CASE REPORT

Posterior Fossa Ependymoma: Unusual Extension into the Internal Auditory Canal in a 32-year-old Woman

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ABSTRACT

Ependymomas are relatively rare glial neoplasms that arise from ependymal cells. The majority of cases (>75%) arise from the fourth ventricle in the posterior fossa. These tumours may expand locally, extend along subarachnoid spaces, and disseminate through the cerebrospinal fluid. Sometimes, they may extend into the cerebellopontine angle and mimic the clinicoradiological appearance of an extra-axial lesion. However, it is extremely unusual for such a lesion to extend into the internal auditory canal and mimic an acoustic neuroma. Such an unusual occurrence of a fourth ventricular ependymoma that extended into the right internal auditory canal through the porus acusticus is described in this case report. Also discussed are the imaging characteristics that could help distinguish between extra-axial lesions of the cerebellopontine angle and exophytic brain tumours.

Key Words: Cerebellar neoplasms; Cerebellopontine angle; Ependymoma; Labyrinth/pathology

INTRODUCTION

Ependymomas are relatively rare glial neoplasms and account for fewer than 5% of all intracranial tumours in adults.1 They have a propensity to arise from the ventricular system, vestigial central canal of the spinal cord, filum terminale, and the white matter adjacent to the ventricular surface. The location depends on the cell of origin — that is, where the ependymal cell is. The majority of cases (>75%) arise from the fourth ventricle in the posterior fossa.2

Ependymomas may expand locally, extend along subarachnoid spaces, and disseminate through the cerebrospinal fluid. Sometimes, posterior fossa ependymomas may extend into the cerebellopontine (CP) angle through the foramen of Luschka,2 thereby mimicking the clinicoradiological appearance of an extra-axial lesion. An extra-axial origin of the ependymoma directly in the CP angle is also possible, although rare.3 However, it is extremely unusual for such a lesion to extend through the porus acusticus into the internal auditory canal (IAC). This report describes such an unusual occurrence of a fourth ventricular ependymoma, which extended into the right IAC through the porus acusticus, thereby closely simulating an acoustic neuroma.

Because only 2 such cases have been previously described in the literature and the optimal treatment strategy is not very well defined, it is imperative that a preoperative diagnosis of the lesion be made to guide to the surgical treatment. This report also discusses imaging characteristics that would help physicians distinguish between extra-axial lesions of the CP angle and exophytic brain tumours extending into the CP angle.

CASE REPORT

A 32-year-old woman presented to the Tata Memorial Hospital, Mumbai, India, in June 2000 with a 6-month history of diminished hearing in the right ear and a 3-month history of headache, vomiting, imbalance while walking, blurred vision, and slurred speech.

Clinical examination revealed that the patient had sensorineural deafness on the right side, right sixth and seventh nerve paresis, and right-sided cerebellar signs
of disease. Bilateral papilloedema was also evident during fundoscopy.

Plain and contrast-enhanced computed tomography (CT) of the brain revealed a large minimally enhancing hypodense lesion that involved the right CP angle and extended into the right cerebellar hemisphere. There was also an intraventricular component in the fourth ventricle that caused distortion and dilation of the third and lateral ventricles.

Magnetic resonance imaging (MRI) revealed a lobulated lesion that was hypointense on T1-weighted images, hyperintense on T2-weighted images, and involved the right CP angle and the right cerebellar hemisphere (Figure 1). The lesion had an intraventricular component in the fourth ventricle and extended into the right IAC, mildly distorting the fourth ventricle and resulting in hydrocephalus. No gadolinium contrast-enhanced MRI scans were available for review.

Thus, a clinicoradiological diagnosis of CP-angle acoustic neuroma was made. There were no detectable neurofibromatosis-associated features. The dura was opened through a right retromastoid suboccipital approach and the cerebellum was retracted medially to expose the tumour. The tumour was firm, was vascular, and had engulfed the seventh and eighth nerve complex. It was debulked from within and partially excised to leave behind a minor residual amount of tumour because the perioperative impression was of a malignant CP-angle tumour. Histopathological examination revealed that the tumour was an ependymoma. The patient subsequently received adjuvant radiotherapy to the tumour bed to a dose of 54 Gy in 30 fractions during 37 days with bilaterally opposing portals using 6-MV photons.

DISCUSSION

This case illustrates the fact that ependymomas can extend to anywhere in the subarachnoid space. It also underscores the often repeated fact that an exophytic brain tumour can closely mimic the clinicoradiological features of an extra-axial mass in the CP angle.

Gross total excision is the therapeutic goal for benign extra-axial CP-angle tumours such as acoustic neuromas and meningiomas. In contrast, maximal safe resection is performed for primary exophytic brain tumours, followed by adjuvant radiotherapy. Aggressive removal of such intrinsic lesions may result in devastating neurological dysfunction. Because there is substantial overlap in the clinical presentation of intra-axial and extra-axial masses within the CP angle, the primary means of preoperatively differentiating between the 2 tumour types lies in tumour imaging. Although an intra-axial lesion may show a similar pattern of signal intensity on an MRI scan (i.e., isointensity to hypointensity on T1-weighted images, and enhancement with gadolinium contrast agent and hyperintensity on T2-weighted images), few features can help distinguish between the 2 tumours.

The presence of calcium in a fourth ventricular tumour is highly suggestive of an ependymoma. Interface irregularity between the lesion and adjacent brain tissue strongly suggests intra-axial lesions, although it may theoretically occur with large and longstanding extra-axial lesions in which the tumour-brain interface becomes blurred because of encephalomalacia. Dilatation of the lateral recess of the fourth ventricle (foramen of Luschka) is an important distinctive finding in fourth ventricular ependymomas. Finally, the absence of IAC involvement can indicate a non-acoustic lesion of the CP angle. On the other hand, acoustic neuromas generally show homogeneous and marked contrast enhancement. The tumor in the patient in this report showed only minimal contrast enhancement during CT examination. Furthermore, despite IAC involvement, the
histology turned out to be that of an intra-axial ependymoma.

A literature search of MEDLINE, EMBASE, and Cancerlit identified only 2 similar case reports. CP-angle ependymoma with IAC enlargement and pineal extension was reported by Ueyama et al6 in a 38-year-old man with a posterior fossa ependymoma. MRI clearly demonstrated the extension from the CP angle to the pineal region through the right ambient cistern. CT using a bone algorithm revealed enlargement of the right IAC. The authors hypothesised that ependymoma could extend anywhere within the subarachnoid space along the pathway of least resistance. The second case was reported by Bonneville et al7 who described a 24-year-old woman with a left-sided CP-angle ependymoma with invasion of the IAC but without widening of the porus acousticus.

CONCLUSION
This case report illustrates that ependymomas have a potential to extend into the IAC and that preoperative imaging may help to distinguish between intra-axial and extra-axial lesions of the CP angle, thereby guiding the clinician to formulate the appropriate therapeutic protocol.

REFERENCES