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
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

lateral ventricle
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 4\textsuperscript{th} 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 4\textsuperscript{th} 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

CT without contrast

T1 with contrast

cystic component

solid component
Pre-operative T1 3D
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

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

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
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

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Total Cost</th>
<th>Avg. Out-of-Pocket</th>
<th># Obtained</th>
<th>Total Out-of-Pocket</th>
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<tbody>
<tr>
<td>Non-contrast head CT</td>
<td>$3,157</td>
<td>$98</td>
<td>4</td>
<td>$392</td>
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<tr>
<td>MRI spine with and without contrast</td>
<td>$23,464</td>
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<td>MRI brain with and without contrast</td>
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<td>$523</td>
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<td>$2,092</td>
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<td>$417</td>
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<tr>
<td>Chest x-ray</td>
<td>$762</td>
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<td>Abdominal x-ray</td>
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<td>Retroperitoneal ultrasound</td>
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<td>Barium swallow video evaluation</td>
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<td><strong>Total Cost</strong></td>
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<td><strong>Out-of-Pocket Expense</strong></td>
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[Link](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

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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</thead>
<tbody>
<tr>
<td>MRI head without and with IV contrast</td>
<td>Usually Appropriate</td>
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<tr>
<td>MRI head without IV contrast</td>
<td>Usually Appropriate</td>
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<tr>
<td>CT head with IV contrast</td>
<td>May Be Appropriate</td>
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<tr>
<td><strong>CT head without IV contrast</strong></td>
<td>May Be Appropriate</td>
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<tr>
<td>CT head without and with IV contrast</td>
<td>May Be Appropriate</td>
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<td>CTA head and neck with IV contrast</td>
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<td>MRA head and neck without and with IV contrast</td>
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<tr>
<td>MRA head and neck without IV contrast</td>
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<tr>
<td>Arteriography cervicocerebral</td>
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<td>CTV head with IV contrast</td>
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<td>MRV head with IV contrast</td>
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<tr>
<td>MRV head without IV contrast</td>
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<tr>
<td>In-111 DTPA cisternography</td>
<td>Usually Not Appropriate</td>
<td>☢</td>
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</tbody>
</table>
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
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

Questions?