Cerebellopontine angle tumors in children

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Introduction

- Posterior fossa tumors is common in children: the fourth ventricle, cerebellar vermis and brain stem, or in the cerebellar hemispheres
- The cerebellopontine angle (CPA) is a rare site and less than 10% of posterior fossa tumors in childhood were located in CPA
- A plethora of tumors can occur in the CPA and CMF, which derive from neuroglial tissues, cranial nerve sheath, meninges, and embryonic remnants
- In the adult: vestibular schwannoma, meningioma, and epidermoid.
- In children: gliomas
Methods

• From January 2014 to May 2016, 34 children younger than 16 years old with CPA tumor were operated.
• All had histologically verified CPA/CMF tumors and underwent radical tumor resection through craniotomy
• The average age was 6.2 years (range: 1.5 to 14 years)
Presentation

- 9 females and 25 males
- Locations: predominantly in the CPA: 18; both in the CPA and the fourth ventricle: 16
- Hydrocephalus: 21/34 (62%)
- Cranial nerve dysfunctions:
  - Hearing loss: 3
  - Dysphagia/dysphonia: 3
  - Facial weakness: 2
- Gait disturbances: 5
- Hemiparesis: 2
- All were evaluated with CT and MRI
- No NF2 in this group
Surgery

- Hydrocephalus: 21/34 (62%)
  - VP: 5/21
  - OMMAYA: 4/21
  - external ventricular drainage: 4/21

- Approach
  - Midline craniotomy: 14
  - Hockeystick incision with midline & lateral craniotomy: 12
  - Retrosigmoid craniotomy: 5
  - Far lateral approach: 3
Results

• Tumor resection: 36 cases (2 relapsed)
  – gross total resection: 32
  – subtotal resection: 4

• tumor origin:
  – lateral recess of the fourth ventricle: 21
  – ventral cerebellar hemisphere: 2
  – cerebellar peduncle: 2
  – brain stem: 4
  – embryonal nests: 2
  – cranial nerves and meninges: 3
### Pathology

#### Malignant: benign = 2.4:1

- **21 cases of ependymomas (II & III) (62%):**
  - Anaplastic ependymoma (III): 15 (44%)
  - Ependymoma (II): 6 (18%)
  - <7yr: 13 case (anaplastic: 11 cases, 85%)

<table>
<thead>
<tr>
<th>Malignant</th>
<th>Benign</th>
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<tbody>
<tr>
<td>ependymomas</td>
<td>glioblastoma</td>
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<tr>
<td>21</td>
<td>1</td>
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<td>24 (70%)</td>
<td>10 (30%)</td>
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Surgical complications

- no mortalities
- vocal cord palsy or dysphagia: 9 (3 were noted preoperatively)
  - Nasogastric tube: 6 (removed in 2-9 w)
  - 2 with a combination of tracheotomy (removed in 8-9 w)
- facial weakness: 3 (2 preop)
Outcome

• For Malignant tumors (24 cases)
  – Chemotherapy: 14
  – Radiotherapy: 15

• follow-up (6-29mo): 5 patients with anaplastic ependymoma relapsed, 2 reop without recurrence; 2 died without op and the last one continue chemotherapy.
medulloblastoma, 1yr, facial weakness 2mo
M, 14yr, vestibular schwannoma

3mo postOP
M, 10yr, Anaplastic ganglioglioma

1.9y postOP
F, 8y, ganglioglioma

1y postOP
F, 11y, meningiomas

1.1y postOP
M, 14Y, epidermoids
M, 1.5Y, inflammatory granuloma
M, 8Y, midbrain pons tumor
Kawase approach, GTR. pilocytic astrocytomas
F, 9y, medulla oblongata pilocytic astrocytomas
M, 3y, Anaplastic ependymoma
M, 13y, ependymomas
6 mo after OP
2 mo after the second OP, Anaplastic ependymoma
M, 7y, Anaplastic ependymoma
M, 2y, Anaplastic ependymoma
M, 5Y, Anaplastic ependymoma

9mo postOP
Conclusion

- Various of tumor types occur in childhood at the CPA/CMF and our experience indicated that 70% tumor are malignant and 62% were ependymomas (Anaplastic ependymoma: 44%).
- If the lower cranial nerves were protected well and carefully dissected from the tumors during the operation, most patients can avoid tracheostomy.
- The follow-up time in our group is short, so the tumor recurrence and survival rate need further observed.
THANK YOU