Juvenile Pilocytic Astrocytoma

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RAD 3030

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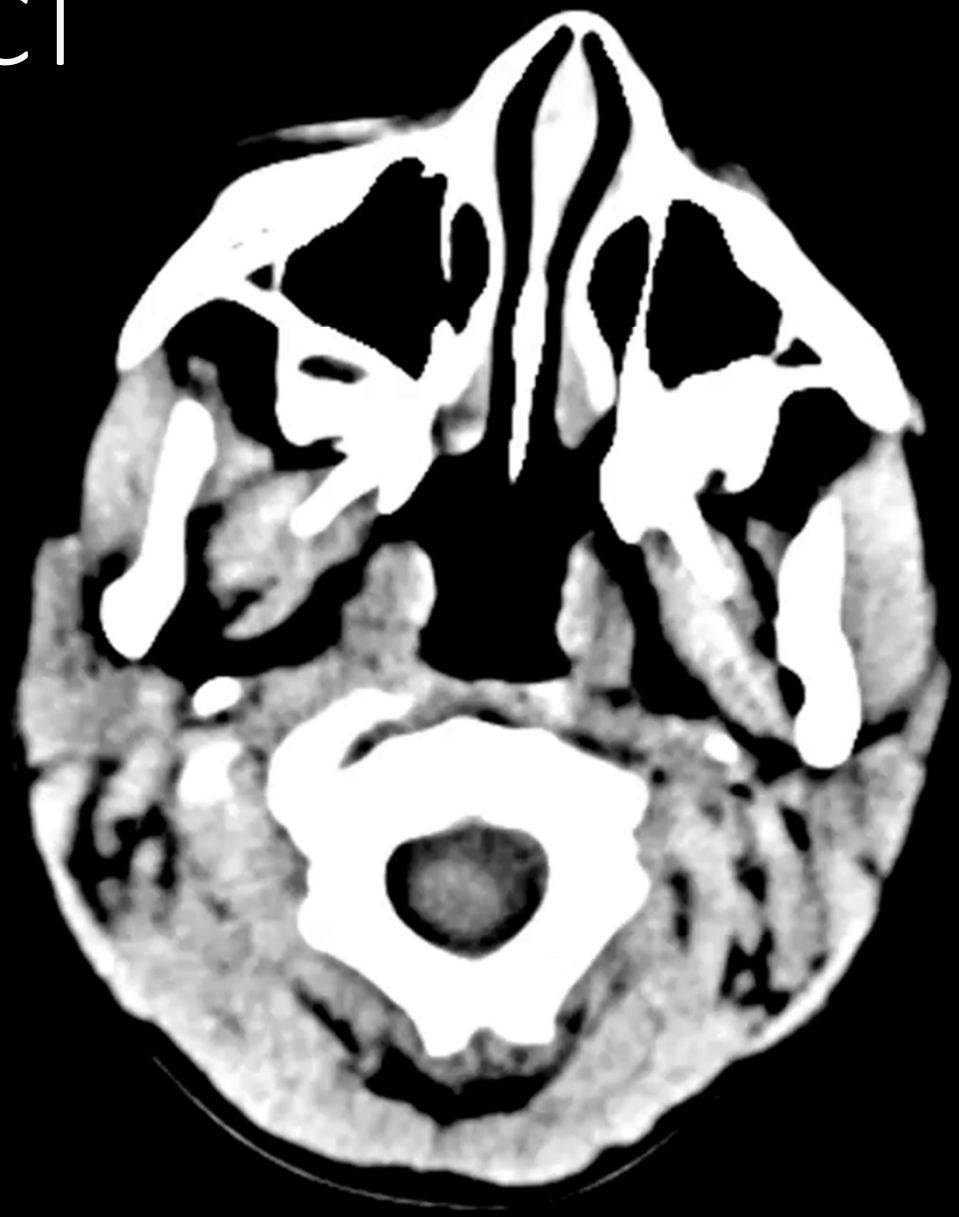
J.M.

- 5 yo M with no relevant past medical history sent to ED from ortho clinic on 7/30/2019 for ataxia
  - Ataxia x 7 weeks
  - Headache x 1 week
  - Vomiting x 2 days
- ED physical exam
  - Ataxic gait
  - Abnormal Romberg
  - Vertical nystagmus
- Sent for emergent CT

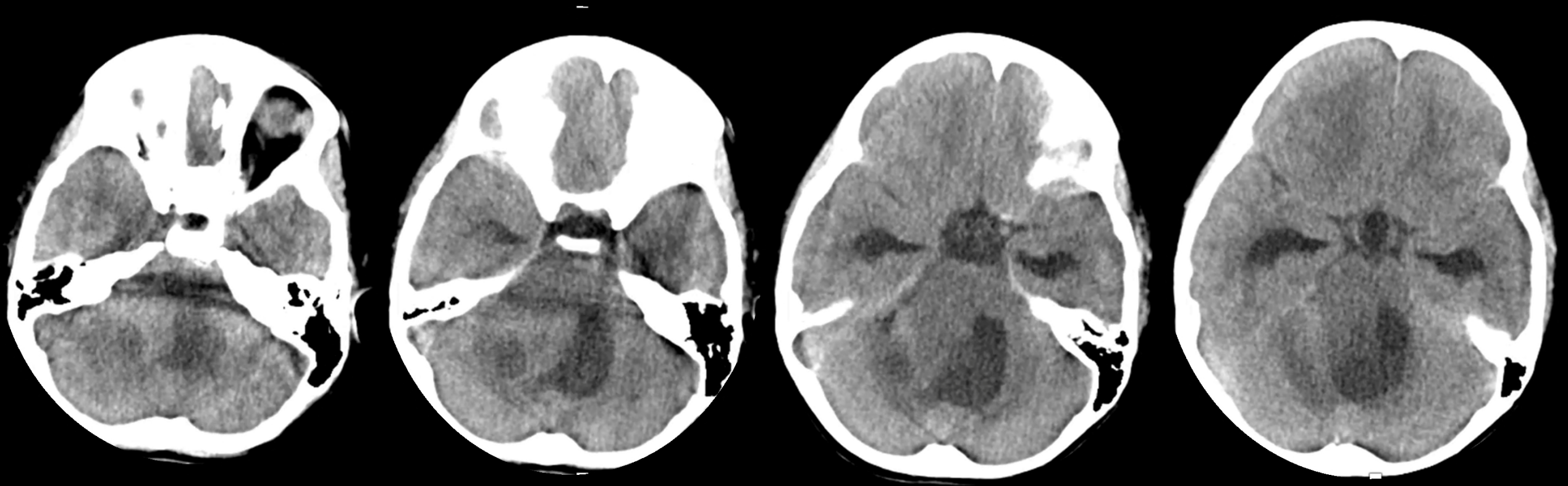
Non-contrast CT

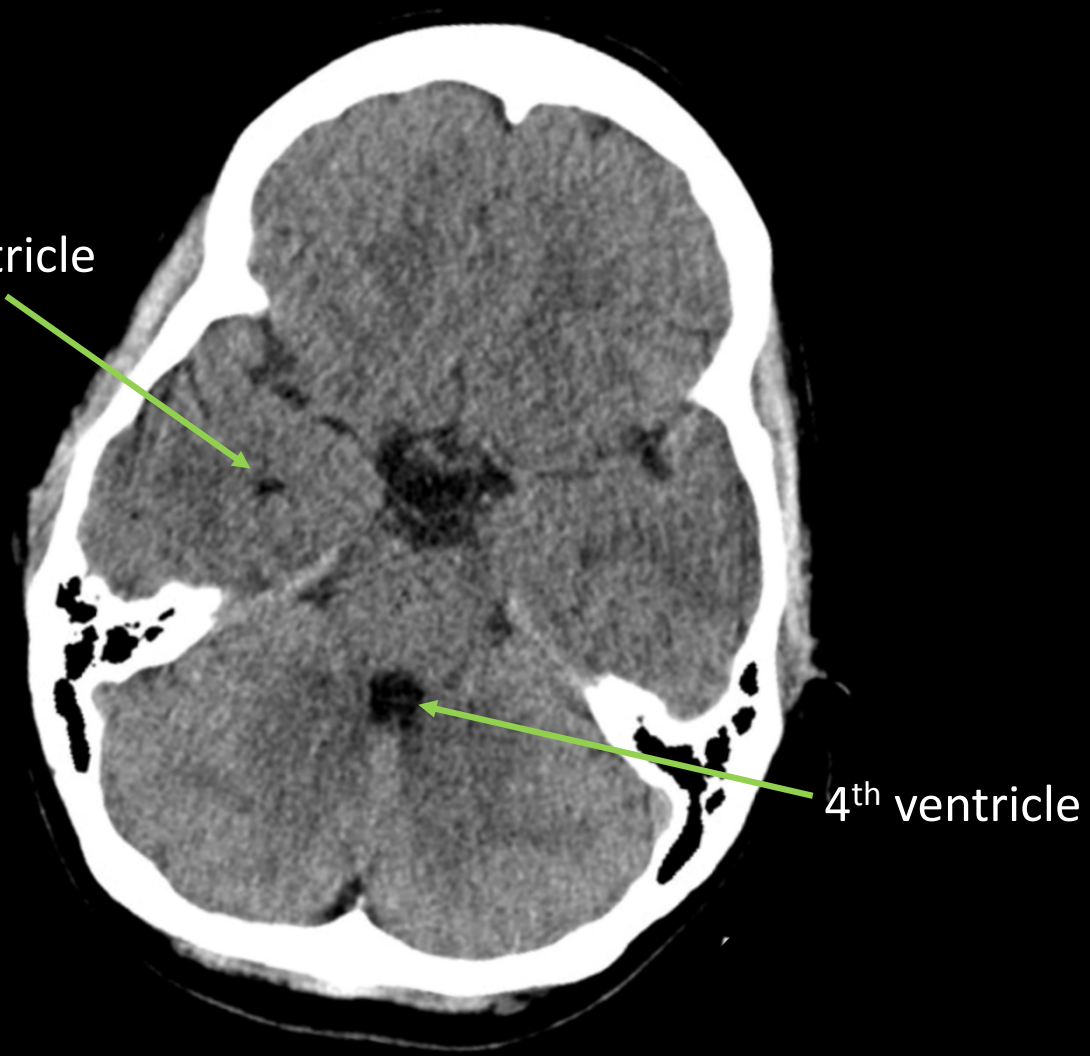
7/30/2019



Non-contrast CT

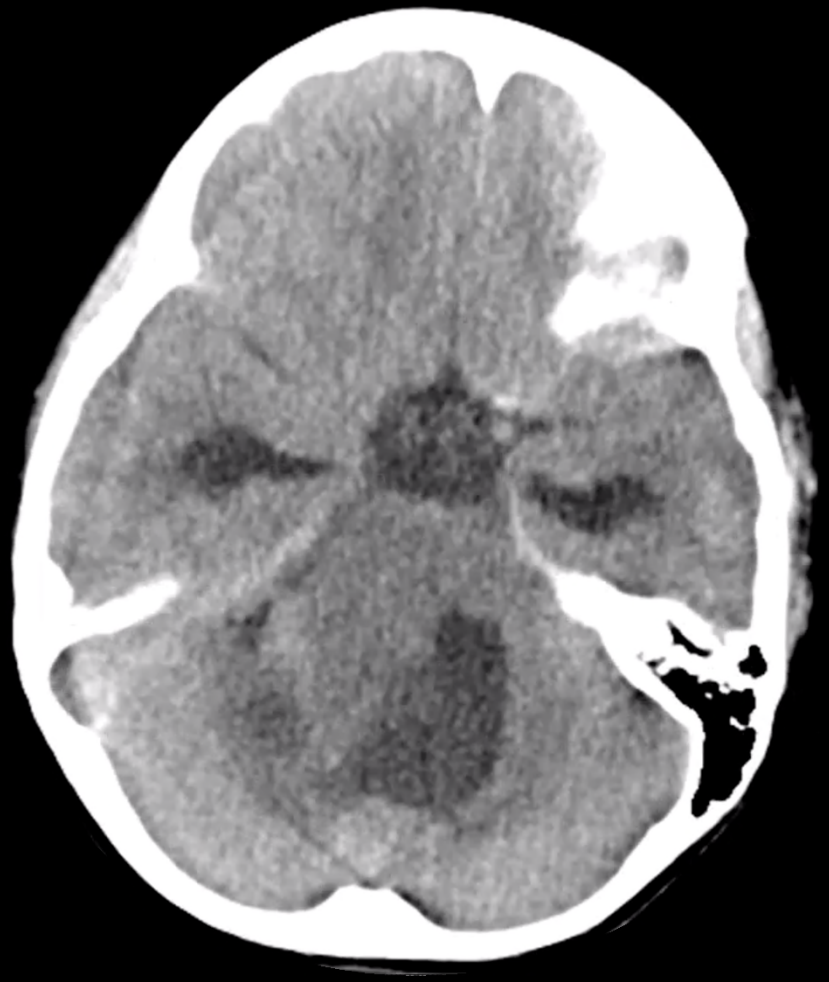
7/30/2019





Normal Anatomy

Case courtesy of Dr Craig Hacking, Radiopaedia.org, rID: 62252



J.M.

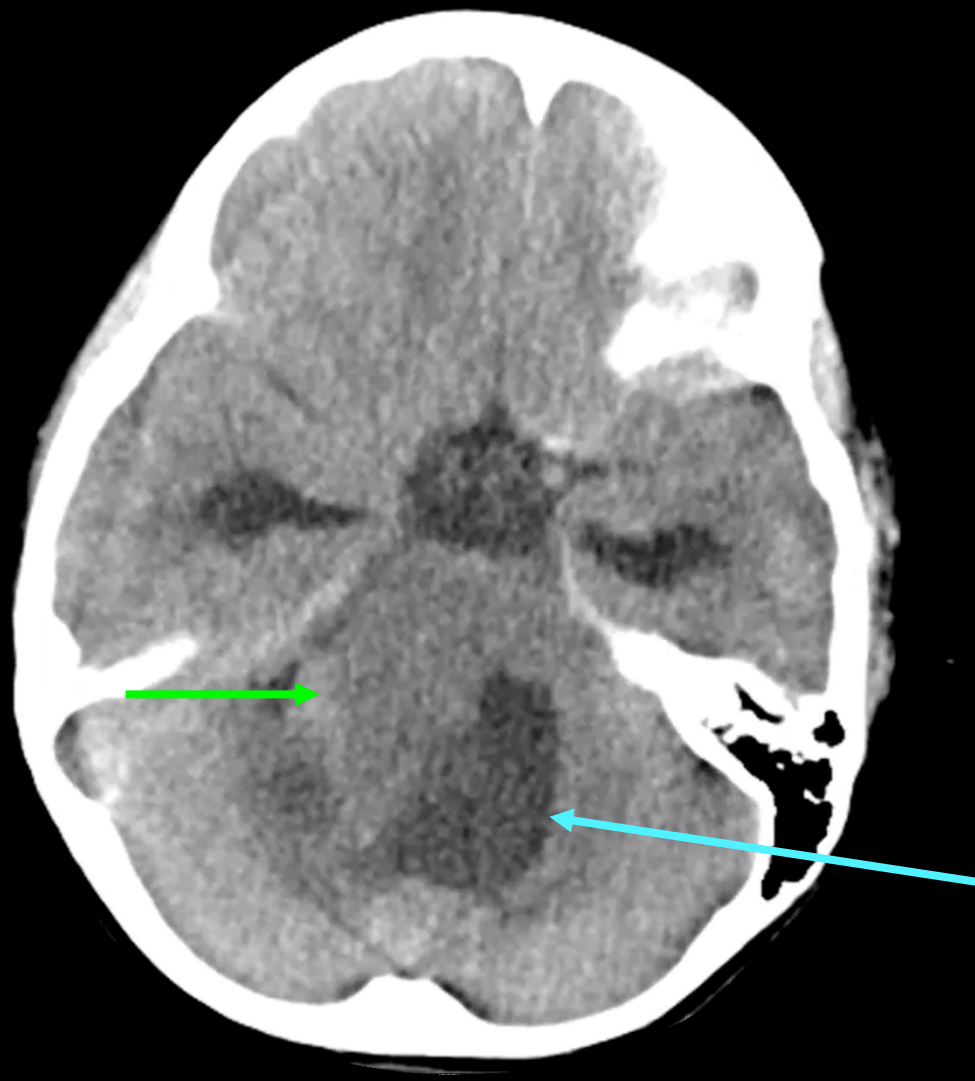
Dilated temporal horns of lateral ventricle

Complete effacement of 4th ventricle

Non-contrast CT

7/30/2019

- Approximately 38 x 43 x 37 mm (AP x TV x CC) heterogenous mass with cystic and solid components centered within R lateral cerebellar hemisphere, R middle cerebellar peduncle and cerebellar vermis causing complete effacement of the 4th ventricle



Non-contrast CT

7/30/2019

- Approximately 38 x 43 x 37 mm (AP x TV x CC) heterogenous mass with cystic and solid components centered within R lateral cerebellar hemisphere, R middle cerebellar peduncle and cerebellar vermis causing complete effacement of the 4th ventricle
- Moderate supratentorial obstructive hydrocephalus and R > L cerebellar tonsillar herniation with effacement of the subarachnoid fluid spaces across the foramen magnum

Differential Diagnosis

Posterior fossa masses in children

- Medulloblastoma
- Pilocytic astrocytoma
- Ependymoma

Differential Diagnosis

Pilocytic astrocytoma

Case courtesy of Prof Frank Gaillard, Radiopaedia.org, rID: 8474

Medulloblastoma

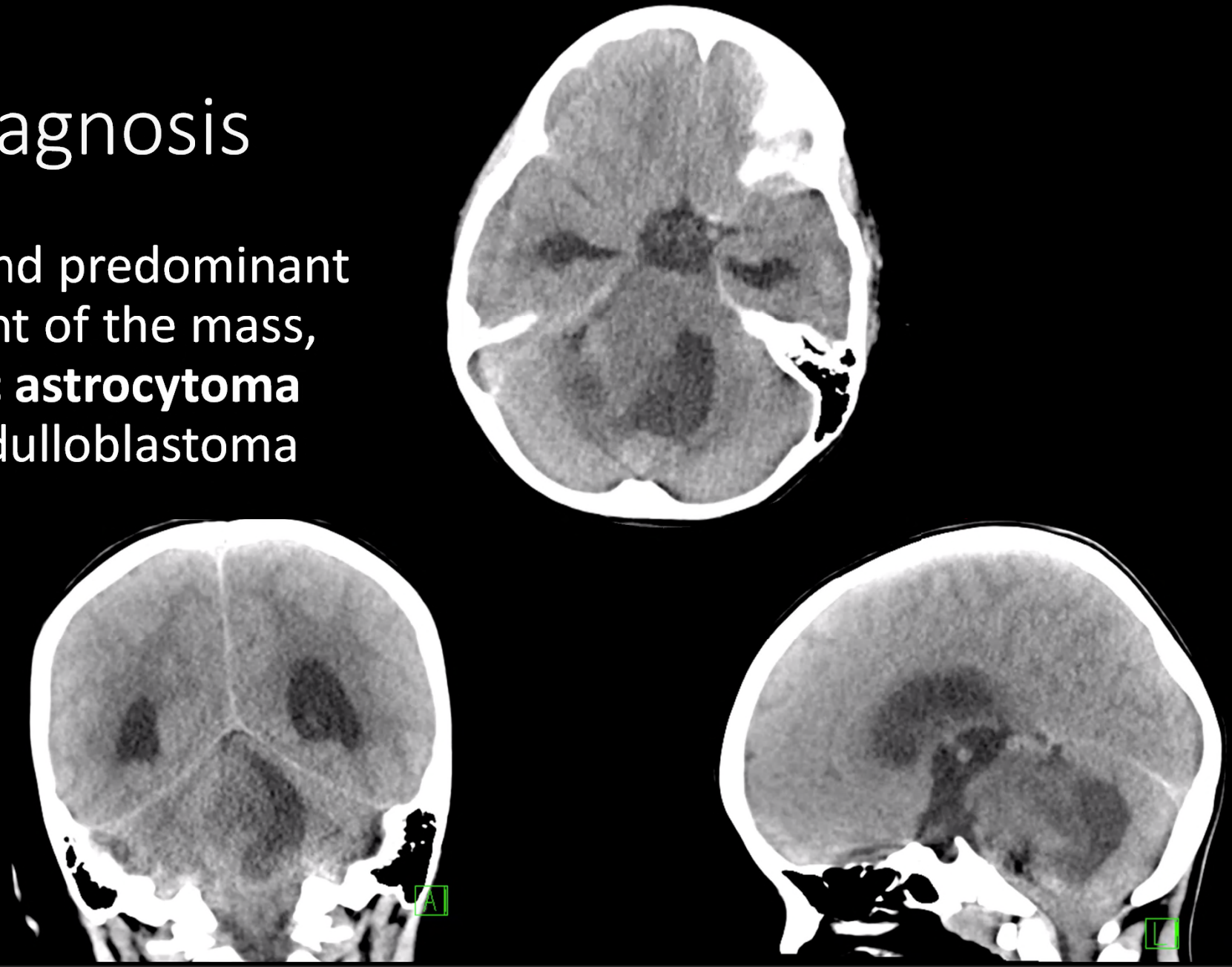
Case courtesy of Prof Frank Gaillard, Radiopaedia.org, rID: 7912

Ependymoma

Case courtesy of Dr Hani Salam, Radiopaedia.org, rID: 15699

Differential Diagnosis

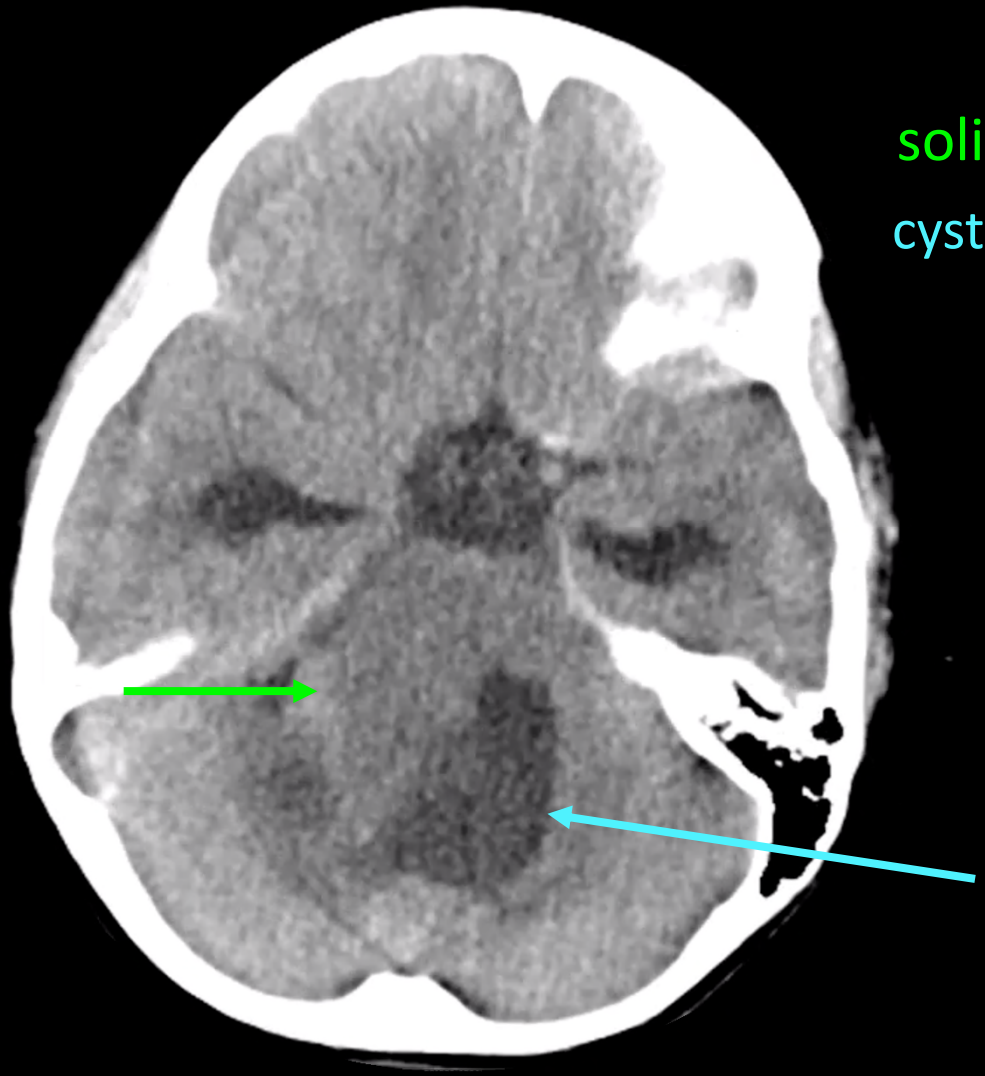
With given location and predominant hypodense component of the mass, possibility of **pilocytic astrocytoma** was favored over medulloblastoma and ependymoma.



J.M. (cont’d)

- Admitted to neurosurgery service
- MRI brain and spine ordered for operative planning

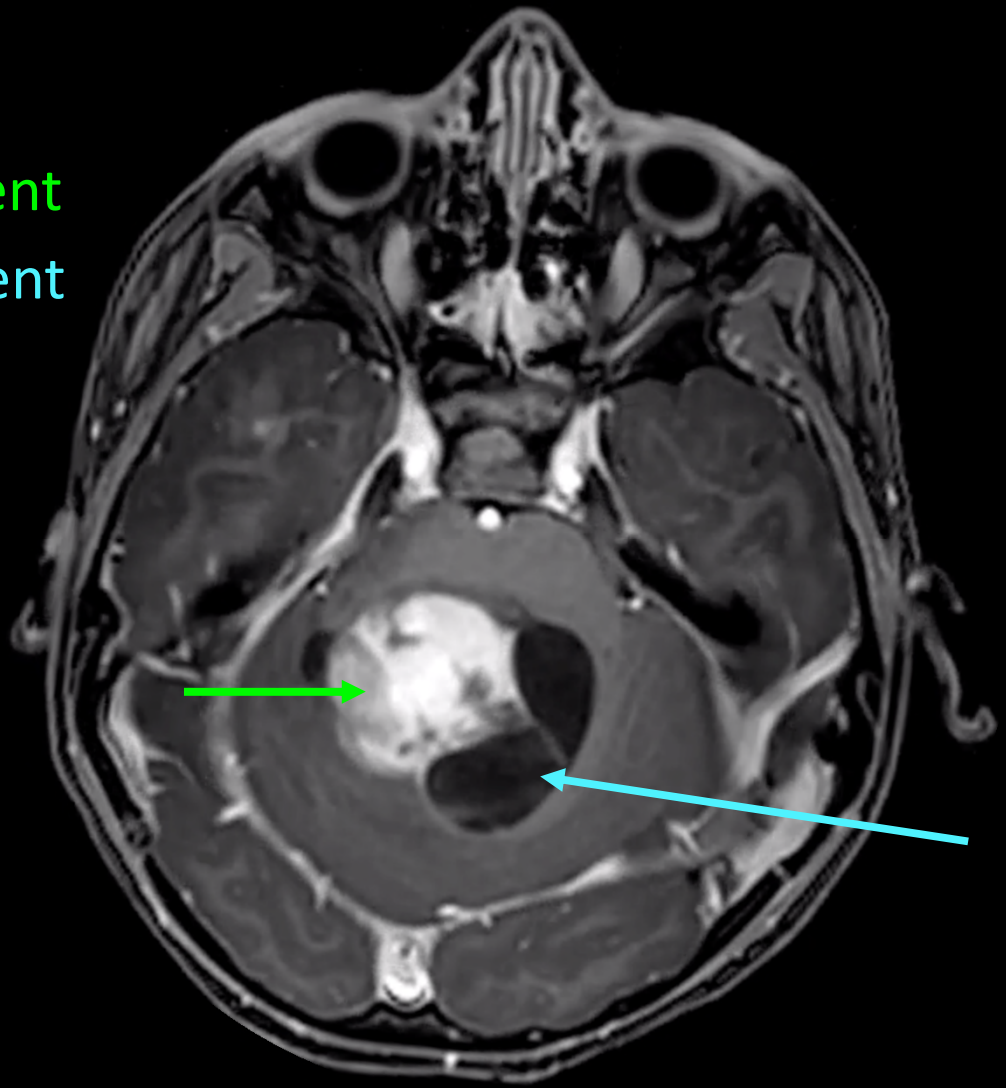
Pre-operative CT vs. MRI



solid component

cystic component

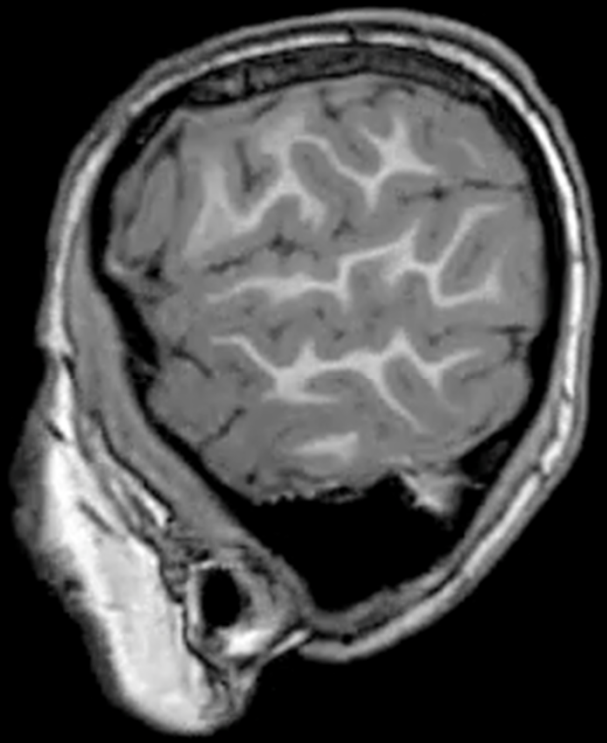
CT without contrast



T1 with contrast

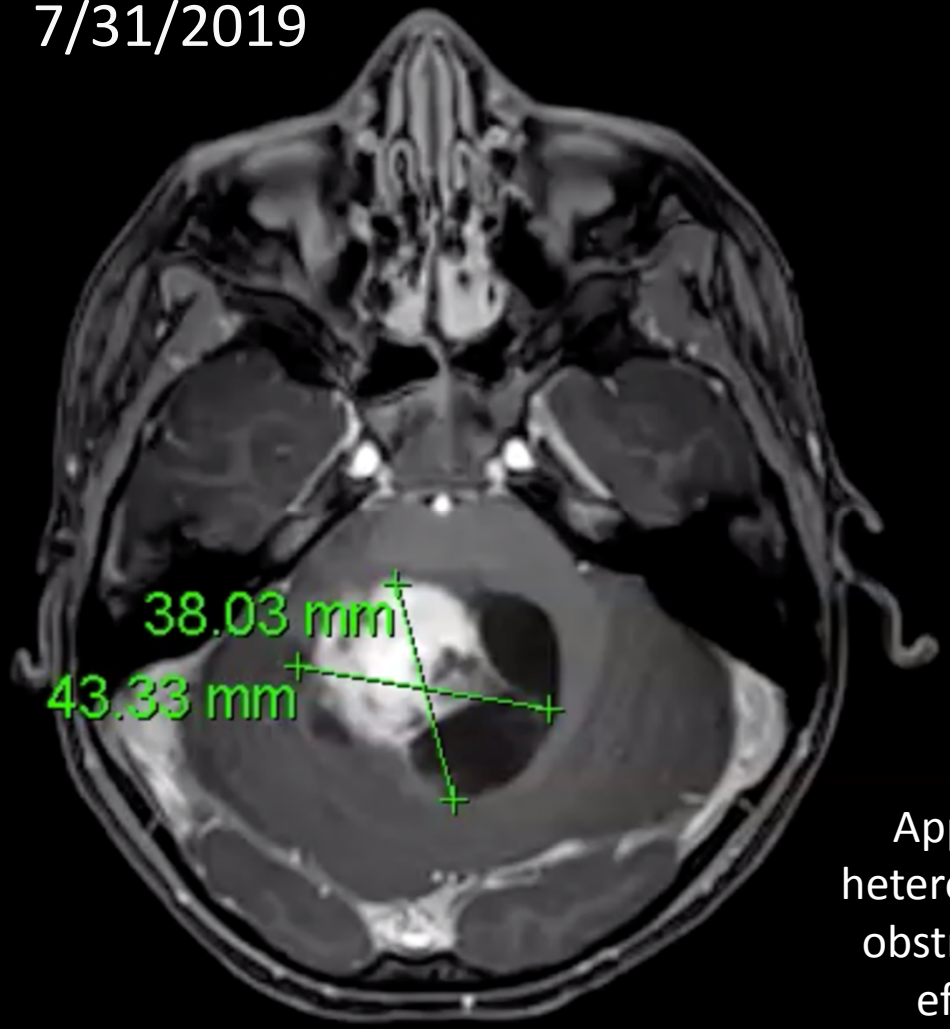
Pre-operative T1 3D

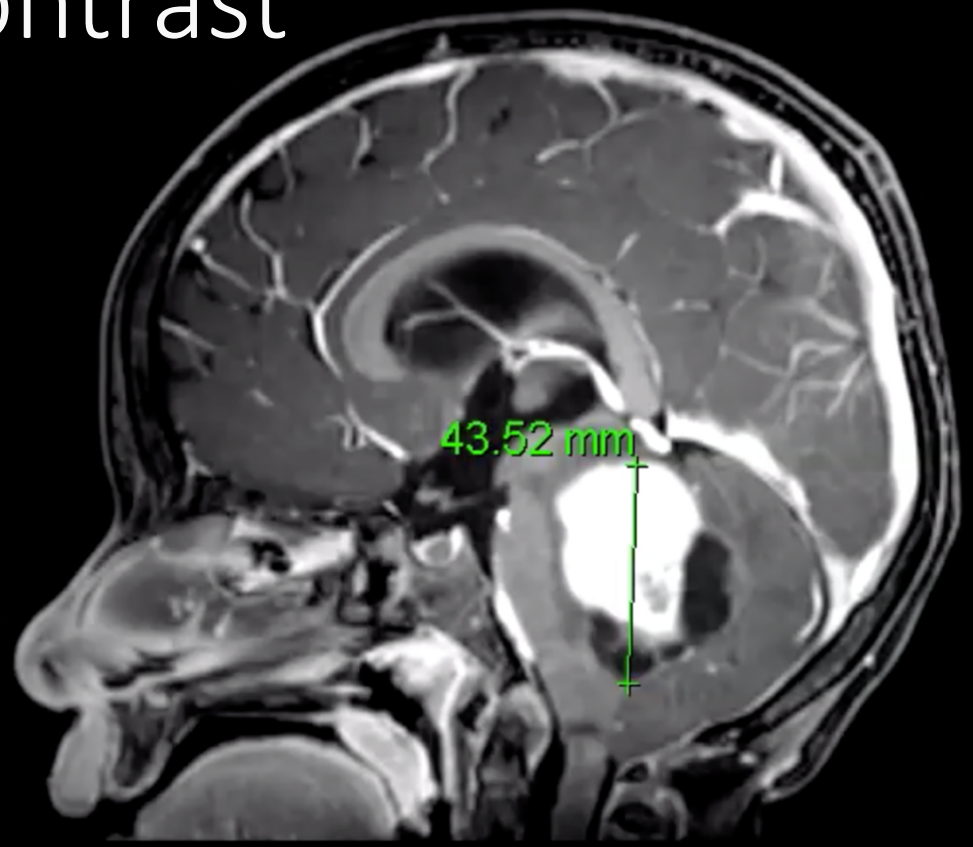
7/31/2019



Pre-operative T1 with contrast

7/31/2019





Approximately 38 x 43 x 44 mm (AP x TV x CC) mixed solid cystic and heterogeneous mass in R posterior fossa causing moderate supratentorial obstructive hydrocephalus and R > L cerebellar tonsillar herniation with effacement of subarachnoid fluid spaces across foramen magnum

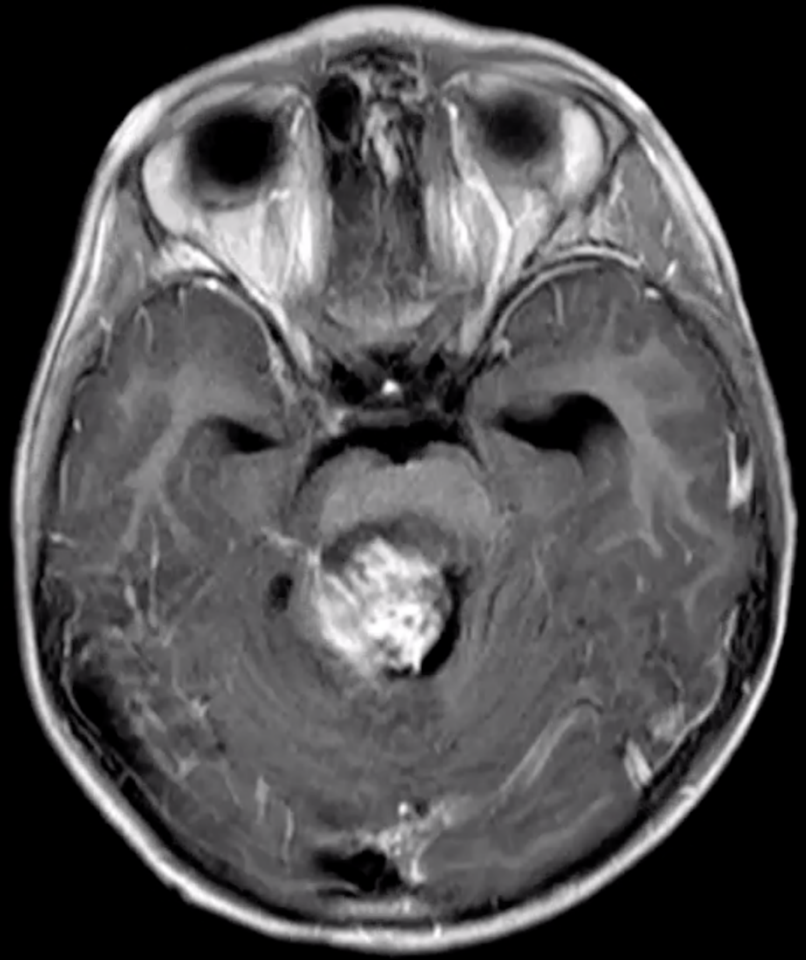
Treatment

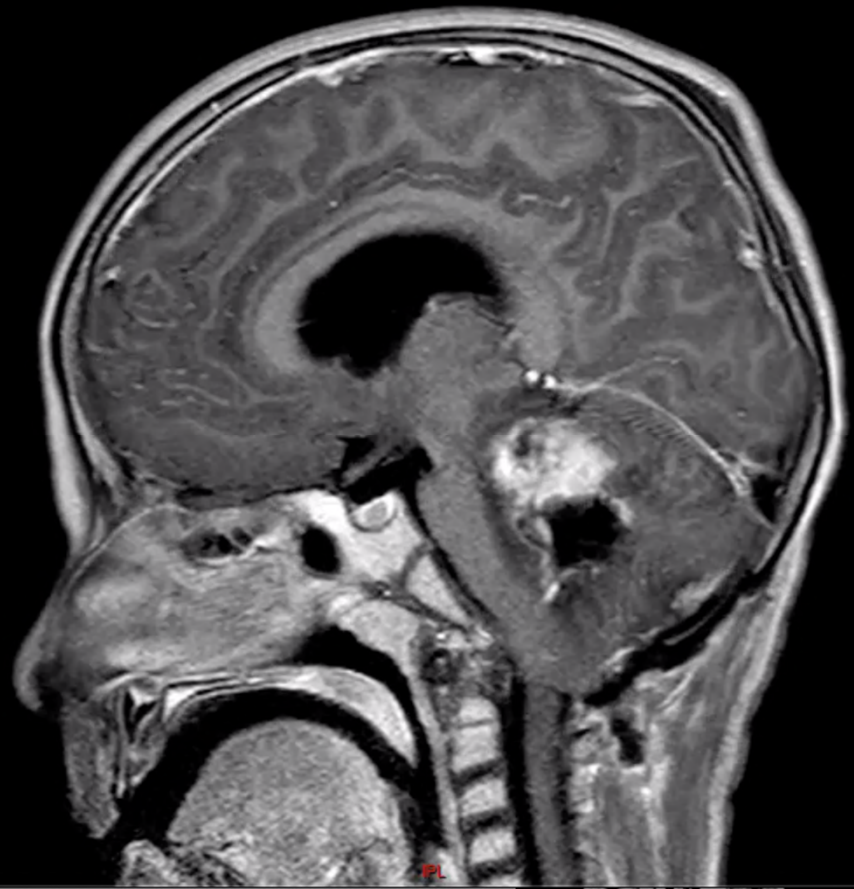
- Tumor debulking and VP shunt placement on 8/2/2019
- 24-hour post-op CT and MRI obtained

Post-operative T1 with contrast

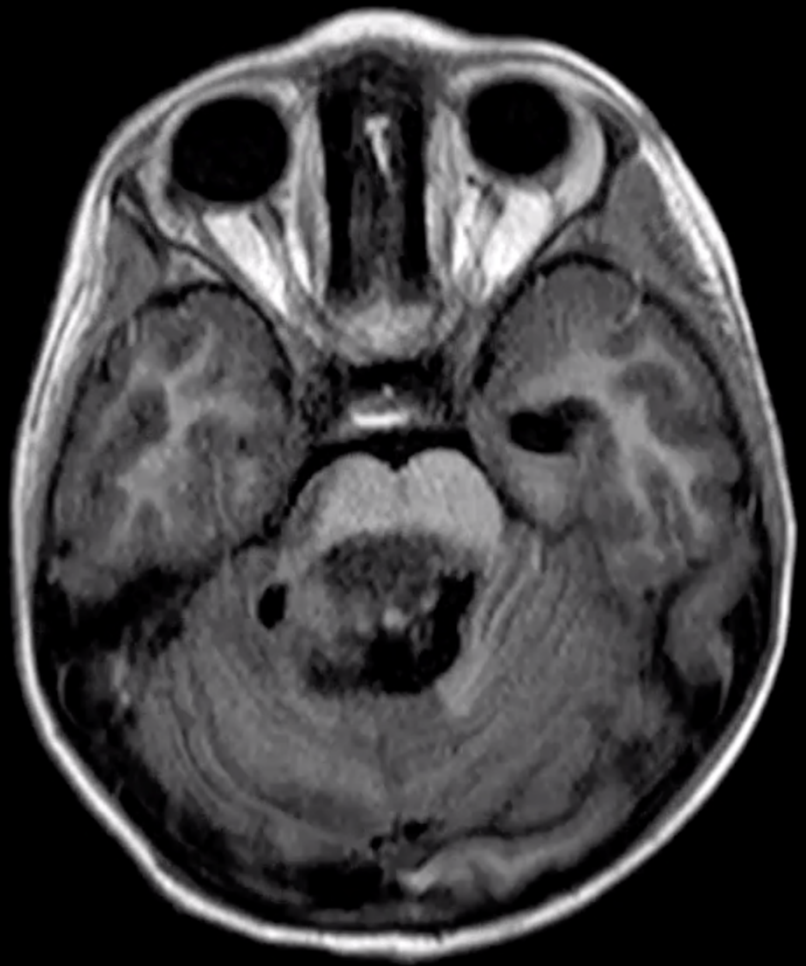
8/2/2019

Post-op coronal T1 with contrast



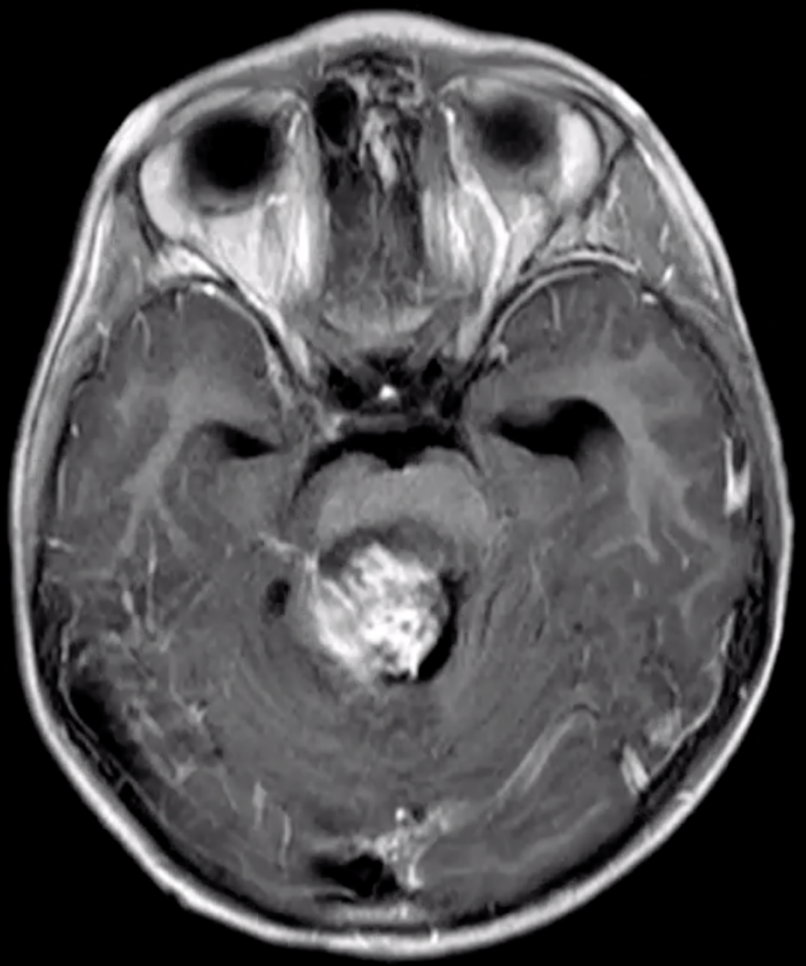


Post-operative T1



T1 without contrast

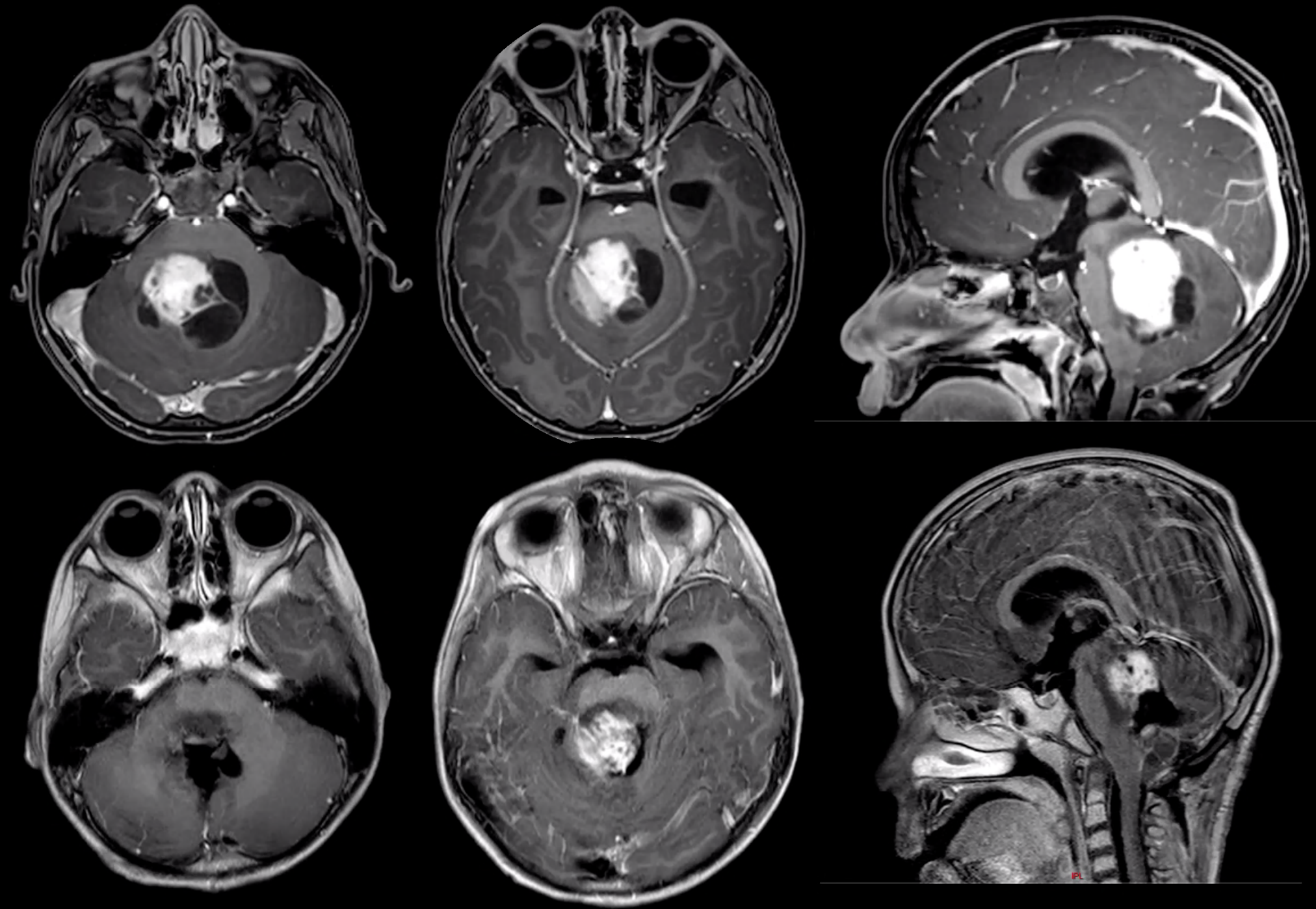
Minimal areas of residual enhancing tumor measuring 24 x 23 x 28 mm (AP x LV x CC)



T1 with contrast

Pre-operative

Post-operative



Brainstem Pilocytic Astrocytoma

- Final diagnosis made by pathology
- WHO grade I tumor
- Often cystic, slow-growing tumor seen in children
- Radical resection is often curative
  - 5-year survival rate: 92%
  - 25-year survival rate: 88%
- Most common residual effects are emotional lability & disequilibrium

Hospital course

- 8/1/2019: tumor debulking, VP shunt placement
- 8/2/2019: post-operative imaging (MRI), hydrocephalus (CT)
- 8/3/2019: hydrocephalus (MRI)
- 8/5/2019: hydrocephalus (MRI)
- 8/9/2019: altered mental status (CT)
- 8/15/2019: Discharged to Shriner’s for inpatient rehab

Post-operative course

- Hydrocephalus

POD 1
8/2/2019

POD 8
8/9/2019

Post-operative course

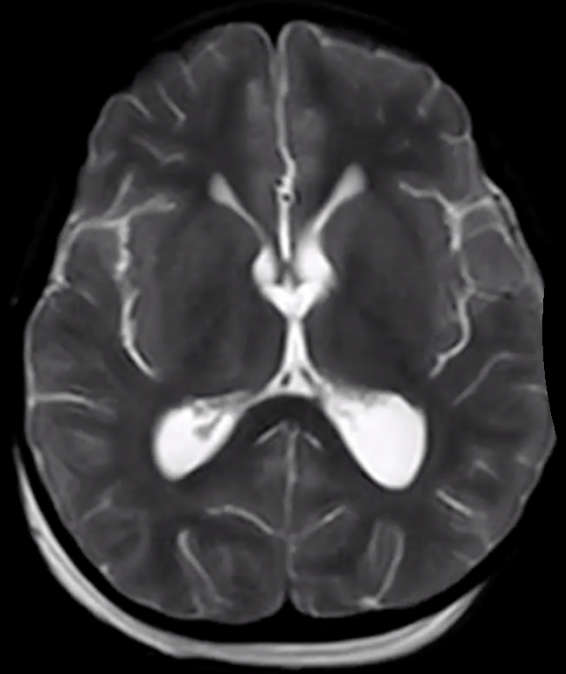
- Hydrocephalus protocol
  - Interval decrease in ventricle size by POD 8
- Discharge disposition
  - Residual left-sided weakness
  - Horizontal gaze palsy

Follow up

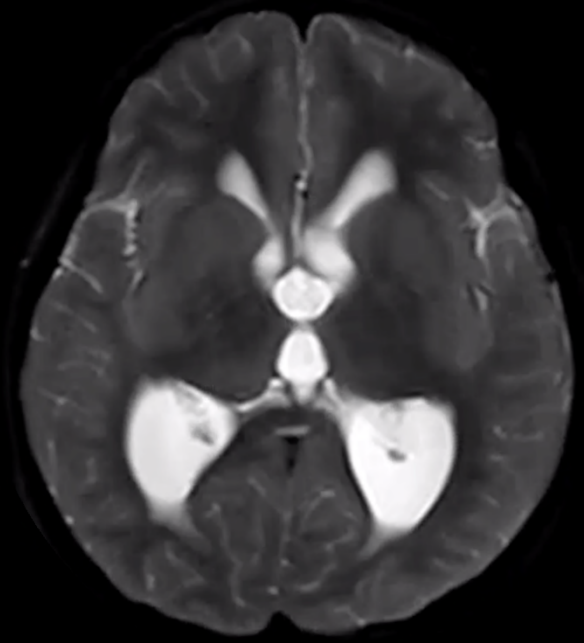
- Readmitted 9/10/2019 for acute aggression
  - Found to have obstructive hydrocephalus
  - 9/11/2019: Right frontal endoscopic third ventriculostomy
  - Discharged on POD #4

Follow up

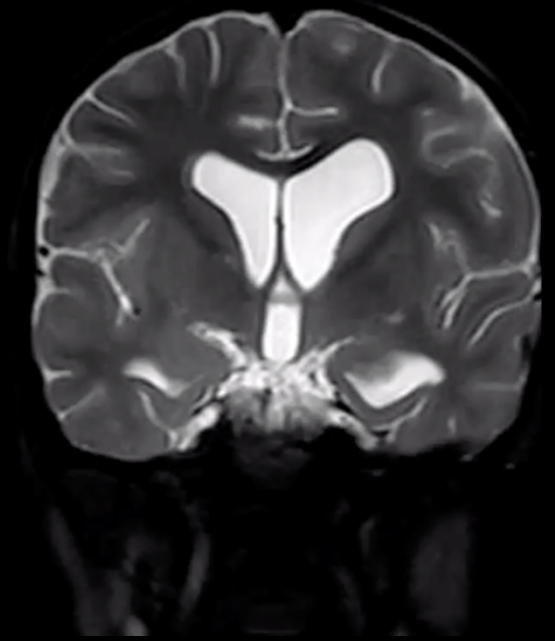
9/10/2019 readmission for acute aggression



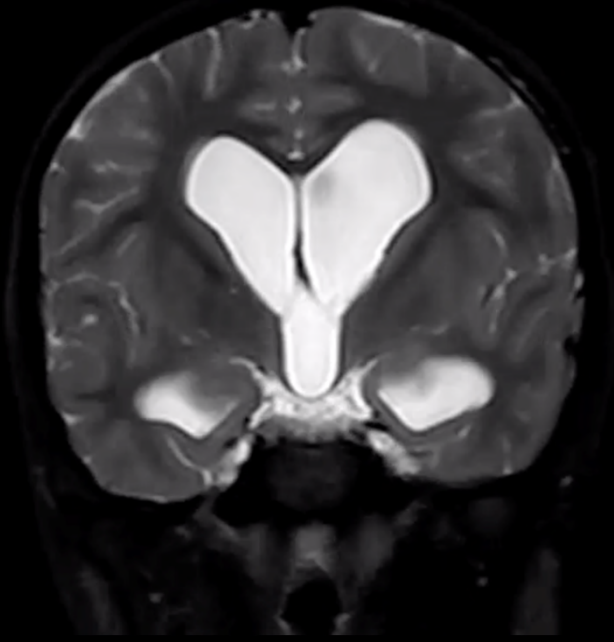
T2 from 8/5/2019



T2 on admission



T2 from 8/5/2019



T2 on admission

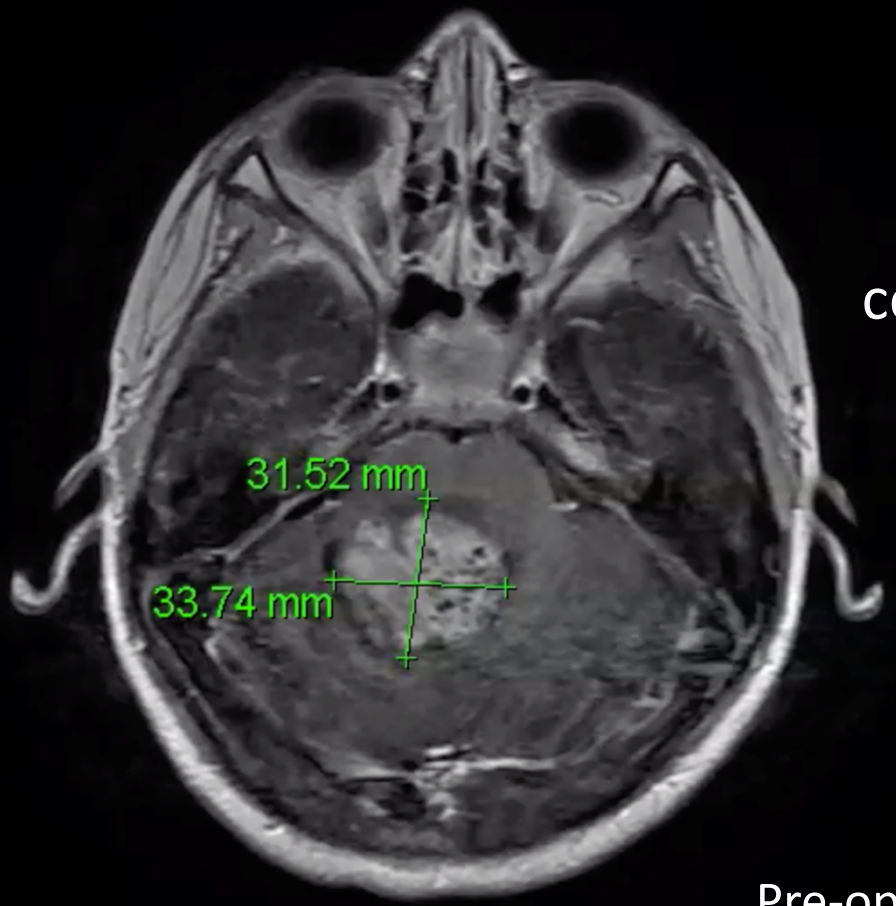
Interval increase in supratentorial ventricular caliber related to noncommunicating obstructive hydrocephalus with new periventricular edema; interval increase in size of residual tumor along superior aspect of 4th ventricle and adjacent R brainstem with associated worsening mass effect on the adjacent brainstem and 4th ventricle

Follow up

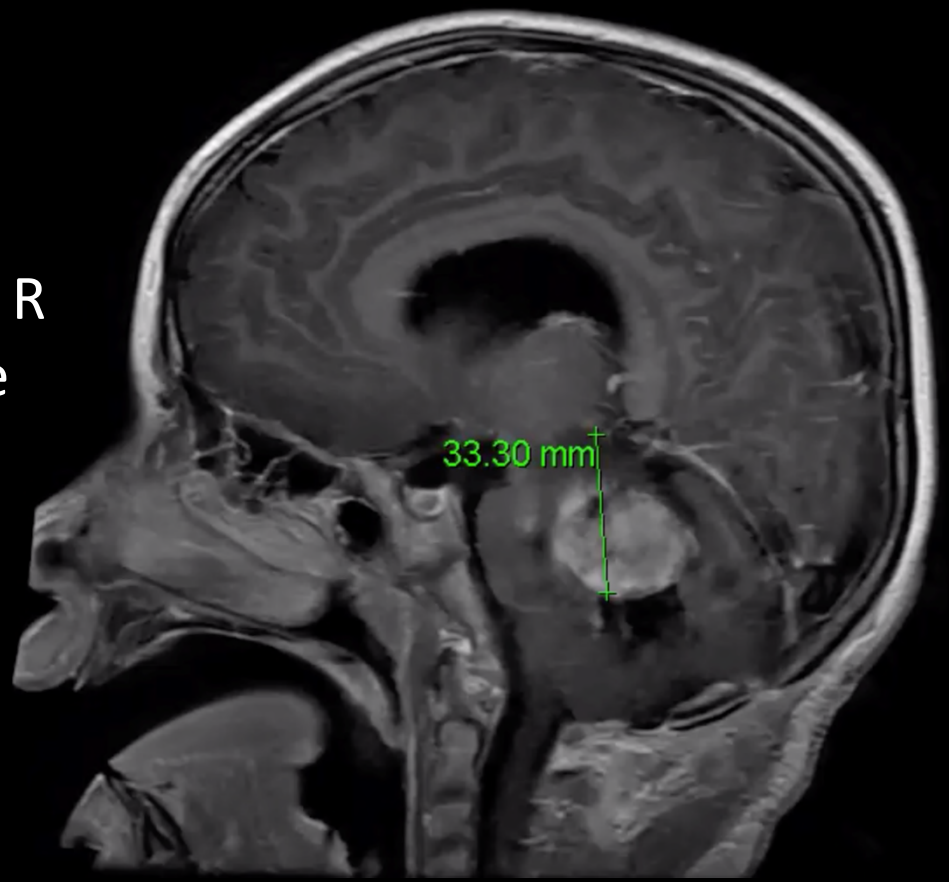
- Readmitted 9/10/2019 for acute aggression
  - Found to have obstructive hydrocephalus
  - 9/11/2019: Right frontal endoscopic third ventriculostomy
  - Discharged on POD #4
- Direct admit from clinic 10/2/2019 for tumor recurrence
  - 10/4/2019: repeat tumor debulking and C1 laminectomy
  - Discharged on POD #3

Follow up

10/2/2019 direct admission from clinic for tumor recurrence



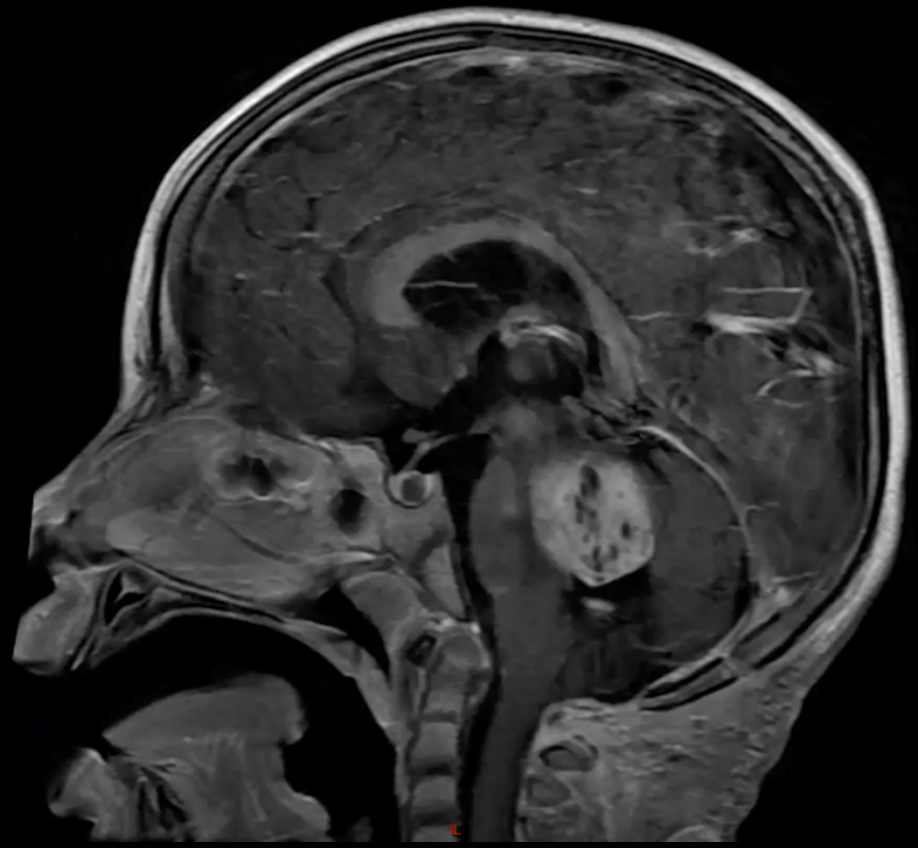
Enhancing tumor in cerebellar vermis and R cerebral hemisphere measuring
31 x 33 x 33 mm



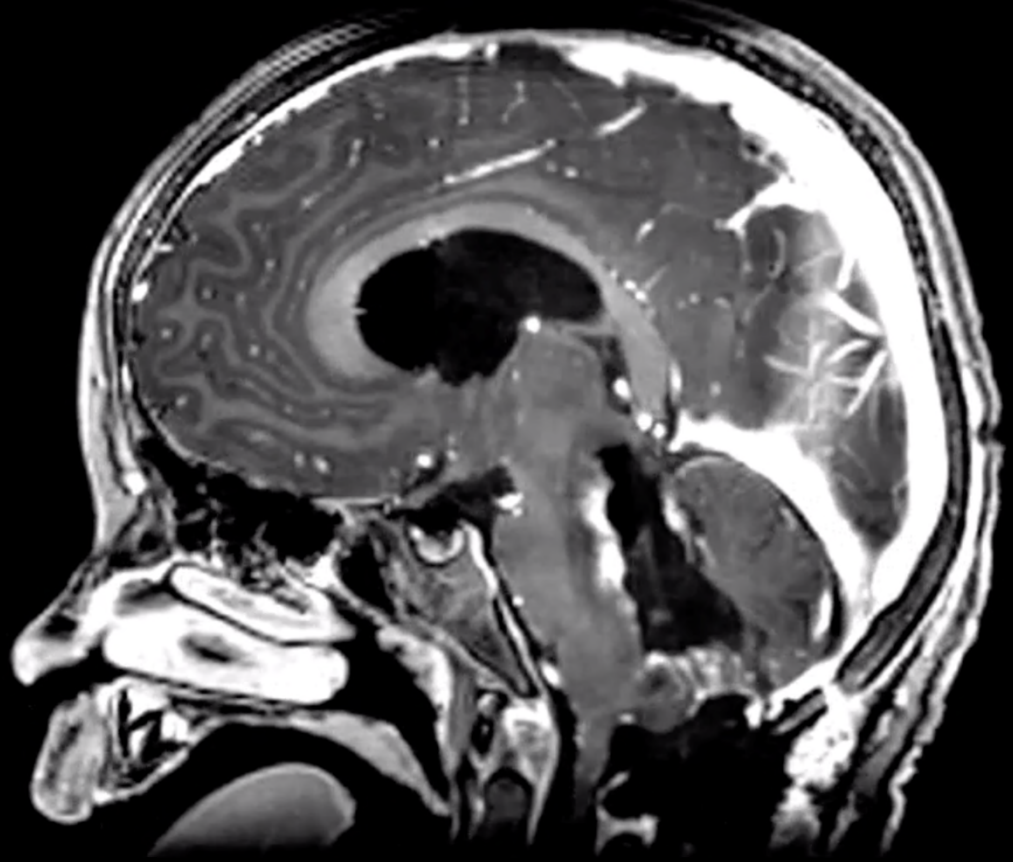
Pre-operative T1 FLAIR with contrast

Follow up

10/2/2019 direct admission from clinic for tumor recurrence



Pre-operative T1 with contrast



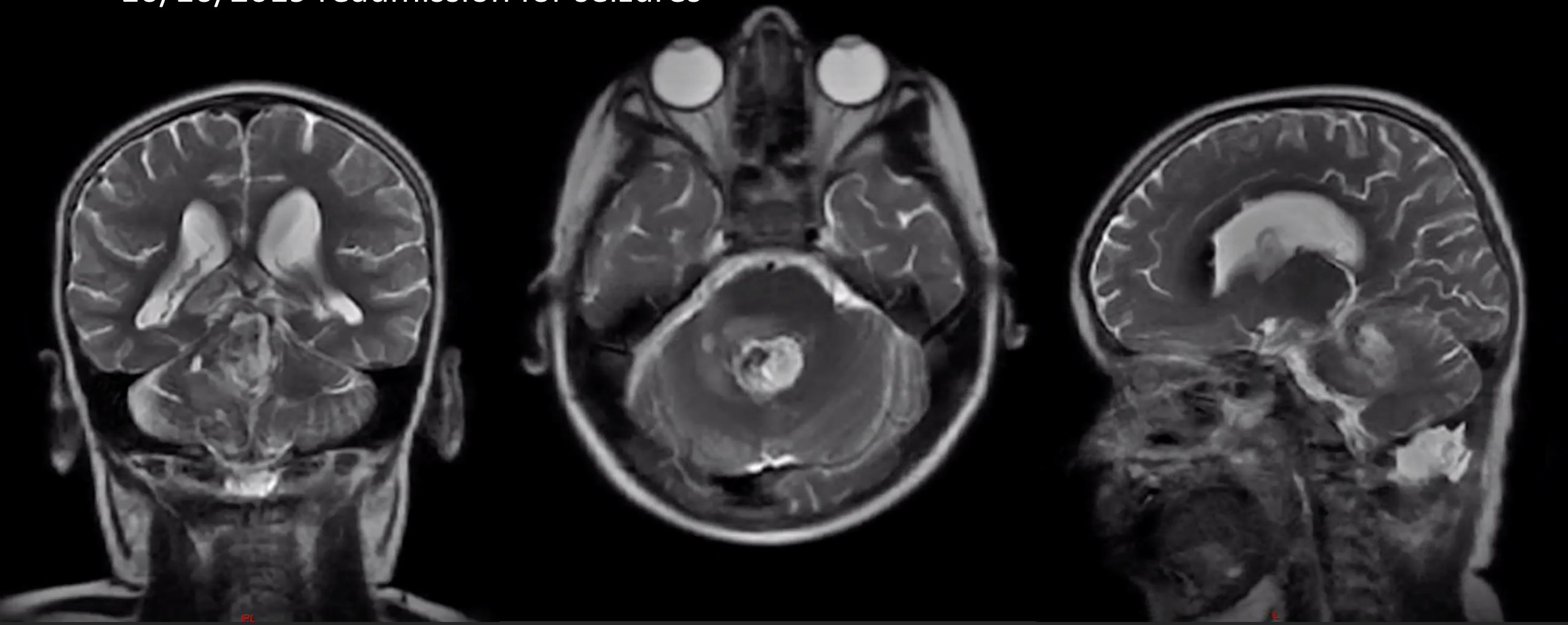
Post-operative T1 with contrast

Follow up

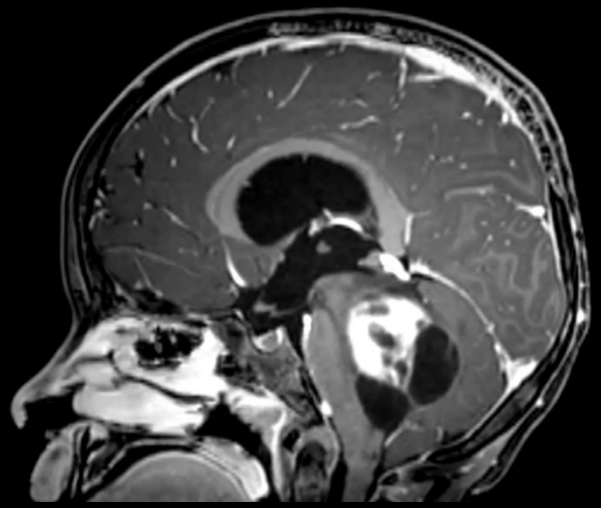
- Readmitted 9/10/2019 for acute aggression
  - 9/11/2019: Right frontal endoscopic third ventriculostomy
  - Discharged on POD #4
- Direct admit from clinic 10/2/2019 for tumor recurrence
  - 10/4/2019: repeat tumor debulking
  - Discharged on POD #3
- Readmitted 10/10/2019 for seizures and visual hallucinations
  - Found to have CSF infection with *S. aureus*
  - Still admitted at time of case presentation

Follow up

10/10/2019 readmission for seizures

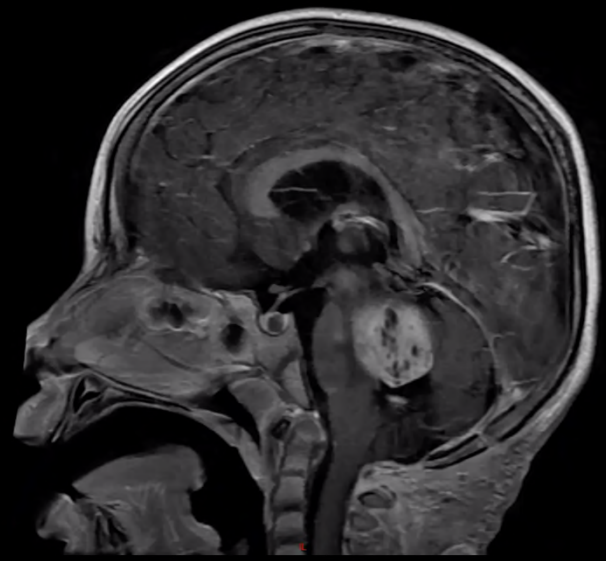


Tumor Progression

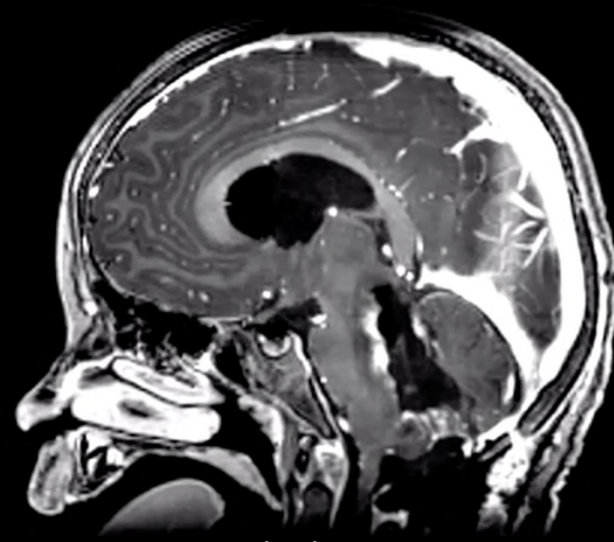


7/31/2019
(Pre-operative)
38 x 43 x 34 mm

8/2/2019
(Post-operative)



10/2/2019
(Pre-operative)
31 x 33 x 33 mm



10/5/2019
(Post-operative)

Imaging Costs

Procedure	Total Cost	Avg. Out-of-Pocket	# Obtained	Total Out-of-Pocket
Non-contrast head CT	$3,157	$98	4	$392
MRI spine with and without contrast	$23,464	$995	1	$995
MRI brain with and without contrast	$7,431	$523	4	$2,092
MRI brain without contrast	$5126	$417	4	$1,668
Chest x-ray	$762	$250	3	$750
Abdominal x-ray	$670	$52	2	$104
Retroperitoneal ultrasound	$1576	$224	1	$224
Barium swallow video evaluation	$918	$221	1	$221
Total Cost	**$92,440**	**Out-of-Pocket Expense**		**$6,446**

https://www.memorialhermann.org/patients-caregivers/memorial-hermann-charge-master/

ACR appropriateness Criteria

- MRI preferred over CT on initial presentation of ataxia in adults
  - Young kids pose a special consideration as MRI may require general anesthesia

Variant 3: **Ataxia. No history of trauma. Suspected intracranial process. Stroke intervention not a consideration. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head with IV contrast	May Be Appropriate	☢☢☢
CT head without IV contrast	May Be Appropriate	☢☢☢
CT head without and with IV contrast	May Be Appropriate	☢☢☢
CTA head and neck with IV contrast	Usually Not Appropriate	☢☢☢
MRA head and neck without and with IV contrast	Usually Not Appropriate	O
MRA head and neck without IV contrast	Usually Not Appropriate	O
Arteriography cervicocerebral	Usually Not Appropriate	☢☢☢
CTV head with IV contrast	Usually Not Appropriate	☢☢☢
I-123 Ioflupane SPECT/CT brain	Usually Not Appropriate	☢☢☢
MRV head with IV contrast	Usually Not Appropriate	O
MRV head without IV contrast	Usually Not Appropriate	O
In-111 DTPA cisternography	Usually Not Appropriate	☢☢☢

ACR appropriateness Criteria

- MRI preferred over CT on initial presentation of ataxia in adults
  - Young kids pose a special consideration as MRI may require general anesthesia

- Per American Academy of Pediatrics, MRI is standard of care for all children with suspected brain tumor
  - Has the major benefit of no radiation exposure

Discussion

- Radiation since original presentation on 7/31/2019
  - 4 Non-contrast CT x 2 mSv = 8 mSv
  - 3 Chest XR x 0.1 mSv = 0.3 mSv
  - 1 Barium swallow x 6 mSv = 6 mSv
  - 2 Abdominal XR x 8 mSv = 16 mSv

*Radiation doses are adjusted for body weight

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Discussion

Pediatric Posterior Fossa Tumors

- Classic triad of gait imbalance, headache, nausea/vomiting
  - Any of the above symptoms + an abnormality on neuro exam warrants a referral for neuroimaging, MRI being the modality of choice
- Treatment depends on type and location of tumor
  - Medulloblastoma often requires resection, radiation, and chemotherapy
  - Ependymoma responds best to surgical resection and post-op radiation
  - Pilocytic astrocytoma requires surgical resection

Discussion

Juvenile Pilocytic Astrocytoma

- Most common posterior fossa tumor in kids > 4 years old
- If near-total resection and/or tumor recurs, repeat surgical resection with option to add chemotherapy or radiation to treat residual tumor
- Long-term follow up when gross total resection is achieved
  - MRI at 6 months, then 1, 2, 3.5 and 5 years
    - Recent studies have suggested 2 consecutive negative MRIs ≥ 3 months apart is sufficient
- Long-term follow up for patients with residual tumor
  - MRI every 6 months for 3 years, annually for 2 additional years, and every other year thereafter indefinitely

Discussion

Case of J.M.

- Patient had been seen by pediatrician for slowly progressing ataxia a couple weeks prior to presentation at ED
  - Referred to ortho clinic for leg length discrepancy
  - Thorough neurological exam should have been completed by pediatrician who then should have placed order for MRI
    - Would have spared cost, time, and radiation exposure
- Will require post-operative surveillance MRIs and consideration of chemotherapy or targeted radiation if tumor recurs

Take Home Points

- There is a low threshold for obtaining neuroimaging for any child presenting with new-onset, progressive ataxia
- MRI carries significantly less radiation risk than CT or XR and is therefore the preferred neuroimaging modality for pediatric patients
- Juvenile pilocytic astrocytomas carry an overall good prognosis following radical resection but can be complicated by recurrence if resection is not complete

References

1. Crawford JC. Childhood bran tumors. *Pediatrics in Review.* 2013;34(2):63-78.
2. Gaillard F, Radiopaedia.org, rID: 8474. 3 Feb 2010.
3. Gaillard F, Radiopaedia.org, rID: 7912. 23 Dec 2009.
4. Hacking C, Radiopaedia.org, rID: 62252. 16 Aug 2018.
5. Juliano AF, Policeni B, Agarwal V, et al. ACR Appropriateness Criteria® Ataxia. Available at https://acsearch.acr.org/docs/69477/Narrative/. American College of Radiology. Accessed 20 Oct 2019.
6. Maher CO, Raffel C. Neurosurgical treatment of brain tumors in children. *Pediatr Clin North Am*. 2004;51(2):327-57.
7. Mitchell RN, Kumar V, Fausto N et al. Chapter 28: The central nervous system. In: Mitchell RN, Kumar V, Fausto N et al. eds. *Pocket Companion to Robbins & Cotran Pathologic Basis of Disease*. 9th ed. Philadelphia, PA: Elsevier Health Sciences; 2016:1309.
8. Ronghe M, Hargrave D, Bartels U, et al. Vincristine and carboplatin chemotherapy for unresectable and/or recurrent low-grade astrocytoma of the brainstem. *Pediatr Blood Cancer*. 2010;55(3):471-7.
9. Salam H, Radiopaedia.org, rID: 15699. 1 Nov 2011.
10. Saunders DE, Phipps KP, Wade AM, Hayward RD. Surveillance imaging strategies following surgery and/or radiotherapy for childhood cerebellar low-grade astrocytoma. *J Neurosurg*. 2005;102(2 Suppl):172-8.
11. Yecies D, Fisher PG, Cheshier S, Edwards M, Grant G. Long-term outcomes of primarily metastatic juvenile pilocytic astrocytoma in children. *J Neurosurg Pediatr*. 2018;21(1):49-53.


Questions?