Craniopharyngioma

Michael Gottschalk, MD, PhD
University of California San Diego
Rady Children’s Hospital
Objectives

• Incidence
• Clinical Presentation
• Treatment Options
• Perioperative concerns
• Long-term endocrine outcome
Craniopharyngioma

Slow-growing, epithelial-squamous, calcified cystic tumor arising from remnants of the craniopharyngeal duct and/or Rathke cleft and occupying the (supra)sellar region
Pathology

• Controversial hypotheses on the embryonal origin:
  – originating from ectodermal remnants of Rathke’s pouch
  – originating from residual embryonal epithelium of the anterior pituitary and of the anterior infundibulum.

• Two histological variants:
  – adamantinuous (ACF), the typical pediatric form,
  – squamous-papillary form (PCF) observed in adults
ACFs arise from neoplastic transformation of embryonic squamous cell nests of the involuted craniopharyngeal duct.

During the process of proliferation and rotation of the cells of Rathke’s pouch leading to the formation of adenohypophysis, cell remnants of the craniopharyngeal duct are spread through the intrasellar and suprasellar region.
Epidemiology

• Incidence is 0.5–2 cases per million persons per year
• Distribution by age is bimodal, with peak incidence in children aged 5-14 years and older adults aged 65-74 years.
• 30–50% of all cases become apparent in childhood and adolescence
• Represent 6-10% of all childhood intracranial tumors.
• Equal sex ratio
Clinical Presentation

• Visual impairment (62–84%)
• Endocrine deficits (52–87%)
  – Growth hormone (75%)
  – Gonadotropins (40%)
  – Adrenocorticotropic hormone (ACTH) (25%)
  – Thyroid-stimulating hormone (TSH) (25%)
  – Antidiuretic hormone (ADH) (17%)
Local Extension

- **Anteriorly** into the prechiasmatic cistern and subfrontal spaces
- **Posteriorly** into the prepontine and interpeduncular cisterns, cerebellopontine angle, third ventricle, posterior fossa, and foramen magnum
- **Laterally** toward the subtemporal spaces
Clinical Presentation

Tumor infiltration into the hypothalamus

Disturbances of:

- memory
- attention
- impulse control
- motivation
- socialization
Clinical Presentation

• Frontal lobe function and integrative activities
  – (inability to withstand frustration, unmotivated anger, emotional lability or cognitive inflexibility)

• Hypothalamic dysfunction
  – (appetite perturbation, frustration at food restriction, uncontrolled violence)
Imagining

• Cystic tumor of the intra- and/or suprasellar region
• CT may be better at revealing calcification, which is found in approximately 90% of tumors
• Solid tumor portions and cyst membranes appear isointense in T1-weighted images, often with a mildly heterogeneous structure
• The most common localization is suprasellar, with an intrasellar portion. 20% of tumors are exclusively suprasellar, 5% exclusively intrasellar
Initial Treatment

Surgery

• The operative approach is generally dictated by the localization and extent of the craniopharyngioma

• A right frontotemporal approach is standard, but purely intrasellar tumors can be operated on by the trans-sphenoidal route.

• Craniopharyngioma arising in childhood usually extend to the suprasellar area and must be removed through a transcranial approach
Perioperative Hormone Deficiencies

Loss of hypothalamic and/or pituitary gland function

Serum levels of pituitary hormones are not a reliable measure of pituitary deficiency
CRF-ACTH-Cortisol
- Diurnal variation

GHRH-GH-IGF-1
- Prepubertal child
  - Nocturnal secretion

TRH-TSH-T4
- Change in glycosylation of TSH alters its bioactivity and immunogenicity

GnRH-LH/FSH-Steroid
- Pubertal stage dependency

Antidiuretic Hormone
- Short ½ life
- Proteolytic degradation
Perioperative Hormone Deficiencies

• Critical hormone deficiencies:
  – Cortisol
    • Often pretreated with decadron
  – Thyroid hormone
  – Antidiuretic hormone
    • Preoperative 17%
    • Postoperative 80-90%
## Long-Term Outcome

<table>
<thead>
<tr>
<th>Condition</th>
<th>S (n = 36)</th>
<th>S + RT (n = 43)</th>
<th>P-value&lt;sup&gt;a&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>Endocrinopathies</td>
<td></td>
<td></td>
<td>0.06</td>
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<tr>
<td>APHD + DI</td>
<td>29 (80.6)</td>
<td>28 (66.7)</td>
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</tr>
<tr>
<td>APHD only</td>
<td>2 (5.6)</td>
<td>10 (23.8)</td>
<td></td>
</tr>
<tr>
<td>DI only</td>
<td>2 (5.6)</td>
<td>0</td>
<td></td>
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<tr>
<td>Visual deficits</td>
<td>20 (56)</td>
<td>23 (53)</td>
<td>1.00</td>
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<tr>
<td>Blindness</td>
<td>4 (11)</td>
<td>2 (5)</td>
<td>0.40</td>
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<tr>
<td>Obesity</td>
<td>17 (47)</td>
<td>24 (56)</td>
<td>0.50</td>
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<tr>
<td>Seizure disorder</td>
<td>5 (14)</td>
<td>4 (9)</td>
<td>0.72</td>
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<td>Moya-Moya Vasculopathy</td>
<td>1 (3)</td>
<td>4 (9)</td>
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<tr>
<td>Second tumor</td>
<td>0</td>
<td>3 (7)</td>
<td>0.24</td>
</tr>
</tbody>
</table>

S, surgery; RT, radiation therapy; APHD, anterior pituitary hormone deficits; DI, diabetes insipidus. <sup>a</sup>P-value for Fisher’s exact test.