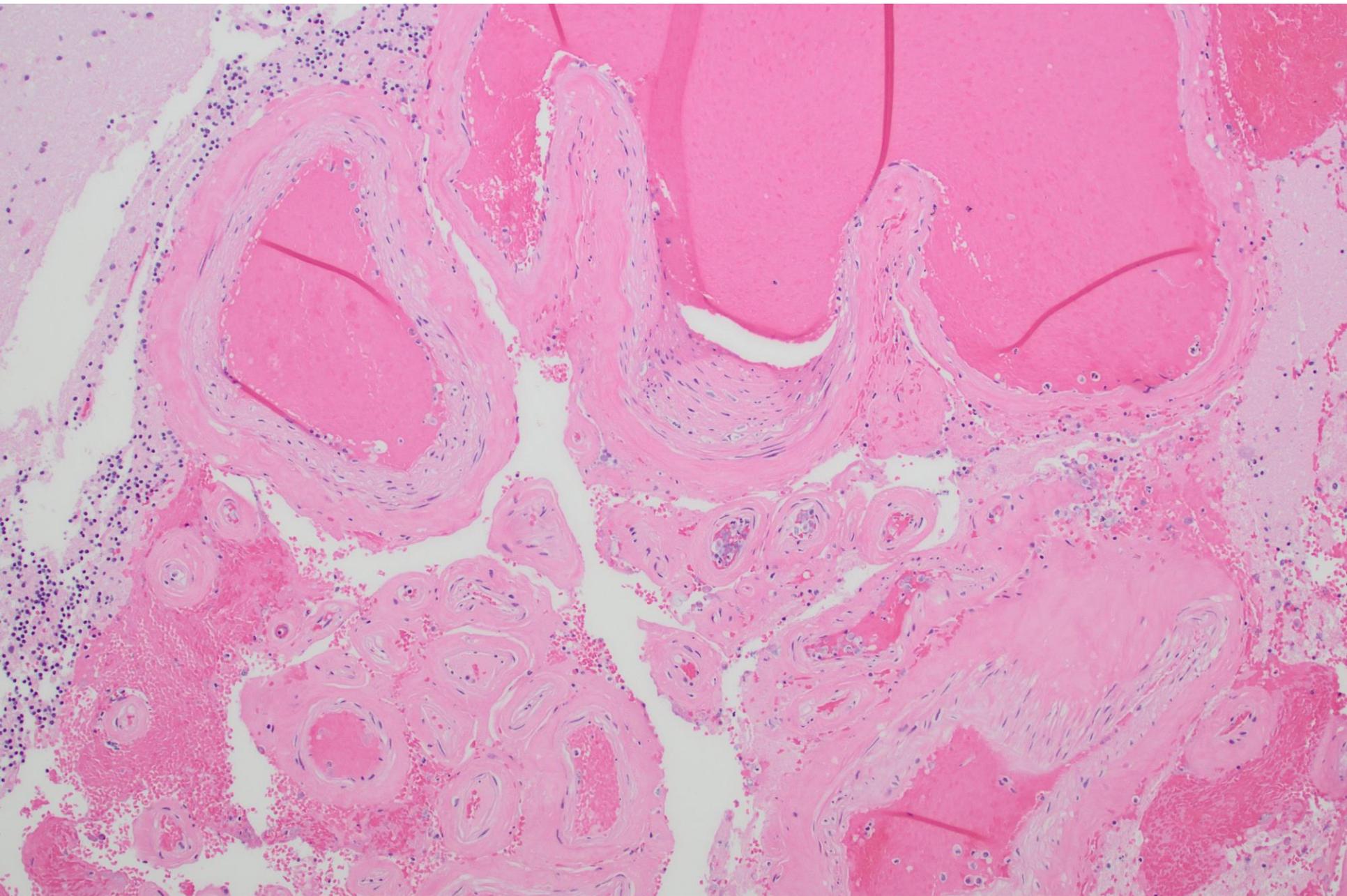


EAC Case #28



Case and images courtesy of Dr. Leo Yenwongfai (PGY2) and Dr. Greg Davis (University of Kentucky)



1. The decedent was a 63-year-old African American male with hypertension and sudden onset severe headache after mowing his lawn. He was rushed to the emergency room and a head CT scan showed cerebellar hemorrhage with obstructive hydrocephalus, transtentorial herniation and cerebellar tonsillar herniation. A lesion located in the cerebellum is shown in the histologic image above.

What is the most likely diagnosis?

- Capillary telangectasia
- Hemangioblastoma
- Arteriovenous malformation
- Glioblastoma
- Intracerebral abscess

A. Capillary telangiectasia (2.7% responses)

These lesions are usually incidental autopsy findings. Microscopically they are composed of small thin-walled vessels with intervening brain parenchyma. Capillary telangiectasias are rarely symptomatic and are almost never hemorrhagic.

B. Hemangioblastoma (4.8% responses)

This neoplasm is associated with von Hippel-Lindau syndrome, and usually seen in patients 20-40 years old. The lesion is most commonly identified in the posterior cranial fossa (cerebellar region) and appears as a well-defined nodule or a cyst with a mural nodule. Microscopically they show a dense cluster of small vascular channels.

C. Arteriovenous malformation (CORRECT ANSWER, 88.89% responses)

Arteriovenous malformations (AVMs) are congenital or acquired vascular malformations arising from arteries draining directly into veins without a capillary bed. AVMs account for 1-2% of all strokes annually. Patients typically present with severe headaches with mass effect. Microscopically, veins are typically more numerous than arteries with abundant hyalinization, calcification and occasional thrombosis. These vessels generally contain thin walls and prominent internal elastic lamina (IEL), or thick walls and no IEL, which are sometimes described as 'arterialized veins. The vascular channels of an AVM are usually, at least partially, embedded within brain parenchyma, but involved brain regions always show reactive changes, including astrocytic gliosis, old hemorrhage, cytooid bodies and even Rosenthal fibers.

This patient had atypical posterior circulation and a conglomerate of anomalous vessels with significant hyalinization and loss of elastic lamina consistent with a vascular malformation. At autopsy, there was extensive ventricular and intraparenchymal hemorrhage of the brainstem with extension into the subarachnoid space (see additional photos).

D. Glioblastoma (1.8% responses)

Glioblastoma multiforme is an aggressive high-grade glioma with predominantly astrocytic maturation that occurs at any age with a short duration of antecedent symptoms and poor prognosis. Imaging studies usually show an enhancing ring lesion. This patient presented with acute CNS symptoms less likely to be a brain tumor. Moreover, CT imaging does not reveal a ring-enhancing lesion.

E. Intracerebral abscess (1.8% responses)

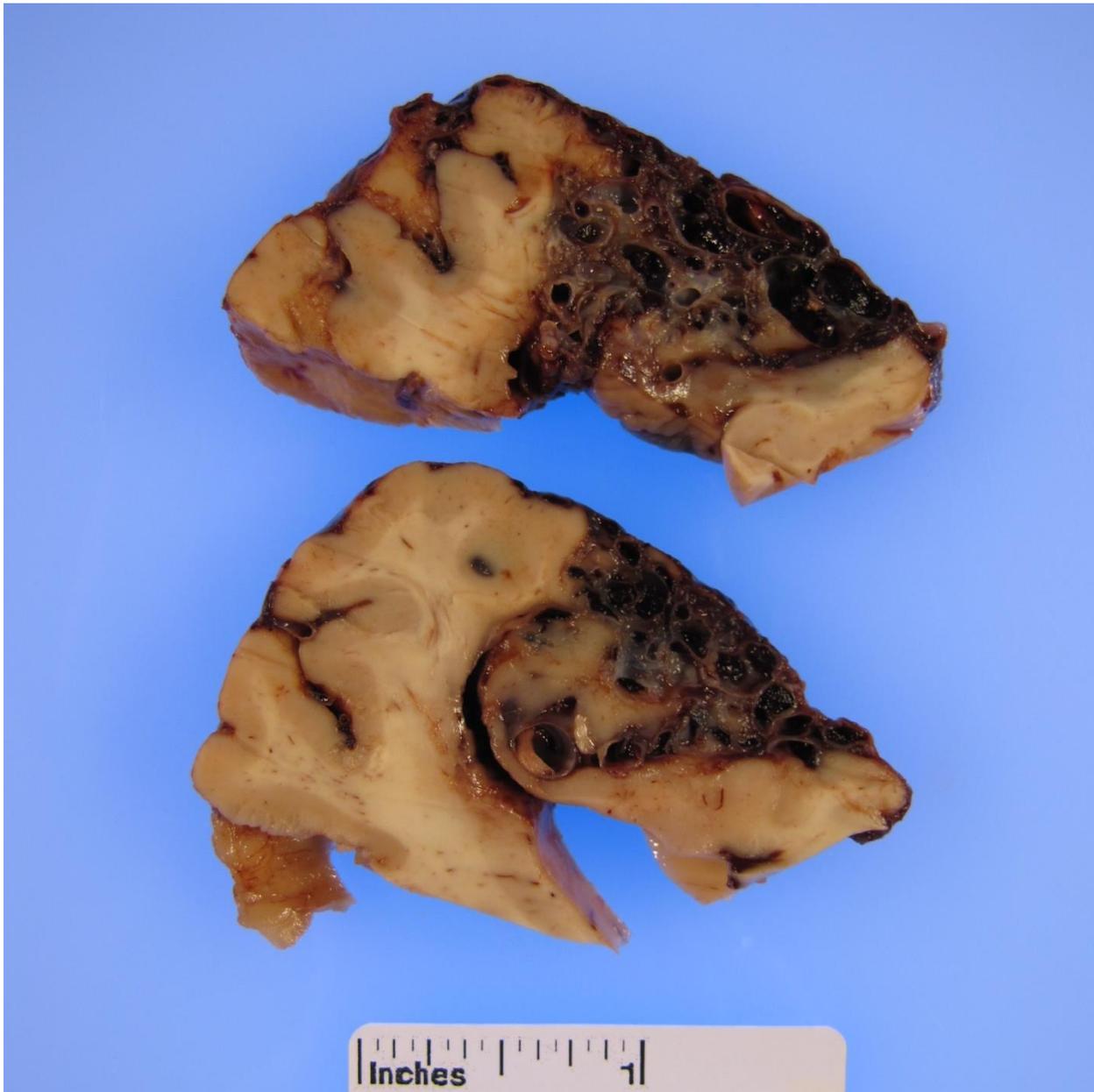
Though a brain abscess can have similar imaging findings as in intracranial hemorrhage and clinical symptoms like headache, the degree of hemorrhage cannot be explained by an abscess alone. Moreover, CNS symptoms will be chronic with most patients having other systemic symptoms such as fever. Our patient had a sudden onset headache with no other systemic symptoms



Basilar subarachnoid hemorrhage
seen at autopsy.



Intraparenchymal hemorrhage of
the brainstem.



Gross example of an AVM (photo provided by Dr. Reade Quinton, Mayo Clinic Rochester)

References:

Batash R, Asna N, Schaffer P, Francis N, Schaffer M. Glioblastoma Multiforme, Diagnosis and Treatment; Recent Literature Review. *Curr Med Chem*. 2017;24(27):3002-3009.

doi:10.2174/0929867324666170516123206

Ellison D. *Neuropathology*. Edinburgh: Mosby; 2013.

Grisold W, Oberndorfer S, Struhal W. Stroke and cancer: a review. *Acta Neurol Scand*. 2009 Jan;119(1):1-16. doi: 10.1111/j.1600-0404.2008.01059.x. PMID: 18616624.

Kim H, Nelson J, Krings T, et al. Hemorrhage rates from brain arteriovenous malformation in patients with hereditary hemorrhagic telangiectasia. *Stroke*. 2015;46(5):1362-1364.

doi:10.1161/STROKEAHA.114.007367

Parizel, Paul M, Makkat, Smitha, Jorens, Philippe G, Özsarlak, Özkan, Cras, Patrick, Van Goethem, Johan W, van den Hauwe, Luc, Verlooy, Jan, & De Schepper, Arthur M. (2002). Brainstem hemorrhage in descending transtentorial herniation (Duret hemorrhage). *Intensive Care Medicine*, 28(1), 85–88.