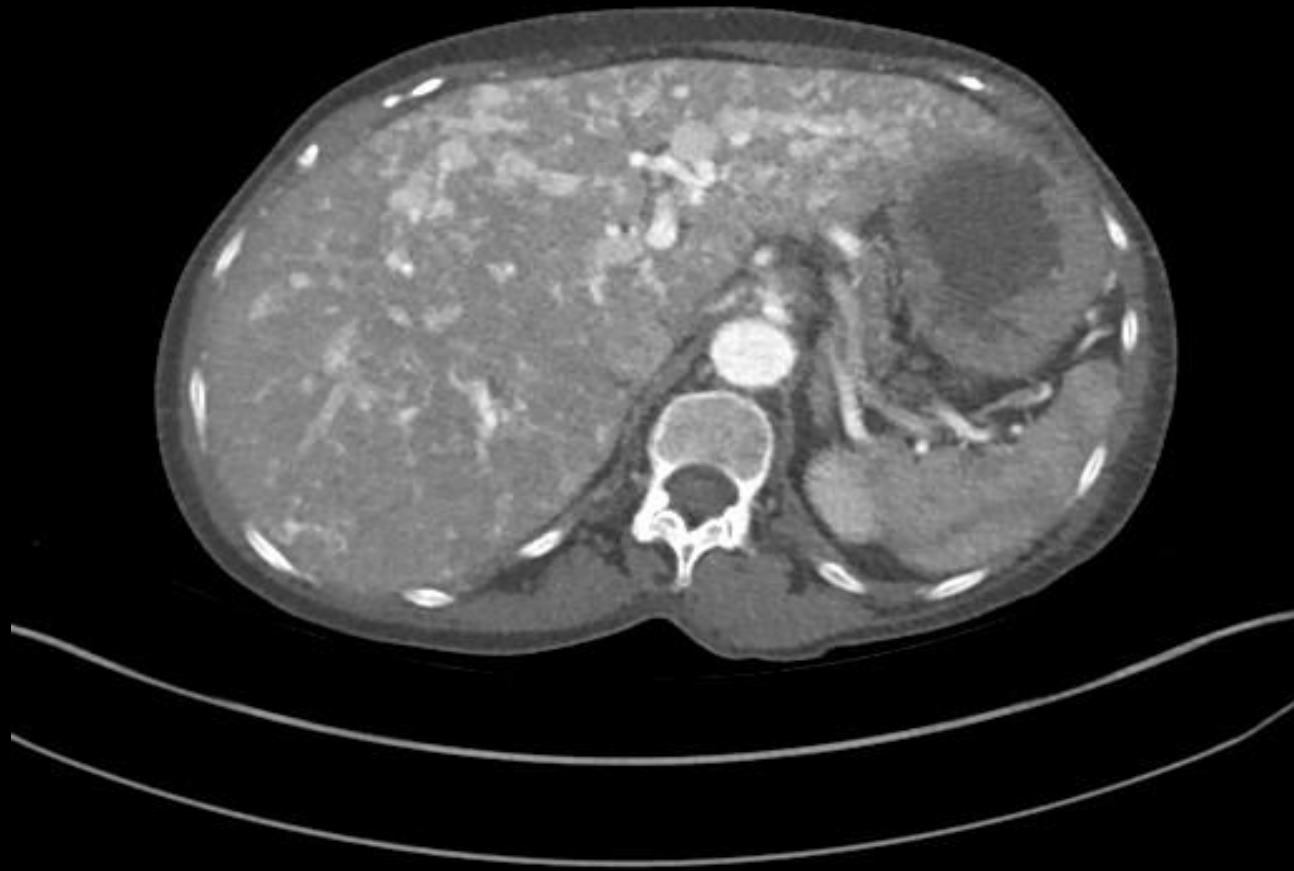
Findings

Numerous bilateral nodular densities (red arrows) in both lungs, the larger of which clearly demonstrate enhancement similar to the pulmonary artery and a few demonstrate a feeding vessel; findings compatible with pulmonary AVMs



Main pulmonary artery is 3.2 cm, can be seen in a setting of pulmonary hypertension.

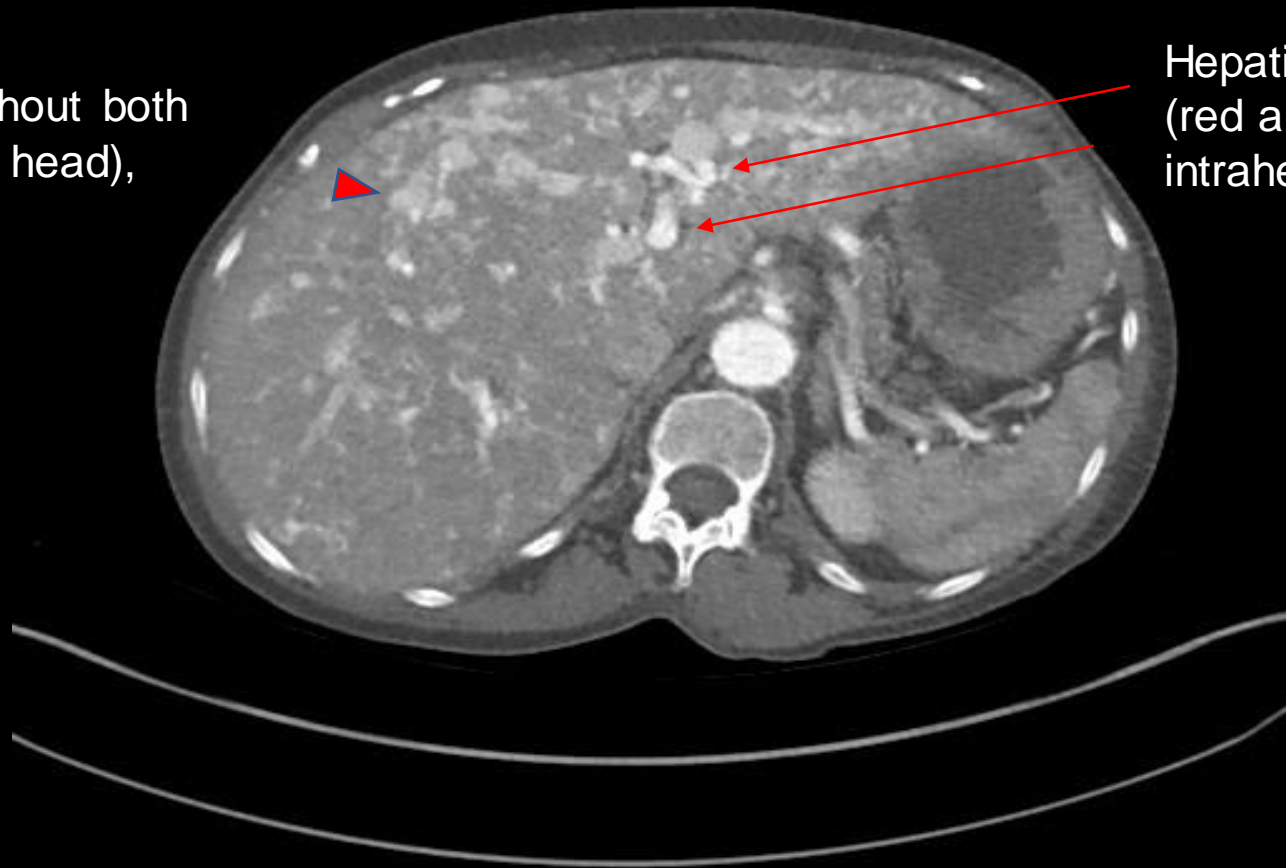
CTA Abdomen/Pelvis – arterial phase (unlabeled)



CTA Abdomen/Pelvis – arterial phase (labeled)

Findings

Extensive AV shunts throughout both lobes of the liver (red arrow head), greater on left.



Hepatic arteries are enlarged (red arrows), related to extensive intrahepatic AV shunts

CTA Abdomen/Pelvis - venous phase (unlabeled)



CTA Abdomen/Pelvis - venous phase (labeled)

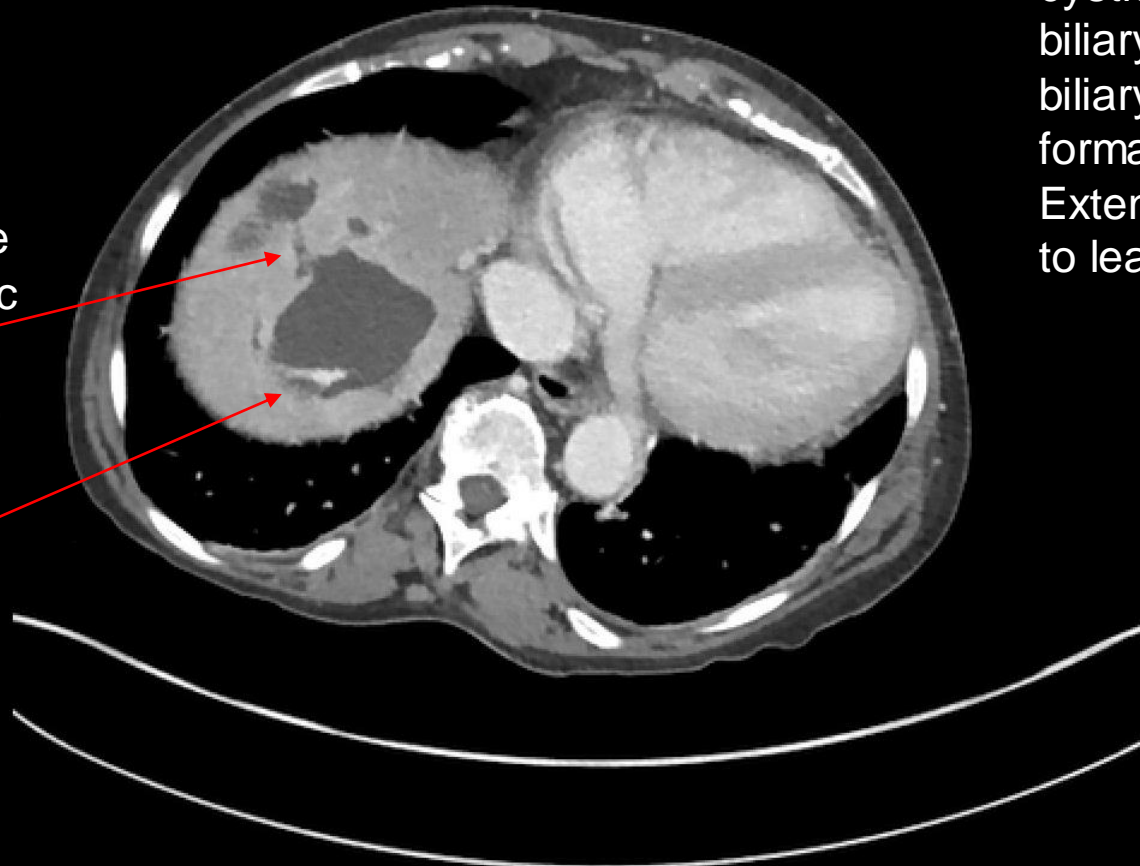
Impression

Large complex right hepatic lobe cystic lesion communicating with biliary system, likely a result of biliary necrosis and bile lake formation.

Extensive AV shunting has shown to lead to biliary necrosis.

In the right hepatic lobe extending to the dome, there is an irregular complex cystic lesion with multiple communicating components.

There is extravasation of contrast in the bile lake, suggesting hemorrhage.



Final Dx:

Hereditary Hemorrhagic Telangiectasia
(Osler-Weber-Rendu Syndrome)

Osler-Weber-Rendu Syndrome

- **Epidemiology:** 1.5 per 100,000 worldwide
- **Clinical Presentation:** classic clinical triad – epistaxis, multiple telangiectasia, and positive family history
- **Pathology:** A rare autosomal dominantly inherited disorder characterized by malformation of blood vessels in the skin (90%), mucous membranes (90%), and organ systems (eg pulmonary (20%), hepatobiliary/GI (40-80%), CNS (5-10%))
- **Diagnosis:** mainly clinical Dx – based 3 of the 4 findings of Curacao criteria. 1) recurrent spontaneous epistaxis 2) multiple mucocutaneous telangiectasia 3) visceral AVMs 4) 1st degree relative with HHT

Osler-Weber-Rendu Syndrome

RADIOGRAPHIC FINDINGS

Pulmonary/GI/Hepatobiliary

CXR – well-circumscribed mass with enlarged draining vein

CT/CTA --vascular mass with enhancing feeding artery and draining vein. Shunting AVMs in the biliary system can cause biliary necrosis, leading to intra or extrahepatic leaks of bile (bile lakes or bilomas)

Brain MRI: cerebral and cerebellar AVMs in superficial locations

MANAGEMENT

- 1) embolization via IR using coils.
- 2) Surgical resection is another option, however, there is a large risk for bleeding.
- 3) Endoscopic ablation/electrocautery for GI bleeding.
- 4) Last resort – liver transplant

Case Discussion

- After finding AVMs in the lungs and liver, a Brain MRI was ordered. Findings showed scattered capillary telangiectasias in the pons and right paramedian cerebellar hemisphere.
- IR, neurosurgery, ENT, and hepatobiliary surgery were consulted. However, IR and neurosurgery could not localize the feeding vessel and were unable to perform embolization
- Hepatobiliary surgery was suspicious for a neoplasm, thus a Liver MRI was ordered. No evidence of neoplasm, and surgery declined to excise due to the large risk of hemorrhage.
- Overall management: Watchful waiting with 3 units of pRBCs per month. ENT recommended nasal sprays without packing or cauterization for ongoing epistaxis. Repeat Liver MRI in 3 months for reassessment.

References

1. Garcia-tsoo G, Korzenik JR, Young L et-al. Liver disease in patients with hereditary hemorrhagic telangiectasia. N. Engl. J. Med. 2000;343 (13): 931-6.
2. Ianora AA, Memeo M, Sabba C et-al. Hereditary hemorrhagic telangiectasia: multi-detector row helical CT assessment of hepatic involvement. Radiology. 2004;230 (1): 250-9.
3. Memeo M, Stabile Ianora AA, Scardapane A, Buonamico P, Sabba C, Angelelli G. Hepatic involvement in hereditary hemorrhagic telangiectasia: CT findings. Abdom Imaging 2004;29:211–220
4. Shovlin CL, Guttmacher AE, Buscarini E, Faughnan ME, Hyland RH, Westermann CJ, Kjeldsen AD, Plauchu H. Diagnostic criteria for hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber syndrome). (2000) American journal of medical genetics. 91 (1): 66-7.