

AMSER Case of the Month

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27F with headache and fever

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Patient Presentation

- 27 year old female with history of polycystic ovarian syndrome and depression presents with frequent headaches for the past 6 weeks that have intensified in the last week, and a fever for the past 3 days. The patient also reports shortness of breath with exertion for the past 4 months and lately cannot walk more than 2 blocks without being short of breath. The headache is intractable, and the fever is persistent. No known sick contacts, no recent travel. Works in a middle school. Unknown family history, no past surgical history.

Pertinent Labs and Physical Exam

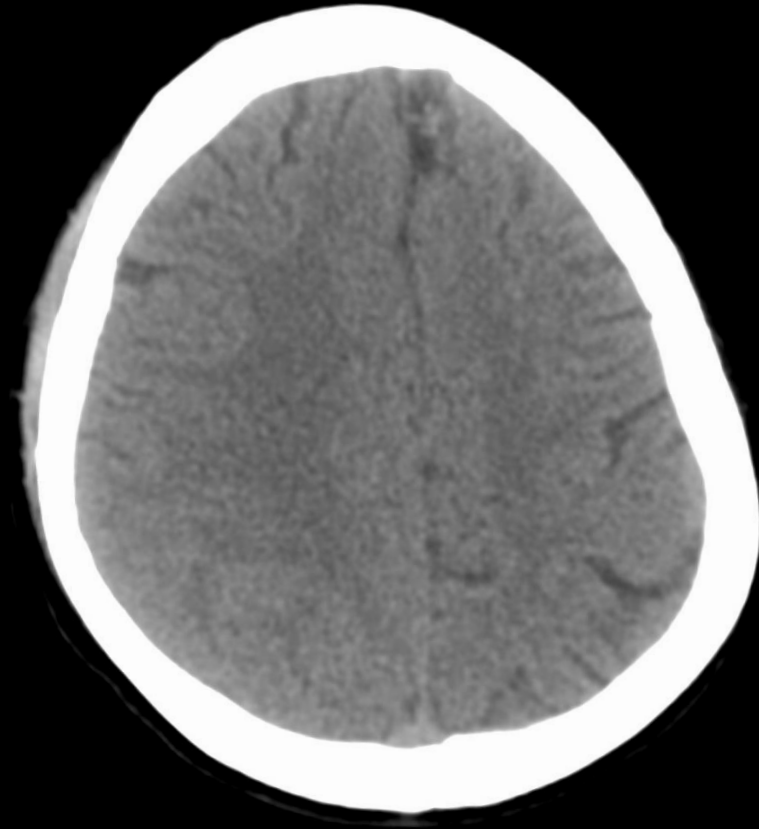
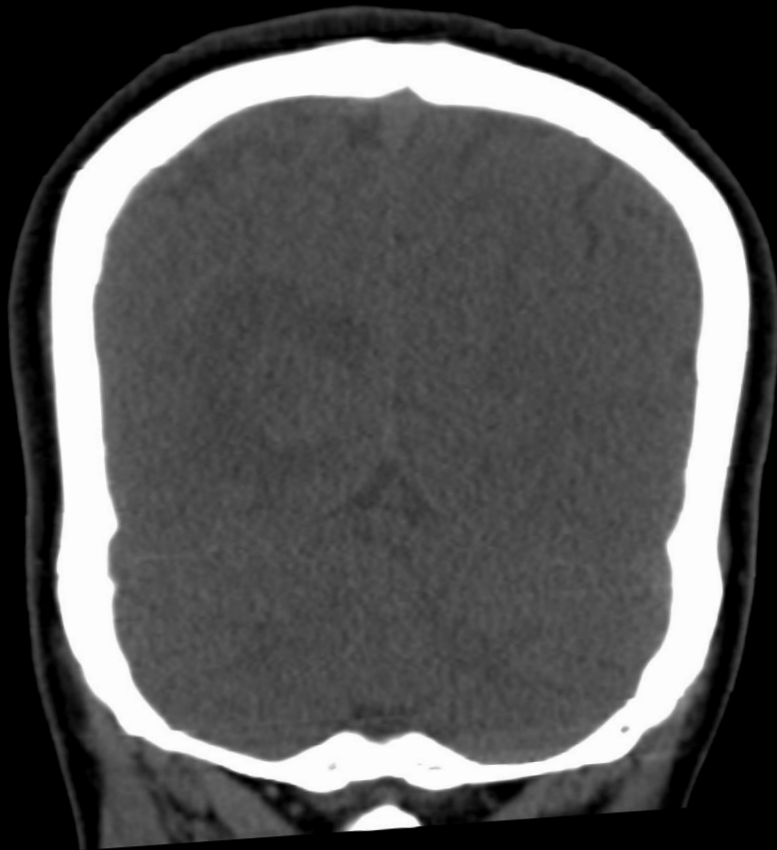
- CBC
 - WBC – 12,200 w/ neutrophil predominance
- Blood culture
 - Gram positive streptococci
- Physical exam findings:
 - Temperature 102.2 F
 - Neurologic Exam – Normal
 - HEENT – Normal
 - Chest - Normal lung sounds. Tachycardic , regular rhythm

What Imaging Should We Order?

ACR Appropriateness Criteria

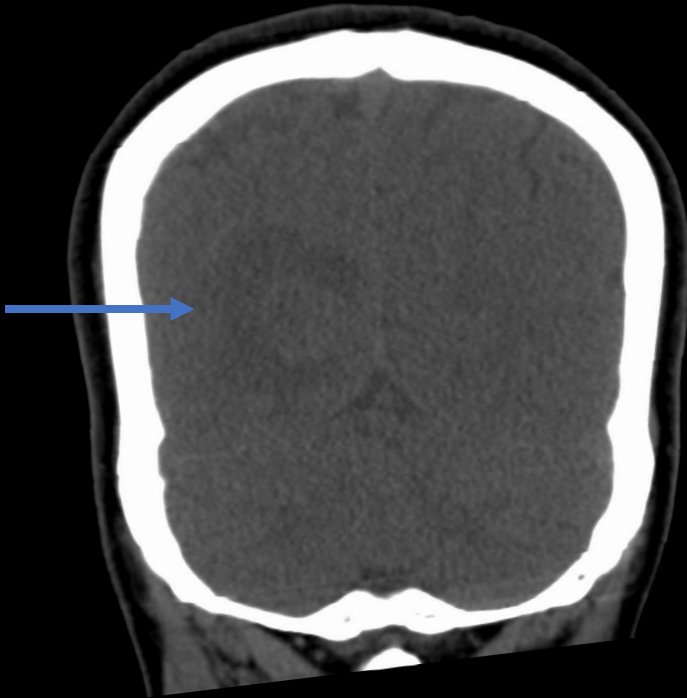
Scenario	Scenario ID	Procedure	Adult RRL	Peds RRL	Appropriateness Category
Headache, fever, initial imaging	3196295	● MRI head without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate
		● MRI head without IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate
		● CT head without IV contrast	1-10 mSv ☼☼☼	0.3-3 mSv [ped] ☼☼☼	Usually appropriate
		● Arteriography cervicocerebral	1-10 mSv ☼☼☼	3-10 mSv [ped] ☼☼☼☼	Usually not appropriate
		● MRA head with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		● MRA head without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		● MRA head without IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		● MRI head with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		● MRV head with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		● MRV head without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		● MRV head without IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		● CT head with IV contrast	1-10 mSv ☼☼☼	0.3-3 mSv [ped] ☼☼☼	Usually not appropriate
		● CT head without and with IV contrast	1-10 mSv ☼☼☼	3-10 mSv [ped] ☼☼☼☼	Usually not appropriate
		● CTA head with IV contrast	1-10 mSv ☼☼☼	3-10 mSv [ped] ☼☼☼☼	Usually not appropriate
		● CTV head with IV contrast	1-10 mSv ☼☼☼	3-10 mSv [ped] ☼☼☼☼	Usually not appropriate

Findings

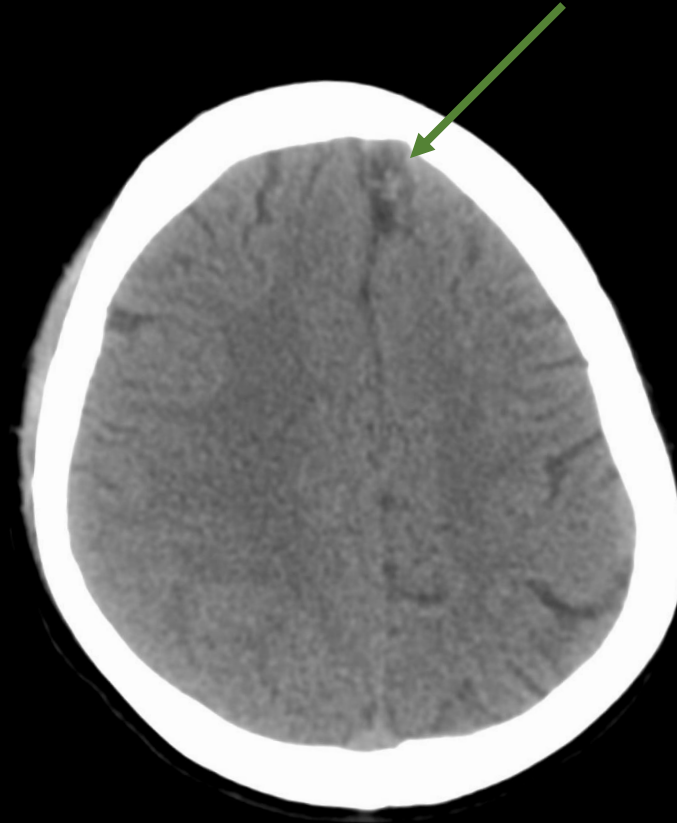


Findings

1.9 cm suspected mass in right occipital lobe medially at gray-white junction with moderate surrounding vasogenic edema.



Possible prominent venous structure anterosuperomedial to the medial left frontal lobe



Final Dx:

Hereditary Hemorrhagic Telangiectasia

Diagnostic Angiograms



Left frontal SM1 AVM, 11.5x10x15 mm. Arterial feeders from the L ACA callosomarginal branch and drainage in the superior sagittal sinus.



Feeding artery to the right upper lobe PAVM measures 7mm. The nidus measures up to 4.4 cm. The outflow vein measures 11 mm.

Case Discussion – Clinical Presentation

- Classic clinical presentation of Hereditary Hemorrhagic Telangiectasia (HHT) is recurrent epistaxis - most common and often earliest symptom, present in >90% of patients [1,4]
- Multiple mucocutaneous telangiectasias - Small, red violaceous lesions with typical sites being lips, oral cavity, fingers, face [1,2]
- **Visceral arteriovenous malformations (AVM)** commonly affect lungs, liver, brain, and gastrointestinal tract. May cause dyspnea, hypoxemia, stroke, GI bleeding, or high-output heart failure [2,4]
- Family history of HHT is common as it has autosomal dominant inheritance [2]
- Patients usually present in childhood/adolescence. Mucocutaneous telangiectasias can appear later in life, often in adolescence or adulthood, while visceral arteriovenous malformations may be present from birth but are often asymptomatic until later in life [2,4]

This patient did not have epistaxis or cutaneous lesions. Her clinical picture warranted brain imaging, and upon visualizing cerebral AVM, it was necessary to screen for additional AVMs. Additional genetic testing was also completed.

Case Discussion – Pathophysiology

Patient was found to have a large pulmonary arteriovenous malformation (PAVM)

Mechanism of brain abscess formation in HHT [3] :

- Right-to-left shunting through pulmonary AVM
- PAVMs create direct connections between pulmonary arteries and veins, bypassing the pulmonary capillary filter
- Loss of filtration allows septic emboli to enter the systemic circulation.

The American College of Radiology recommends percutaneous transcatheter embolization as the primary treatment for PAVMs in HHT [5]

Embolization occludes the feeding artery, eliminating the right-to-left shunt and reducing risk of hypoxemia, septic emboli, and life-threatening hemorrhage [2]

Case Discussion – Management

- PAVM embolized with microcoils with the goal of closing the right to left shunt and reducing risk of downstream sequelae
- Brain abscess initially treated with antibiotics with neurosurgery following, future cerebral AVM resection planned
- The American College of Radiology recommends follow-up CT angiography at 6–12 months post-embolization [5]
- SIR guidelines for routine follow up: bubble echo every 3-5 years to screen for additional PAVMs, annual lab tests to assess for hepatic AVMs, cerebral AVMs are screened at time of diagnosis [6]



PAVM embolized with
0.018" microcoils

References:

1. Li, S., Wang, S. J., & Zhao, Y. Q. (2018). Clinical features and treatment of hereditary hemorrhagic telangiectasia. *Medicine*, 97(31), e11687. <https://doi.org/10.1097/MD.00000000000011687>
2. Hetts, S. W., Shieh, J. T., Ohliger, M. A., & Conrad, M. B. (2021). Hereditary Hemorrhagic Telangiectasia: The Convergence of Genotype, Phenotype, and Imaging in Modern Diagnosis and Management of a Multisystem Disease. *Radiology*, 300(1), 17–30.
3. Roberts, J. I., Woodward, K., Kirton, A., & Esser, M. J. (2022). Pearls & Oysters: Cerebral Abscess Secondary to Pulmonary Arteriovenous Malformation in Hereditary Hemorrhagic Telangiectasia. *Neurology*, 98(7), 292–295.
4. Geisthoff, U. W., Mahnken, A. H., Denzer, U. W., Kemmling, A., Nimsky, C., & Stuck, B. A. (2024). Hereditary Hemorrhagic Telangiectasia (Osler's Disease): Systemic, Interdisciplinary, Relatively Common—and Often Missed. *Deutsches Arzteblatt international*, 121(18), 601–607. <https://doi.org/10.3238/arztebl.m2024.0111>
5. ACR Appropriateness Criteria® Pulmonary Arteriovenous Malformation (PAVM): 2023 Update. Pillai, Anil K. et al. *Journal of the American College of Radiology*, Volume 21, Issue 6, S268 - S285
6. 2009 Treatment Guidelines for Hereditary Hemorrhagic Telangiectasia Trerotola, Scott O. et al. *Journal of Vascular and Interventional Radiology*, Volume 21, Issue 2, 179