A single centre experience of managing a series of childhood macro/giant prolactinomas


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Introduction: Childhood prolactinomas often occur as aggressive macro (1-4cm) or giant (>4cm) tumours, with little consensus regarding timing of different therapies

Aim: To highlight the phenotype and the outcome of childhood macroadenomas

Subjects & Method:
- Collected data from 10 children (<18 years) presenting to our centre between 2009-2016 with hyperprolactinaemia due to macro/giant prolactinomas.

Results:
- Median age at diagnosis was 13.9 years (11.6-16.3).
- Median duration of symptoms prior to diagnosis in months was 24 (1-84).
- Presenting symptoms included headaches (10/10), visual deficit (5/10) and endocrinological signs (8/10).
- Positive family history identified in 4/10 cases. One proved heterogeneous for an MEN1 mutation. None harboured an AIP mutation.
- At diagnosis, all children had 1-4 anterior pituitary hormone deficiencies.

Figure 1. MRI slices for each case demonstrating extensive invasion to surrounding structures.

Table 2. Effects of treatment on 10 patients with macroadenoma

<table>
<thead>
<tr>
<th>Therapy</th>
<th>CBG mg/wk</th>
<th>Initial/Max</th>
<th>Pituitary hormone deficiencies at diagnosis</th>
<th>Vision at diagnosis</th>
<th>Vision post Rx</th>
<th>PRL basal mL/L</th>
<th>PRL nadir mL/L</th>
<th>% Tumour size change post Rx</th>
<th>Duration Rx</th>
<th>Genetics</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. CAB</td>
<td>0.5/7.5</td>
<td>Normal fields and VA</td>
<td>Normal fields and VA</td>
<td>44,782</td>
<td>7,756</td>
<td>24.3</td>
<td>37</td>
<td>36</td>
<td>Negative</td>
<td></td>
</tr>
<tr>
<td>2. CAB</td>
<td>2.5/10.5</td>
<td>Left homonymous hemianopia; bilateral papilloedema</td>
<td>Registered blind with VA R 6/90</td>
<td>321,498</td>
<td>14,535</td>
<td>36</td>
<td>Negative</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. CAB</td>
<td>1/1</td>
<td>Bi-temporal upper quadrant papilloedema</td>
<td>Pathfy blind loss on VA L 5/18</td>
<td>9,176</td>
<td>572</td>
<td>70</td>
<td>Negative</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. CAB</td>
<td>0.25/7</td>
<td>Bi-temporal hemianopia; small L superior quadrant papilloedema</td>
<td>Registered blind</td>
<td>27,624</td>
<td>2,268</td>
<td>70</td>
<td>Negative</td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

Conclusions:
- Cabergoline should be the first-line treatment in childhood-onset macroadenomas.
- Dose-escalation along with prolonged administration may be necessary for achieving a delayed response and controlling disease, but needs monitoring for side effects.
- In resistant disease, surgery increases endocrine deficiencies and constitutes the treatment of last resort.

*Occurring following: 1. surgical intervention; 2. occurring following second surgical intervention; PRL prolactin; CAB, Cabergoline; TCS, Trans-sphenoidal surgery; TCS, Trans-sphenoidal surgery; RT, radiation therapy; PBRT, Proton Beam Radiotherapy; Go, Gonadotrophins; THY, Thyroid Stimulating Hormone; GH, Growth Hormone; AVP, Arginine Vasopressin; VA, visual acuity; L, left; R, right; GH, Growth hormone; ACTH, Adrenocorticotropic hormone; BT, bitemporal; CF, counting fingers; NPL, No Perception of Light.