

CLINICAL VIGNETTE

Cerebellar Tumor

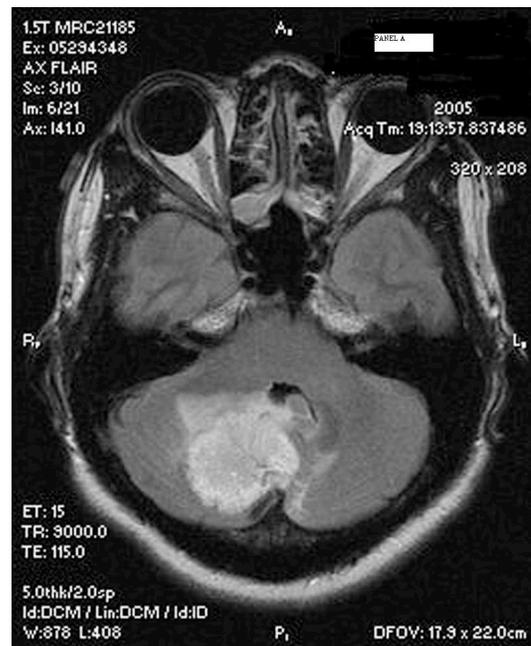
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A 38- year- old female presented to the emergency room with headache and dizziness of 10 days duration. The headache had increased in intensity and frequency and was usually frontal, and at times generalized. On further questioning, she had similar headaches off and on for the past year following a humanitarian mission in Africa but dismissed it as due to the heat and intensity of the work. The headache improved with rest and lying down, rated 8/10 at its worst and 1 -2 /10 at its best. It worsened with defecation or deep breathing. She also reported uncharacteristic fatigue with need to rest at lunchtime. There was no associated nausea or vomiting and screening laboratory tests were normal. A computerized tomography scan of the head, chest, abdomen and pelvis was done then a Magnetic Resonance Imaging (MRI) of the head (Panel A) was done and revealed an intracranial mass.

She was referred to neurosurgery and was noted to have elevated intracranial pressure prompting urgent surgical resection.

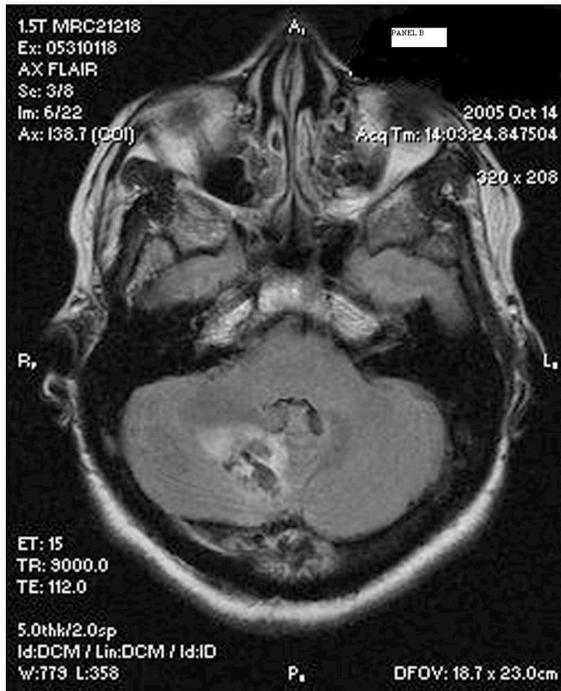
Pathology reported highly vascular tumor.

Panel A:



MRI of the brain showing, large, posterior fossa, right cerebellar, intensely enhancing mass with well defined borders and mild mass effect. Measures 4.3 x 2.1 x 3.8 cm

Panel B:



MRI of the brain, following surgical resection. Lesion is completely gone but slight residual edema still present.

Questions:

Based on the history and physical, which of the following is the most likely diagnosis?

- A. Glioblastoma multiforme
- B. Hemangioblastoma
- C. Cerebellar astrocytomas
- D. Metastasis from the breast
- E. Oligodendroglioma

The answer is B: Hemangioblastoma.

Discussion:

Hemangioblastomas are histologically benign lesions that comprise 1-2 % of primary nervous system tumors and 8-12 % of all posterior fossa lesions in the adult. They commonly occur in young adults with a peak incidence corresponding to the fourth decade of life.¹

Hemangioblastomas, with heterogenous enhancement are benign tumors of uncertain origin that are located predominantly in the posterior cranial fossa and the spinal cord. Although most hemangioblastomas are sporadic, they are sometimes associated with autosomally dominant von Hippel-Lindau (VHL) disease in approximately 25 % of cases. Most hemangioblastomas can be cured with surgical resection, and long-term recurrence rates seem to depend on the presence of VHL disease and multicentric lesions.

Discussion of incorrect answers:

Glioblastoma multiforme is extremely rare in the cerebellum and on imaging has heterogenous enhancement with ill-defined borders. Cerebellar astrocytomas are usually found in children and are heterogeneously enhancing with ill-defined borders on

imaging as well. Metastasis on the other hand, usually has single or multiple lesions, well-defined borders and moderate surrounding edema. Oligodendrogliomas have seizure as their most common presenting symptom and are almost never found in the cerebellum.

Differential Diagnosis Table

Photo Quiz
Author

Selected Differential Diagnosis of Hemangioblastoma

<i>Condition</i>	<i>Characteristics</i>
Cerebellar Astrocytoma	Mainly in children; Heterogeneous enhancement and ill defined borders on imaging
Glioblastoma Multiforme	Exceedingly rare in cerebellum; Heterogenous enhancement and ill defined borders on imaging
Metastasis	Single or multiple lesions; usually well defined borders; surrounding edema
Oligodendroglioma	Seizure is most common symptom; Almost never found in the cerebellum

REFERENCES

1. Gläscher S, Berlis A, Pagenstecher A, Vougioukas VI, Van Velthoven V. Characterization of hemangioblastomas of spinal nerves. Neurosurgery. 2005 Mar;56(3):503-9; discussion 503-9. Review. PubMed PMID: 15730575.

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