AMSER Case of the Month October 2024

37-year-old female with progressively worsening headache, nausea, blurry vision, and ataxia

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Patient Presentation

- HPI: 37-year-old female presenting with a 12-month history of progressively worsening headaches, nausea, vomiting, blurry vision, and ataxia. Patient reports she now dry heaves 1-2 per day and occassionally misses steps while going downstairs. Patient describes blurred and double vision in all quadrants.
- Past Medical History: Bipolar disorder
- Medications: Fluoxetine, Aripiprazole



Patient Presentation

- Vital Signs: BP 134/90, Pulse 60, Temp 98.7F, Resp 16, SpO2 98%
- Physical Exam: No acute distress, AOx4. Normal to light-touch and propioception in all extremities. Mild left-sided pass-pointing and leftsided nystagmus. PERRL. EOMI. Visual fields intact to confrontation. Normal facial sensation in V1-V3. No dysarthria. No focal weakness. 5/5 strength in all extremities. Wide-based gait.



What Imaging Should We Order?



Select the applicable ACR Appropriateness Criteria

Variant 7:

Headache with one or more of the following "red flags": increasing frequency or severity, fever or neurologic deficit, history of cancer or immunocompromise, older age (>50 years) of onset, or posttraumatic onset. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	0
MRI head without IV contrast	Usually Appropriate	0
CT head without IV contrast	Usually Appropriate	€€€
Arteriography cervicocerebral	Usually Not Appropriate	€€€
MRA head with IV contrast	Usually Not Appropriate	0
MRA head without and with IV contrast	Usually Not Appropriate	0
MRA head without IV contrast	Usually Not Appropriate	0
MRI head with IV contrast	Usually Not Appropriate	0
MRV head with IV contrast	Usually Not Appropriate	0
MRV head without and with IV contrast	Usually Not Appropriate	0
MRV head without IV contrast	Usually Not Appropriate	0
CT head with IV contrast	Usually Not Appropriate	€€€
CT head without and with IV contrast	Usually Not Appropriate	€€€
CTA head with IV contrast	Usually Not Appropriate	***
CTV head with IV contrast	Usually Not Appropriate	***





Findings (unlabeled)



Post-Contrast T1 Axial

Post-Contrast T1 Sagittal

T2 FLAIR Axial



Partially solid (enhancing), partially cystic mass within the left cerebellar hemisphere with surrounding vasogenic edema and mass effect on the 4th

Findings (labeled)



Post-Contrast T1 Axial

Post-Contrast T1 Sagittal

T2 FLAIR Axial



Final Dx:

Hemangioblastoma



Hemangioblastoma

- Definition: Benign, highly vascularized neoplasm of the brain, spinal cord, or retina
- Etiology:
 - Sporadic
 - ~20-25% occur in association with Von Hippel-Lindau disease (VHL)
 - ~45% of those with VHL develop hemangioblastomas

• Clinical Features:

- Cerebellum most common site → ataxia, dysmetria, nystagmus, and other cerebellar defects
- Often compresses the 4th ventricle → non-communicating hydrocephalus → features of intracranial hypertension (i.e. headache, papilledema)
- Paraneoplastic EPO production \rightarrow secondary polycthemia



Hemangioblastoma

• Imaging Findings:

- Sharply demarcated homogenous mass composed of a cystic component (60% of cases) and a vividly enhancing mural nodule with prominent serpentine flow voids
 - CT and MRI are the preferred imaging modalities
- CT
 - Mural nodule is isodense to the brain on non-contrast CT with a fluid density surrounding the cyst
 - Post-contrast demonstrates homogenous enhancement of the mural nodule
 - Cyst walls usually do not enhance
- MRI
 - T1: hypointense to isointense mural nodule with CSF signal of the cystic component
 - T2: hyperintense mural nodule with hyperintense fluid-filled cyst
 - Flow voids due to enlarged vessels may be evident, especially at the periphery of the cyst (60% of cases)



Hemangioblastoma

• Treatment:

- Surgical resection is often curative
 - Recurrence <20% in sporadic cases
 - Adjuvant radiotherapy may be used in cases of incomplete resection
- Cystic drainage is insufficient for definitive management



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