17yo female with pituitary mass

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Chief Complaint

- 17 5/12yo F with h/o pituitary mass s/p transsphenoidal partial hypophysectomy
HPI

- Presented to PCP with various sx including dizziness, fatigue, temperature intolerance, loss of taste, palpitations, twitching
- Work-up (labs, EKG) were nL
- Saw a traditional Chinese doctor who recommended various herbs
- MRI done showed pituitary hyperplasia (8mm) at upper limits of nL for pubertal F but no clear mass
- Referred to NSGY and endo
ROS

- Constitutional: Negative for fever, +fatigue
- Endo: Negative for galactorrhea, polyuria
- HEENT: Negative for neck pain, +tinnitus, visual disturbances
- CV: +palpitations
- GU: Negative for menstrual abnormalities
- Skin: +flushing
- Neurol: Negative for HAs and syncope, +dizziness
- Psych: Negative for behavioral changes
Physical Exam

- Vitals: T 37°C, HR 87, RR 15, bp 109/53, wt 51.2 kg (50th%), ht 157.4 cm (25th%), BMI 21.4 kg/m² (54th%)
- General: well-developed, NAD
- HEENT: normocephalic, PERRL, intact visual fields
- Neck: thyroid palpable, not enlarged, smooth
- CV/Pulm/Chest: RRR, CTAB, Tanner 4 breasts, no discharge
- Neuro: alert, no focal deficits, 2+DTRs
- Skin: normal pigmentation, mild acanthosis on neck, comedonal facial acne
Labs

- TSH 1.85
- Total T4 6.6
- Free T4 1.15
- ACTH 14.5
- Cortisol 12.2
- Prolactin 10.5
- FSH 7.4
- LH 7.1
- 17OHP 47

- DHEAS 103
- Total testosterone 19
- Free testosterone 0.6
- SHBG 22
- Insulin 9.9
- HbA1C 5.2
- Urine cortisol 9.6
- Urine metanephrines 71
- CMP normal
Assessment/Plan

- Pituitary hyperplasia with intact pituitary function—monitor clinically
- Follow-up with NSGY and endo
- Repeat MRI in 3 months
3-month NSGY f/u

- HPI and PE stable
- MRI: interval growth of pituitary gland now abutting optic chiasm without compression (10 mm)
- Assessment: Pituitary hyperplasia vs. adenoma
- Plan: Repeat MRI in 6 months
Interval History

- Next 2.5 years: Pt followed up at Northwestern
- Developed peripheral visual field deficit, irregular menses, mildly elevated prolactin
- Repeat MRI showed R sided pituitary adenoma
- Surgery was recommended and she returned to U of C for second opinion
Interval History- cont’d

- ROS: Negative for fever, galactorrhea, polyuria, HAs +peripheral vision loss, heavy menses Q2weeks for last year
- FH: No change
- SH: Now in 11th grade. Still doing well in school.
- Meds: None
- PE: +bitemporal hemianopsia, Tanner 5 breasts, no discharge, no focal neuro deficits
Imaging
Labs

- TSH 1.07
- Free T4 1.05
- ACTH 9.9
- Cortisol 9.3
- Prolactin 71.34
- IGF₁ 333
- IGFBP₃ 5.0
- FSH 5.1
- LH 13.3
- Estradiol 136
- 17OHP 67
- DHEAS 176
- Total testosterone 23
- Free testosterone 8
- SHBG 17
- Androstenedione 187
Plan?
Management

- Dopamine agonist was considered
- Given prolactin level below <100s, progressive visual field deficits, surgical intervention was recommended by NSGY
Post-op concerns?
AVP

DI:
- Polyuria (>300 mL/kg/d)
- ↑ Serum osmolality (>300 mOsm/kg)
- ↓ Urine osmolality (<600 mOsm/kg)
- Hypernatremia
- polydipsia
Triphasic DI

Diabetes insipidus
Antidiuretic interphase
Diabetes insipidus

POD#1

- Increasing UOP since early AM: ~350 mL/hr (6.7 mL/kg/hr)
- Increasing Na: 140 → 141 → 150
- Increasing thirst but PO limited by nausea
Serum Na and UOP

POD#
Hospital Course - POD #6

Labs:
- TSH 0.02, fT4 0.71
- ADH < 0.5
- ACTH 14.1, Cortisol 0.4
- Prolactin 7.64
- FSH 0.7
- LH < 0.1
- Estradiol 5
- DHEAS < 15
Hospital Course- cont’d

- Drinking to thirst
- Possible D/C home
- Down-trending Na
Serum Na and UOP

POD#
Discharge

- Fluid restriction of 500 mL/d
- Pituitary labs to be rechecked as outpatient
- Stress-dose hydrocortisone instructions
Pathology

- All cell types present with nL architecture
- One small nest of monomorphic cells positive for prolactin
- nL MIB-1 activity
Clinical Questions

• What is the natural history of pituitary hypertrophy?
• How common is post-op DI?
• What are risk factors for developing DI?
Physiologic Pituitary Hypertrophy

- Pituitary enlargement (>9mm) in a young woman or adolescent girl should be considered **normal hypertrophy** if:
  - Pituitary MRI and labs are **normal**

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**TABLE 1.** Baseline gonadotrophic hormonal and neuroradiological characteristics in seven patients with physiological enlargement of the pituitary gland

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age at diagnosis (yr)</th>
<th>Basal LH (IU/L)</th>
<th>LH peak* after GnRH (IU/L)</th>
<th>Basal FSH (IU/L)</th>
<th>FSH peak after GnRH* (IU/L)</th>
<th>Basal α-subunit (IU/L)</th>
<th>α-Subunit peak after GnRH* (IU/L)</th>
<th>Basal PRL (μg/L)</th>
<th>PRL peak after TRH† (μg/L)</th>
<th>Height (mm)</th>
<th>Width (mm)</th>
<th>Follow-up duration (yr)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>15</td>
<td>3.5</td>
<td>20</td>
<td>7</td>
<td>17</td>
<td>0.46</td>
<td>0.62</td>
<td>9</td>
<td>38</td>
<td>10.6</td>
<td>14</td>
<td>4</td>
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<tr>
<td>2</td>
<td>17</td>
<td>0.6</td>
<td>33</td>
<td>2.2</td>
<td>7</td>
<td>0.33</td>
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<td>15</td>
<td>154</td>
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<td>14</td>
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<tr>
<td>3</td>
<td>19</td>
<td>9.4</td>
<td>22</td>
<td>6.2</td>
<td>8.3</td>
<td>0.28</td>
<td>0.6</td>
<td>8</td>
<td>107</td>
<td>10</td>
<td>12</td>
<td>2</td>
</tr>
<tr>
<td>4</td>
<td>23</td>
<td>4.8</td>
<td>14</td>
<td>3.9</td>
<td>6.6</td>
<td>0.3</td>
<td></td>
<td>13</td>
<td>66</td>
<td>10</td>
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<td>7</td>
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<td>24</td>
<td>16</td>
<td>59</td>
<td>8.4</td>
<td>11</td>
<td>0.3</td>
<td></td>
<td>20</td>
<td>43</td>
<td>12</td>
<td>12</td>
<td>2</td>
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<tr>
<td>6</td>
<td>27</td>
<td>2.8</td>
<td>174</td>
<td>3.2</td>
<td>11.5</td>
<td>0.52</td>
<td>4.4</td>
<td>22</td>
<td>107</td>
<td>9</td>
<td>11</td>
<td>8</td>
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<tr>
<td>7</td>
<td>19</td>
<td>14</td>
<td>174</td>
<td>5</td>
<td>11.5</td>
<td>0.52</td>
<td>4.4</td>
<td>22</td>
<td>107</td>
<td>12</td>
<td>16</td>
<td>4</td>
</tr>
</tbody>
</table>

How common is post-op DI?

<table>
<thead>
<tr>
<th>Study</th>
<th># of Procedures</th>
<th>Transient DI</th>
<th>Chronic DI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Berker et al.</td>
<td>624</td>
<td>29 (4.6%)</td>
<td>3 (0.5%)</td>
</tr>
<tr>
<td>Frank et al.</td>
<td>381</td>
<td>N/A</td>
<td>6 (1.6%)</td>
</tr>
<tr>
<td>Zhou et al.</td>
<td>375</td>
<td>14 (3.7%)</td>
<td>N/A</td>
</tr>
<tr>
<td>Gondim et al.</td>
<td>341</td>
<td>15 (4.4%)</td>
<td>4 (1.2%)</td>
</tr>
<tr>
<td>Yano et al.</td>
<td>213</td>
<td>10 (4.7%)</td>
<td>2 (0.9%)</td>
</tr>
<tr>
<td>Dehdashti et al.</td>
<td>200</td>
<td>5 (2.5%)</td>
<td>2 (1%)</td>
</tr>
</tbody>
</table>

### Risk factors for DI

#### TABLE 1. Incidence of postoperative diabetes insipidus in patients as determined by sex, tumor size, prior pituitary surgeries, intraoperative cerebrospinal fluid leaks, postoperative cerebrospinal fluid leaks, and preoperative apoplexy

<table>
<thead>
<tr>
<th></th>
<th>Total no.</th>
<th>No DI</th>
<th>Transient DI (&lt;6 mo)</th>
<th>Permanent DI (&gt;6 mo)</th>
<th>Overall DI</th>
<th>Incidence within subtype (%)</th>
<th>P value</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sex</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>53</td>
<td>46</td>
<td>5</td>
<td>2</td>
<td>7</td>
<td>13.2%</td>
<td>0.545</td>
<td>0.23–1.79</td>
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<tr>
<td>F</td>
<td>57</td>
<td>46</td>
<td>10</td>
<td>1</td>
<td>11</td>
<td>19.3%</td>
<td>0.545</td>
<td>0.56–4.41</td>
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<tr>
<td><strong>Adenoma size</strong></td>
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<td></td>
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<tr>
<td>Macroadenoma</td>
<td>78</td>
<td>66</td>
<td>11</td>
<td>1</td>
<td>12</td>
<td>15.4%</td>
<td>0.997</td>
<td>0.28–6.74</td>
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<tr>
<td>Microadenoma</td>
<td>17</td>
<td>15</td>
<td>2</td>
<td>0</td>
<td>2</td>
<td>11.8%</td>
<td>0.997</td>
<td>0.15–3.63</td>
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<tr>
<td><strong>Intraoperative CSF leak</strong></td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>19</td>
<td>12</td>
<td>5</td>
<td>2</td>
<td>7</td>
<td>36.8%</td>
<td>0.021</td>
<td>1.38–13.07</td>
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<tr>
<td>No</td>
<td>91</td>
<td>80</td>
<td>10</td>
<td>1</td>
<td>11</td>
<td>12.1%</td>
<td>0.021</td>
<td>0.08–0.73</td>
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<tr>
<td><strong>Postoperative CSF leak</strong></td>
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<td></td>
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<tr>
<td>Yes</td>
<td>11</td>
<td>10</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>9.1%</td>
<td>0.797</td>
<td>0.06–4.02</td>
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<tr>
<td>No</td>
<td>99</td>
<td>82</td>
<td>14</td>
<td>3</td>
<td>17</td>
<td>17.2%</td>
<td>0.797</td>
<td>0.25–17.3</td>
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<td><strong>Previous pituitary resection</strong></td>
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<td></td>
<td></td>
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<td>Previous nonendoscopic transsphenoidal</td>
<td>14</td>
<td>8</td>
<td>4</td>
<td>2</td>
<td>6</td>
<td>42.9%</td>
<td>0.01</td>
<td>1.55–17.77</td>
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<tr>
<td>Previous endoscopic transsphenoidal</td>
<td>5</td>
<td>5</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0%</td>
<td>0.984</td>
<td>0.02–8.12</td>
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<tr>
<td>Previous craniotomy</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0%</td>
<td>0.519</td>
<td>0.07–42.07</td>
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<tr>
<td>All previous</td>
<td>20</td>
<td>14</td>
<td>4</td>
<td>2</td>
<td>6</td>
<td>30%</td>
<td>0.137</td>
<td>0.90–8.65</td>
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<tr>
<td>No previous</td>
<td>90</td>
<td>78</td>
<td>11</td>
<td>1</td>
<td>12</td>
<td>13.3%</td>
<td>0.137</td>
<td>0.12–4.12</td>
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<tr>
<td><strong>Apoplexy</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>5</td>
<td>5</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0%</td>
<td>0.984</td>
<td>0.02–8.12</td>
</tr>
<tr>
<td>No</td>
<td>105</td>
<td>87</td>
<td>15</td>
<td>3</td>
<td>18</td>
<td>17.1%</td>
<td>0.984</td>
<td>0.12–43.92</td>
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<tr>
<td><strong>Tumor types</strong></td>
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<tr>
<td>Nonfunctioning adenoma</td>
<td>61</td>
<td>52</td>
<td>9</td>
<td>8</td>
<td>1</td>
<td>14.8%</td>
<td>0.52</td>
<td>0.27–1.67</td>
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<tr>
<td>GH-secreting</td>
<td>15</td>
<td>13</td>
<td>2</td>
<td>2</td>
<td>0</td>
<td>13.3%</td>
<td>0.70</td>
<td>0.12–2.68</td>
</tr>
<tr>
<td>RCC</td>
<td>12</td>
<td>6</td>
<td>6</td>
<td>4</td>
<td>2</td>
<td>50%</td>
<td>0.003</td>
<td>2.0–25.8</td>
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<td>ACTH-secreting</td>
<td>9</td>
<td>8</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>11.1%</td>
<td>0.75</td>
<td>0.19–4.6</td>
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<td>Prolactinoma</td>
<td>9</td>
<td>9</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0%</td>
<td>0.36</td>
<td>0.01–3.1</td>
</tr>
<tr>
<td>Craniopharyngioma</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0%</td>
<td>0.55</td>
<td>0.04–17.5</td>
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<tr>
<td>Chordoma</td>
<td>2 (1.8%)</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0%</td>
<td>0.55</td>
<td>0.04–17.5</td>
</tr>
<tr>
<td>FAS-secreting</td>
<td>1 (0.9%)</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0%</td>
<td>0.48</td>
<td>0.05–34.7</td>
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<td><strong>Total</strong></td>
<td>110</td>
<td>92</td>
<td>18</td>
<td>15</td>
<td>3</td>
<td>19.2%</td>
<td></td>
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</tr>
</tbody>
</table>

Risk factors for DI

- Na > 145 mmol/L in the first 5 days post-op → increased risk of permanent DI
  - A single serum Na of > 145 → 23.3% risk of permanent DI
  - Sensitivity 87.5%, specificity 83.5%

- 4 out of 96 (0.04%) of pts with Na < 145 mmol/L developed transient DI
  - NPV 99.5%

Summary

• Pituitary hyperplasia is a physiologic phenomenon that occurs in adolescent/young adult females
  ▫ Suggests normal variations in size, which persist over time
  ▫ Characteristics that are not supportive of benign etiology include: AbL MRI, AnL labs, clinical sx
• Risk factors for DI: Na > 145 mmol/L, previous non-endoscopic pituitary surgery, RCC pathology
References