A case series of craniopharyngioma patients in the Puerto Rico Medical Center, 2004-2014: epidemiology and management analysis

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Craniopharyngioma in Puerto Rico, (USA)

Reason for the Study

- Limited literature worldwide
- No literature for PR
- Epidemiological data for PR is often considered equivalent to that of the mainland USA
- There is no consensus in the management of CP
Methods

- Records were identified using 2012 ICD-9-CM Diagnosis Code 237.0 for Neoplasm of Uncertain Behavior of the Pituitary Gland or Craniopharyngeal Duct.

- 64 records were screened for criteria:
  - Diagnosed and treated at Puerto Rico Medical Center during 2004-2014 (University Adult’s Hospital, University Pediatric Hospital, UPR School of Medicine OPD Clinics)
  - Pathology report confirming diagnosis of Craniopharyngioma

- 33 records were included: 20 adult and 13 pediatric

- 31 records excluded due to diagnostic uncertainty, outlier time frames, incomplete records
Caniopharyngioma in PR: Recovered Variables

- Gender
- Town of residence
- Age at diagnosis
- Date of Diagnosis
- Tumor location
- Clinical manifestations
- Initial treatment
- Method of excision
- Recurrence
- Date of last contact
- Status at last contact
Incidence

Annual incidence data of craniopharyngioma, 2004-2014, according to gender*

<table>
<thead>
<tr>
<th>Year</th>
<th>Number of new diagnoses</th>
<th>Male</th>
<th>Female</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>2004</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>0.523</td>
</tr>
<tr>
<td>2005</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0.262</td>
</tr>
<tr>
<td>2006</td>
<td>3</td>
<td>1</td>
<td>2</td>
<td>0.788</td>
</tr>
<tr>
<td>2007</td>
<td>4</td>
<td>2</td>
<td>2</td>
<td>1.057</td>
</tr>
<tr>
<td>2008</td>
<td>7</td>
<td>2</td>
<td>5</td>
<td>1.861</td>
</tr>
<tr>
<td>2009</td>
<td>4</td>
<td>2</td>
<td>2</td>
<td>1.069</td>
</tr>
<tr>
<td>2010</td>
<td>5</td>
<td>3</td>
<td>2</td>
<td>1.344</td>
</tr>
<tr>
<td>2011</td>
<td>3</td>
<td>1</td>
<td>2</td>
<td>0.815</td>
</tr>
<tr>
<td>2012</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0.275</td>
</tr>
<tr>
<td>2013</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0.835</td>
</tr>
<tr>
<td>2014</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>33</td>
<td>15</td>
<td>18</td>
<td>0.802</td>
</tr>
</tbody>
</table>

* Incidence rates are per 1,000,000 person-years.
† Total incidence rate is an averaged incidence using a mid-point population estimate.

Population Age One Year and Over

Incidence Age-specific

n = 135 cases
Clinical Presentation

Presenting symptoms in the pediatric and adult population diagnosed with craniopharyngioma, according to descending frequency, 2004-2014

<table>
<thead>
<tr>
<th>Presenting Symptoms</th>
<th>Pediatric</th>
<th>Adult</th>
</tr>
</thead>
<tbody>
<tr>
<td>92% Headache</td>
<td></td>
<td>Visual disturbance 85%</td>
</tr>
<tr>
<td>85% Visual disturbance</td>
<td></td>
<td>Headache 60%</td>
</tr>
<tr>
<td>38% Nausea/Vomit</td>
<td></td>
<td>Cognitive impairment 25%</td>
</tr>
<tr>
<td>23% Dizziness</td>
<td></td>
<td>Dizziness 15%</td>
</tr>
<tr>
<td>15% Weight loss</td>
<td></td>
<td>Ataxia 15%</td>
</tr>
<tr>
<td>15% Seizure</td>
<td></td>
<td>Extremity weakness 15%</td>
</tr>
<tr>
<td>15% Personality changes</td>
<td></td>
<td>Nausea/Vomit 10%</td>
</tr>
<tr>
<td>7.7% Ataxia</td>
<td></td>
<td>Endocrine disturbance 10%</td>
</tr>
<tr>
<td>7.7% Involuntary movement</td>
<td></td>
<td>Diplopia 10%</td>
</tr>
<tr>
<td>7.7% Polydipsia &amp; Polyuria</td>
<td></td>
<td>Amenorrhea 10%</td>
</tr>
</tbody>
</table>
Management

- Most common tumor location:
  - Suprasellar (40% adult - 46.2% pediatric)

- Most common surgical resection approaches:
  - Frontal (45% adult - 38.5% pediatric)
  - Frontotemporal (35% adult - 30.8% pediatric)
  - Transsphenoidal (10% adult - 15.5% pediatric)

- Most common surgical complication:
  - Diabetes insipidus (45% adult - 84.6% pediatric)
  - Panhypopituitarism (61.5% pediatric)
Recurrences

- Surgical resections:
  - Adults: 85% total - 15% subtotal
  - Pediatric: 31% total - 31% subtotal - 38% unspecified

- Recurrence rate:
  - 45.6% (45% adult - 46.2% pediatric)
  - 53.3% of cases that recurred developed 2nd recurrence
  - 75% of cases with 2nd recurrence developed additional recurrences
Survival: Recurrence-free

- Average follow-up time: (47.6 months adult - 64.3 months pediatric)
- Survival rates: Adult 87.4% 3-year; 79.5% 5-year; 59.6% 7.5-year
Conclusions

- Incidence in pediatric population peaked at 0-4 yo versus 10-14 reported in literature.
- Incidence peak at 55-64yo versus >65yo reported in literature.
- Even with adjuvant radiotherapy, tumor recurrence was a concern, with 45.6% of patients presenting residual or recurrent tumor after treatment.
- Future studies could serve to:
  - further understand particularities in PR CP patients;
  - consider a pathology revisions to characterize this variable.
Thank you