Case Report

Extra-Axial Ependymoma Presenting as a Cerebellopontine Angle Mass: A Rare Case Report

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Abstract:
Posterior fossa ependymomas characteristically originate from the ependymal cells in the fourth ventricle and present as midline tumors, but many ependymomas arise from the lateral medullary velum and extend into the cerebellopontine (CP) angle cistern. By maintaining a high index of suspicion and through careful analysis of preoperative MRI images, the surgeon should be able to distinguish exophytic intraaxial lesions from extra-axial masses in the vast majority of cases of cerebellopontine angle. We present a rare case of isolated extra-axial ependymomas at the cerebellopontine angle with features of raised intracranial pressure and signs of cerebellar lesion.

Key words: cerebellopontine angle, extra-axial, ependymoma.


Introduction:
Posterior fossa ependymomas are common in pediatric age group, but uncommon in adults.1,2 They usually arise in the fourth ventricle from the roof, floor, lateral medullary velum or its lateral recesses and present as midline tumors.3,4 Although extension of this tumor into the cerebellopontine angle (CPA) and subarachnoid space through the foramina of Luschka or Magendie is well described, a primary extra-axial cerebellopontine angle location of a posterior fossa ependymoma is distinctly uncommon.5 Vestibular schwannomas and meningiomas combine for 80% to 95% of CPA lesions, with epidermoid cysts being the next most common.6 Only rarely do ependymomas present as primary CPA lesions and, for this reason, these tumors are often initially misdiagnosed. As the optimal management strategy for treating parenchymal tumors differs substantially from that for extra-axial lesions, it is essential that the surgeon have a preoperative awareness of the lesion’s nature before embarking on a surgical endeavor.7

Case Report:
A 32-year-old female presented with a 2-months history of worsening headaches, nausea, vomiting, visual disturbance, lightheadedness, and progressive ataxia. There was no hearing loss or any facial hypo or hyperesthesia. On examination visual acuity was 6/18 in both eyes as measured with Snellen’s chart with fundus examination showing bilateral papilledema. Horizontal jerk nystagmus on lateral gaze and ataxia were present with no clinical evidence of seventh, eighth or lower cranial nerve involvement.

Magnetic resonance imaging (MRI) revealed a well-defined intensely enhancing homogeneous extra-axial mass lesion (3.0 x 3.3 x 2.8 cms) within the enlarged right cerebellopontine angle cistern (Figure 1B), exerting mild perilesional edema and mass effect over cerebellum, brain stem and fourth ventricle (Figure 1C). The mass is almost homogeneously isointense...
on short TR (Figure 1A) and hyperintense on long TR images with central foci of calcifications (Figure 1C). There is no intracanalicular extension of the mass. The cranial nerves VII & VIII are outlined separately from the mass with normal diameter of the right internal acoustic canal (IAC). There is fourth ventricle outlet foraminolobstruction with quadriventricular enlargement and periventricular hyperintensity (Figure 1D).

A diagnosis of cerebellopontine angle tumor most likely a meningioma, acoustic neuroma or lower cranial nerve schwannoma was considered.

Initially, left sided ventriculoperitoneal shunting was performed followed by right sided retromastoid retrosigmoid suboccipital approach for removal of the tumor after 3 weeks. Intraoperatively there was a totally extra-axial moderately vascular, soft, suctionable tumor with areas of necrosis and hemorrhages having a well demarcated plane with the cerebellum and no obvious connection with the fourth ventricle. Gross total removal of the tumor was achieved (Figure 2).

The histopathology revealed papillary ependymoma, World Health Organization (WHO) grade II8 showing perivascular pseudorosettes with a small component of rare foci of few bizarre cells with hyperchromatic nuclei and the number of mitoses were < 5 per 10 high power fields. Few areas of necrosis without palisading were encountered (Figure 3).

**Fig.-1:** Axial Magnetic resonance (MR) image (A) T1 weighted image showing a well-defined isoointense mass lesion in the right cerebellopontine angle (CPA), (B) Contrast image showing the heterogeneously enhancing lesion with cystic areas compressing the brain stem and fourth ventricle, (C) T2-weighted image showing the extra-axial hyperintense lesion in right CPA with mild perilesional oedema, and (D) FLAIR image showing hydrocephalus with periventricular hyperintensity.
Rest of the postoperative course was uneventful. Her facial nerve and hearing have remained intact postoperatively and she has remained in good health since surgery without further treatment. The patient is awaiting adjuvant radiation treatment.

Discussion:
Ependymomas are glial tumors constituting approximately 5% to 6% of all intracranial tumors, most commonly located within the posterior fossa, classically originating from the ependymal cells of the fourth ventricle. It may extend into the cerebellopontine angle or subarachnoid space by means of an exophytic component through the foramina of Luschka and Magendie and can have both intraaxial and extraaxial components. Isolated extra-axial ependymoma at the cerebellopontine angle as described by us is very rare, particularly when no fourth ventricle involvement is identified. It has been suggested that deposition of heterotopic ependymal rests in surrounding tissue during fetal development...
Extra-Axial Ependymoma Presenting as a Cerebellopontine Angle Mass

Chowdhury D et al.

could give rise to extra-axial ependymomas. They assume a large size before becoming symptomatic. Initial presentation is with features of raised intracranial pressure (ICP) because of hydrocephalus. Involvement of cranial nerves and brain stem occurs at a later stage. Differential diagnosis of lesions at CPA includes common extra-axial lesions like acoustic neuromas, meningiomas. Less common lesions are epidermoid and other schwannomas as well as metastases, paragangliomas, and arachnoidal cysts. Intra-axial tumors in the area of the cerebellopontine angle include the medulloblastoma, astrocytoma, and the ependymoma.

The imaging appearance of this lesion is inconsistent with that of the more common CPA lesions, meningioma and schwannoma, as well with the less frequently encountered choroid plexus papilloma.

Schwannomas tend to be isointense to the brain on precontrast T1, have variable intensity on T2 based on cystic degeneration of the tumor, and enhance intensely with contrast. They also usually have a concentric growth around the component extending into the internal auditory canal.

Meningiomas tend to be isointense to brain on T1 and T2 sequences, brightly enhance with contrast, are dural based, and have a sessile growth pattern. They also may contain calcifications. Choroid plexus papillomas are typically isointense to hyperintense on T1 and T2 and show robust homogeneous enhancement. Internal calcification is present in about 20% of these tumors.

CPA ependymomas on the plain CT scan appear as large extra-axial masses in posterior fossa pushing the brain stem and fourth ventricle to the opposite side. Although no Magnetic resonance imaging features are pathognomonic of ependymoma, these generally appear as lobulated tumors which are hypointense on T1W images and hyper intense on T2W image, demonstrate irregular enhancement and are markedly heterogeneous due to calcification, hemorrhage, cystic components or necrosis. T1 and T2 sequences are required to exactly delineate the encasement of vessels and to appreciate the rotation and positioning of brain stem for correct surgical planning. Full spinal cord MRI is also extremely important to rule out spinal cord metastasis in ependymoma patients, though we found no such metastasis in our case. Ependymomas may be cystic or demonstrate areas of necrosis and hemorrhage. On histologic examination, pleomorphic cells with rare mitoses form perivascular pseudorosettes and occasional ependymal canals.

Surgical resection with subsequent radiotherapy is the primary treatment strategy for patients with ependymoma. Gross or near total excision offers the patient maximal chances of prolonged overall survival as well as progression-free survival. Adjuvant radiotherapy and chemotherapy have been used post operatively to prevent tumor recurrence. The efficacy of conventional chemotherapy for this disease remains uncertain; however, temozolomide appears to be a promising adjuvant therapeutic approach for multifocal anaplastic ependymoma following surgical resection. Proton radiotherapy although dosimetrically superior to photon radiotherapy is likely to become available after its clinical significance and population subset likely to benefit by its use is clearly defined. The role of neuraxis irradiation is limited to cases, which have proven metastasis at presentation.

Conclusion: Although primary cerebellopontine angle ependymoma is extremely rare and difficult to diagnosis, pre operative suspicion of ependymoma especially with a heterogeneous tumor in a patient with well-preserved hearing is important for surgical planning, given its more malignant nature and poorer prognosis compared with acoustic neuroma.

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Ethical approval: There is no ethics issue in this paper.

Reference:


