Giant Extra-Axial Cerebellopontine Angle Ependymoma with Sub-Axial Cervical Extension: Infratentorial Ependymomas as the ‘Many-Faced God’ in the ‘Game of Neurosurgery’

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Disclosures

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Introduction

- *Ependymomas are glial tumours* that typically arise from ependymal lining of ventricles and central canal

- Although most are infra-tentorial, it is uncommon for Infratentorial ependymoma to arise away from fourth ventricle, such as in the cerebellopontine (CP) angle

- We report findings of one such patient with a *giant extra-axial ependymoma with epicentre in right CP angle*, and further review literature of all such reported cases
Case description

• 37-year old gentleman with occipital headache and pain at the back of neck for 4 months, with features of raised intracranial pressure

• Had undergone ventriculoperitoneal shunt at another hospital

• On examination, he had papilloedema and bilateral cerebellar signs, including nystagmus and appendicular ataxia

• No other cranial nerve deficits
• **MRI** - Single, well-defined, extra-axial lesion, originating in the right CP angle and extending from tentorium superiorly to upper body of C4 inferiorly; occupying perimesencephalic, prepontine and cerebellomedullary cisterns

• A working diagnosis of Schwannoma - based on preoperative radiology

• Underwent *right suboccipital craniotomy with C1-C3 laminectomy and near-total tumour excision*

• Tumour was yellowish, soft-to-firm, moderately vascular, extra-axial, and had a well-defined plane separating it from the cord parenchyma and cerebellum
Figure 1: A) Pre-operative T2 sagittal MR: giant extraaxial heterointense tumor in CP angle extending from tentorium superiorly to upper body of C4 body inferiorly. 
B) T2 axial MR: Extraaxial mass in right CP angle with compression and shifting of fourth ventricle. No extension into IAC is seen. 
C) and D) axial and sagittal T1 contrast MR: ill-defined heterogenous contrast enhancement. 
E) and F) Intra-operative microscopic image showing the cervical extension of the tumor with well-defined plane from the cord parenchyma and the accessory nerve adherent to the tumor (black arrow).

Figure 2: A) and B) Post-operative T1 contrast axial and sagittal MR showing gross total excision of the tumor. 
C) H&E at 10x: tumor cells with round to oval nuclei, evenly dispersed granular chromatin in fibrillary background. Few perivascular pseudo rosettes seen. 
D) Immunohistochemistry (IHC) for EMA at 40X magnification with dot and ring-like positivity. 
E) IHC for S100 at 10x: positive. 
F) Microscopic image at 10x showing pseudorosettes marked with an arrow.
• Postoperative course was uneventful with no new neurological deficits

• **Histopathological examination** - Cellular tumour with extensive fibrillary background, microcytic spaces, vague fascicles, and occasional cells with pleomorphic nuclei

• Immunopositivity for S100, vimentin, and para nuclear dot-like positivity for EMA

• Findings consistent with a diagnosis of *Ependymoma*

• Planned for adjuvant radiotherapy

• Patient remains asymptomatic at 9 months follow-up. CEMRI at 6 months post-surgery showed a 2x1 cm residual in cerebellomedulary cistern
Discussion

• Pure extra-axial CP angle ependymomas are extremely rare - have no connection to ventricle/ependymal surface

• Believed to arise from embryological ependymal cell rests present in extra-ventricular regions (like CP angle, falx, other extra/intracranial sites)

• Literature review revealed 7 reported cases (Table 1) of extra-axial ependymomas in or originating from CP angle - all cases did not have typical features of CP angle syndrome (such as hearing loss)

• Mean age of presentation is higher (46.25 years) with extra-axial CP angle ependymomas, as compared to typical 4th ventricle ependymomas (23 years)
<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Sex/Age</th>
<th>Clinical manifestations</th>
<th>Tumor location &amp; extension</th>
<th>Tumor size</th>
<th>Adjuvant therapy</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cosgrove et al(^3), 1985</td>
<td>78/M</td>
<td>Headache, cerebellar signs</td>
<td>CPA</td>
<td>3 x 4 cm</td>
<td>No</td>
<td>36 months, tumor-free</td>
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<tr>
<td>Fukui et al(^4), 1997</td>
<td>66/M</td>
<td>Headache, cerebellar signs</td>
<td>CPA</td>
<td>2.5 x 1.5 cm</td>
<td>RT</td>
<td>30 months, tumor-free</td>
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<tr>
<td>Ueyama et al(^6), 1997</td>
<td>38/M</td>
<td>V, VI, VIII, cerebellar signs</td>
<td>CPA, IAC, and pineal region</td>
<td>NA</td>
<td>RT</td>
<td>6 weeks, death</td>
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<tr>
<td>Donich et al(^7), 1999</td>
<td>22/F</td>
<td>Headache, facial weakness</td>
<td>CPA and cavernous sinus</td>
<td>3 x 4 cm</td>
<td>RT</td>
<td>18 months, tumor-free</td>
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<tr>
<td>Torun et al(^8), 2004</td>
<td>31/M</td>
<td>Headache, facial paresis</td>
<td>CPA</td>
<td>4.5 x 3.5 x 3.5 cm</td>
<td>RT</td>
<td>NA</td>
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<tr>
<td>Kasliwal et al(^9), 2007</td>
<td>50/F</td>
<td>Headache, vomiting</td>
<td>CPA</td>
<td>NA</td>
<td>RT</td>
<td>NA</td>
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<tr>
<td>Gill et al(^10), 2014</td>
<td>48/M</td>
<td>Headache, vomiting</td>
<td>CPA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Present case, 2019</td>
<td>37/M</td>
<td>Headache, vomiting</td>
<td>CPA, cervical region</td>
<td>9.4 x 5.5 x 6 cm</td>
<td>RT</td>
<td>3 months</td>
</tr>
</tbody>
</table>

**Table 1:** Summary of eight cases of pure extra-axial CPA ependymomas till date
• Ependymomas are WHO Grade II or III tumours; however, all reported extra-axial CP angle ependymomas have been of a lower grade (Grade II)

• These patients have a better prognosis than typical fourth ventricle ependymomas - tumour are low grade and possible to safely resect them completely

• Patients with sub-total or near total resection require radiotherapy

• To conclude, ependymoma should be suspected in ta CP angle mass with no hearing loss in absence of typical radiological features of meningioma/epidermoid

• Due to their plastic nature, they can attain a massive size at presentation and can extend to many other areas, including the cervical spine