

AMSER Case of the Month

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63-year-old male with worsening pulsatile tinnitus and
disequilibrium

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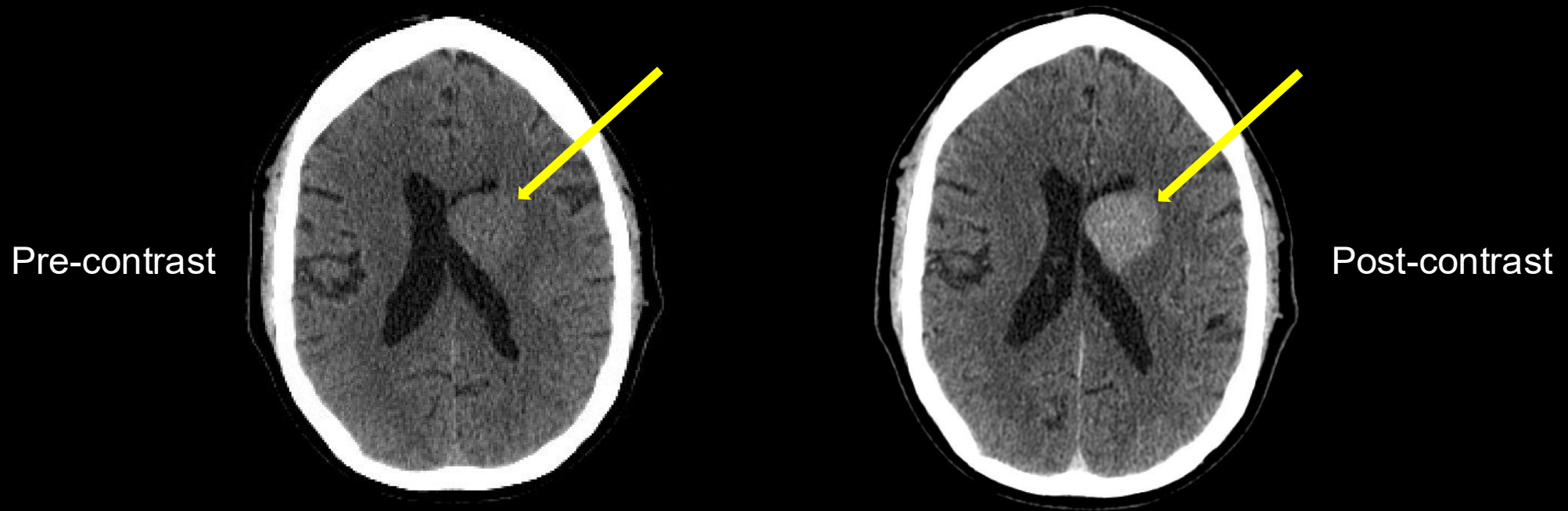
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Patient Presentation

- **HPI:** 63-year-old male with a past medical history of Lynch Syndrome (MSH2 mutation) presents with episodic bilateral pulsatile tinnitus described as “whooshing” over the past 4 years. The episodes are increasing in duration and are associated with feeling off balance. He also endorses mild expressive aphasia and occasional forgetfulness of insidious onset.
- **Exam:** No focal neurologic deficits. Motor strength and sensation intact. Cranial nerves grossly intact.
- **Labs:** CBC, metabolic panel, TSH within normal limits.
- PCP ordered a **CT head** for initial evaluation.

Initial CT Head without and with contrast



- These axial CT images reveal a **mass** centered in the left basal ganglia involving the caudate. It appears **dense** on the pre-contrast CT due to its **hypercellularity**, and **enhances** on the post-contrast image. There is **mass effect** on the left lateral ventricle without hydrocephalus. Findings are **highly suspicious for malignancy**.
- Patient was referred to neurosurgery and neuro oncology for further evaluation and treatment planning.

What Imaging Should We Order?

ACR Appropriateness Criteria

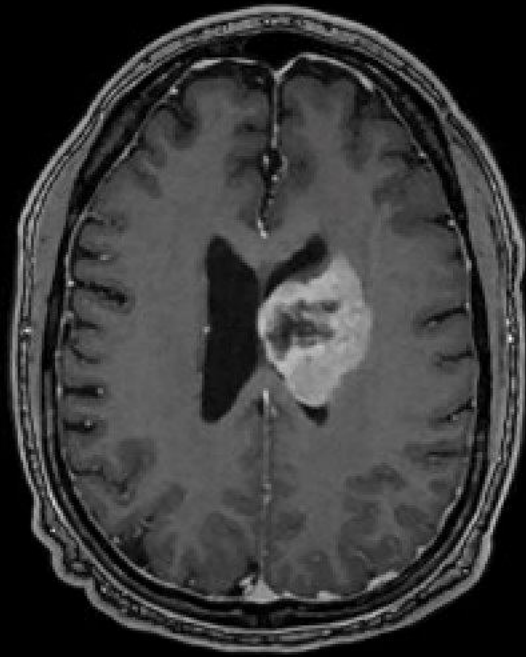
Variant 3: Adult. Suspected intraaxial brain tumor based on prior imaging. Pretreatment evaluation.

Procedure	Appropriateness Category	Relative Radiation Level
MRI head perfusion with IV contrast	Usually Appropriate	O
MRI head without and with IV contrast	Usually Appropriate	O
MR spectroscopy head without IV contrast	May Be Appropriate	O
MRI complete spine without and with IV contrast	May Be Appropriate	O
MRI functional (fMRI) head without IV contrast	May Be Appropriate	O
MRI head perfusion without IV contrast	May Be Appropriate	O
MRI head without IV contrast	May Be Appropriate	O
MRI head without IV contrast with DTI	May Be Appropriate	O
Fluciclovine PET/MRI brain	May Be Appropriate	⚠⚠⚠
Fluciclovine PET/CT brain	May Be Appropriate	⚠⚠⚠⚠
MRI complete spine with IV contrast	Usually Not Appropriate	O
MRI complete spine without IV contrast	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head without and with IV contrast	Usually Not Appropriate	⚠⚠⚠
CT head without IV contrast	Usually Not Appropriate	⚠⚠⚠
DOTATATE PET/CT brain	Usually Not Appropriate	⚠⚠⚠
DOTATATE PET/MRI brain	Usually Not Appropriate	⚠⚠⚠
FDG-PET/CT brain	Usually Not Appropriate	⚠⚠⚠
FDG-PET/MRI brain	Usually Not Appropriate	⚠⚠⚠

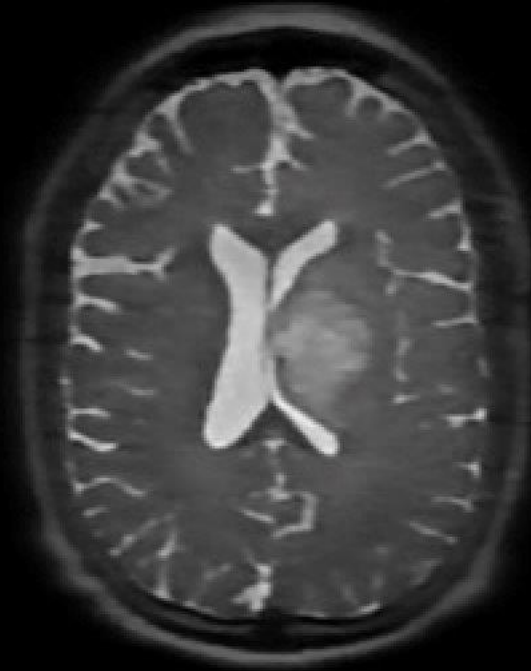
This imaging modality was ordered by the neurosurgeon



Findings (unlabeled)

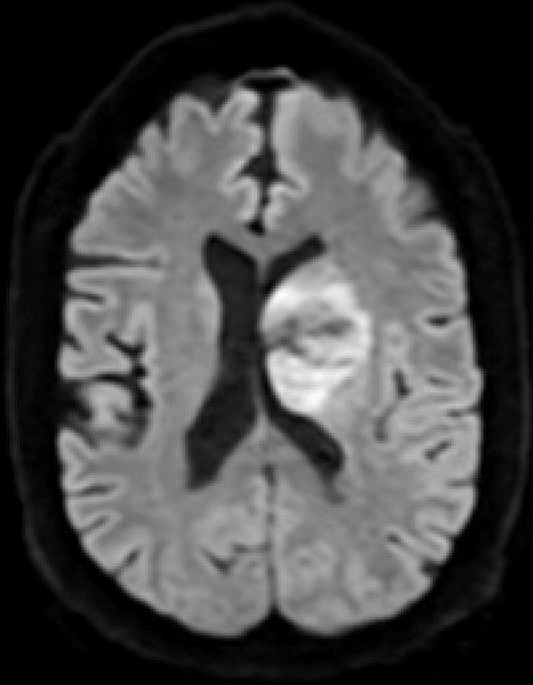


Axial T1 w/
contrast

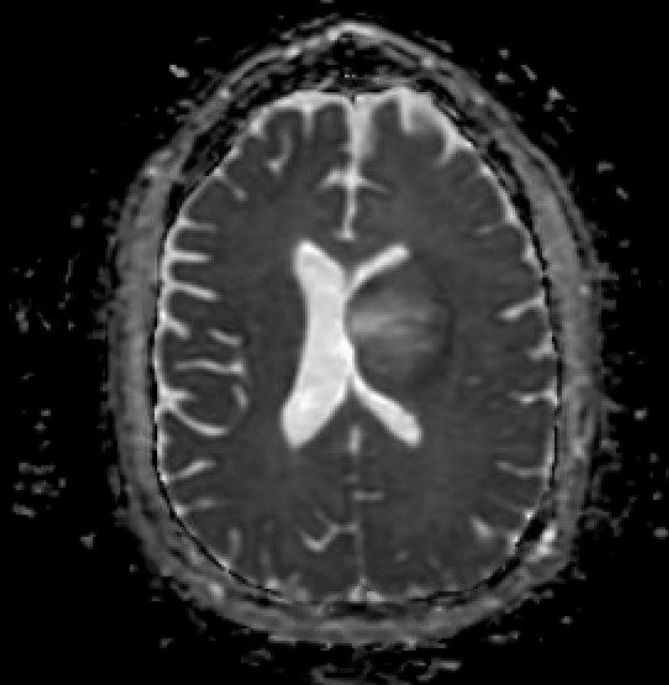


Axial T2

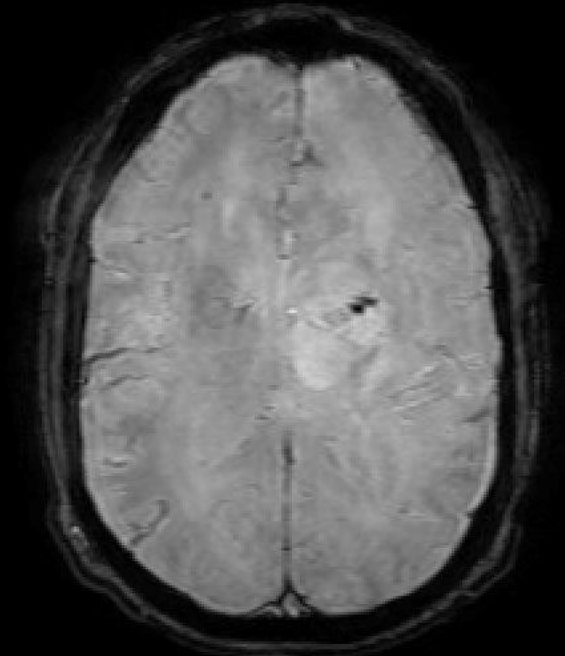
Findings (unlabeled)



DWI

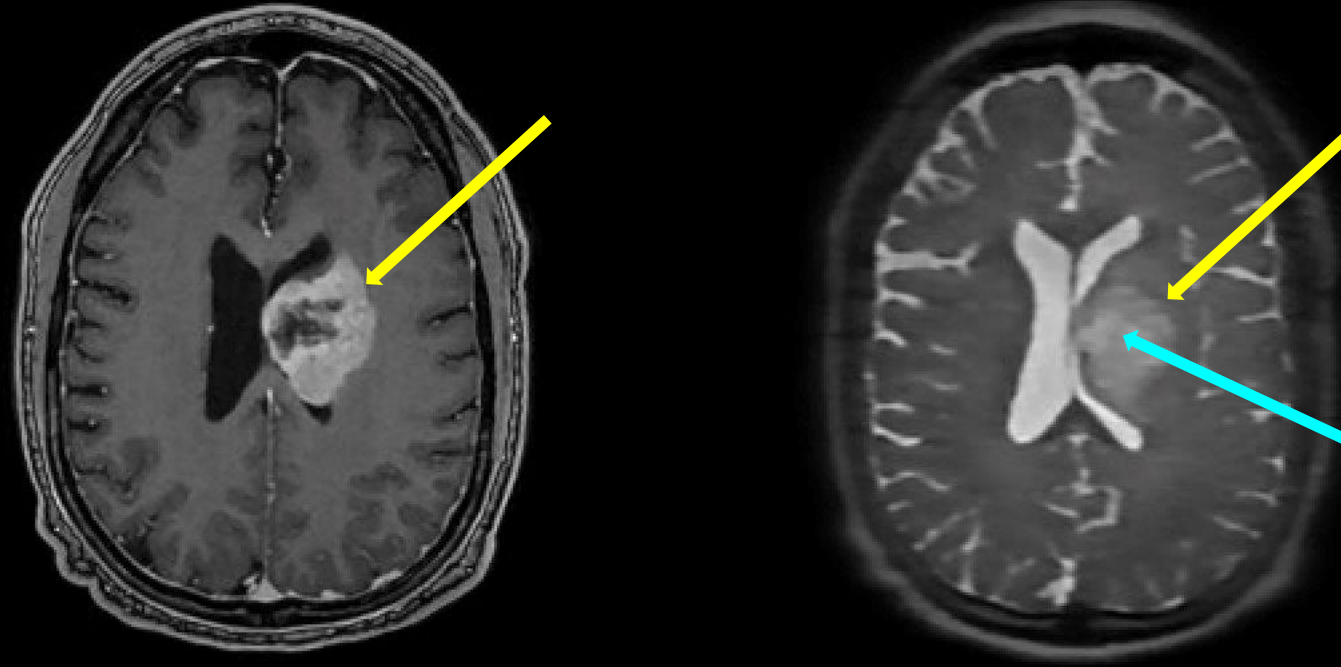


ADC



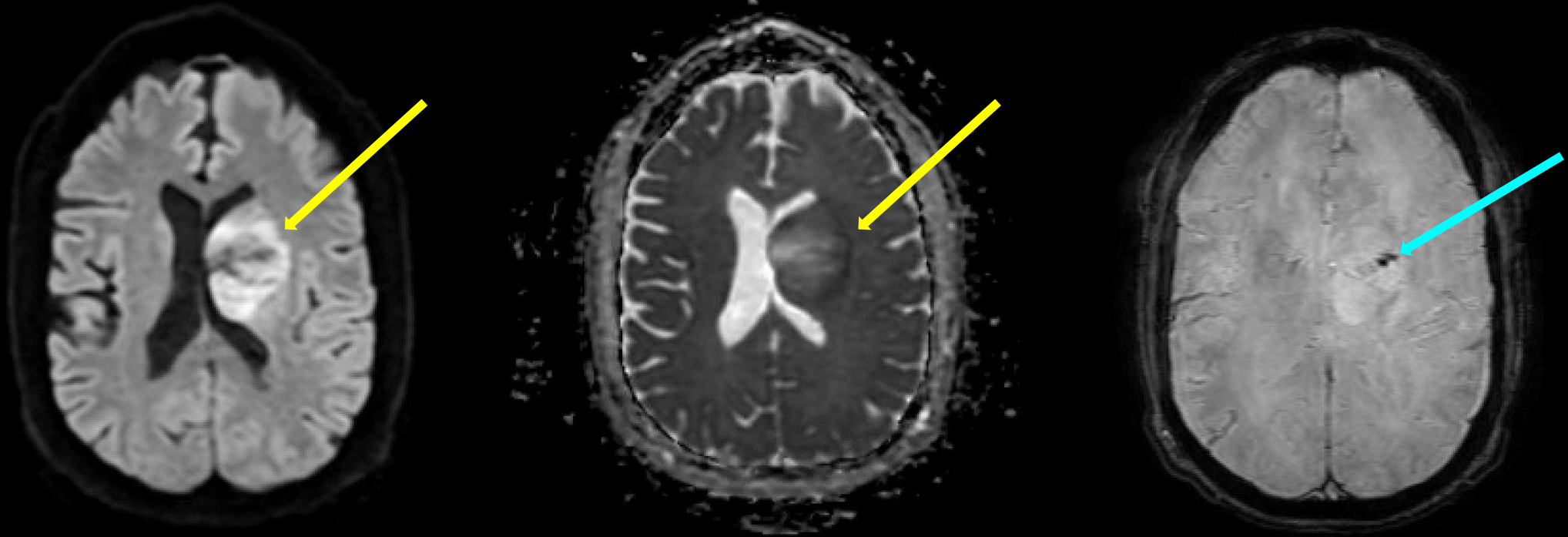
SWI

Findings (labeled)



- These axial CT images redemonstrate a left periventricular gangliocapsular mass that is **T1 hypointense, T2 hyperintense, with contrast enhancement**, measuring 4.7 x 3.4 cm. Associated **mass effect** on the left lateral ventricle without midline shift or herniation (yellow arrows).
- The areas of relative T2 hyperintensity represent **internal necrosis** (blue arrow).

Findings (labeled)



- The mass shows **diffusion restriction** (yellow arrows) due to hypercellularity (confirmed on ADC) and trace **foci of susceptibility** representing **microhemorrhages** (blue arrow).

Differential Diagnosis

- Primary CNS lymphoma
- Glioblastoma
- Low grade astrocytoma, IDH-mutant
- Cerebral metastasis
- Cerebral abscess

Stereotactic Biopsy Pathology Findings

- High grade glioma with a variety of histopathologic patterns including pleomorphic astrocytic, myxoid, anaplastic small cell, multinucleated giant cell, and epithelioid.
- Mitoses are numerous, microvascular proliferation is multifocal, and there are patches of necrosis with pseudopalisading.
- IDH-1 stain is negative, indicating IDH wildtype. A Ki-67 preparation labels about 75% of neoplastic nuclei.
- **Interpretation:** high grade glioma with histologic features of glioblastoma, WHO grade 4.

Final Dx:

Glioblastoma (GBM)

Case Discussion

- **Epidemiology**

- GBM is the most common primary brain cancer in adults, with an age-adjusted incidence rate of 3.2 per 100,000. [1]

- **Risk Factors**

- Ionizing radiation, environmental exposures (pesticides, smoking, etc.), and genetic syndromes including neurofibromatosis, tuberous sclerosis, Li-Fraumeni syndrome, retinoblastoma, Turcot syndrome put patients at increased risk of GBM. [2]
- While Lynch Syndrome is not a proven risk factor for GBM, the incidence in this population is up to 7.8%. [3]
 - In those with the MSH2 gene mutation, such as this patient, there is a 3- to 5-fold increased risk of brain tumors compared to other mutations. [3]

Case Discussion

- **Clinical Presentation**

- Patients often present with symptoms of increased intracranial pressure: headache, nausea, vomiting, seizure, focal/progressive neurologic deficits correlating to the involved area of the brain. [4]

- **Diagnosis: Neuroimaging & Histopathologic Correlation**

- On imaging, GBM appears as an irregularly shaped heterogeneous, ring-enhancing lesion with central necrosis and surrounding vasogenic edema, which may cause mass effect. [4]
 - Necrosis is required for a brain tumor to be WHO grade IV or classified as GBM.
- Diagnosis is confirmed with histopathologic examination of biopsied tissue.
 - IDH wildtype on immunohistochemistry can help differentiate GBM from IDH-mutant astrocytoma. [5]

Case Discussion

- **Management & Prognosis**

- Standard treatment involves surgical resection, radiation, & chemotherapy. [4]
 - Steroids can be used to alleviate neurologic symptoms caused by peritumoral edema. [2]
 - Thromboembolic events are a common complication; a low threshold for investigation and treatment should be maintained. [2]
 - Despite treatment, median survival time is 15 months; palliative care should be initiated at time of diagnosis. [4]

- **Key Takeaways**

- The presenting symptoms of GBM are variable & depend on brain regions involved.
- While it may not improve survival, early diagnosis and treatment is important to facilitate symptomatic treatment and complication management.
- Multidisciplinary coordination between radiology, neurosurgery, neuro oncology, palliative care is essential.

References:

1. Ostrom QT, et al. CBTRUS statistical report: primary brain and central nervous system tumors diagnosed in the United States in 2008-2012. *Neuro-oncology*. 2015;17(suppl_4):iv1-iv62.
2. Omuro A, DeAngelis LM. Glioblastoma and other malignant gliomas: a clinical review. *JAMA*. 2013;310(17):1842-1850. doi:10.1001/jama.2013.280319
3. Therkildsen C, et al. Glioblastomas, astrocytomas and oligodendrogliomas linked to Lynch syndrome. *European journal of neurology*. 2015;22(4):717-724.
4. Alexander BM, Cloughesy TF. Adult Glioblastoma. *J Clin Oncol*. 2017;35(21):2402-2409. doi:10.1200/JCO.2017.73.0119
5. Reuss DE, et al. Adult IDH wild type astrocytomas biologically and clinically resolve into other tumor entities. *Acta neuropathologica*. 2015;130(3):407-417.