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Case Report

Reappraisal of cerebellopontine angle medulloblastomas: Report of a fatal case and lessons learned

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ABSTRACT

CPA Medulloblastomas are rare in adults. We describe a case of medulloblastoma in the right CPA in a middle aged male patient who presented with short history and was preoperatively diagnosed as vestibular schwannoma. Intra-operatively it turned out to be a highly vascular and malignant lesion. Histopathology demonstrated medulloblastoma with high ki67 index. Patient was re-explored for hematoma evacuation and remained vegetative postoperatively. This is a unique case considering the older age at presentation, short duration of symptoms with atypical presentation of only 7th, 8th and lower cranial nerve involvement. MRI showed a lobulated heterogeneous extra axial right CPA mass lesion extending in the right acoustic meatus and causing mass effect and effacement of the fourth ventricle with heterogeneous enhancement post-gadolinium, suggestive of vestibular schwannoma. The aggressive behaviour of this tumor with short history has to prompt one to keep less common lesions like medulloblastomas as differential diagnosis in the CPA region. Though medulloblastomas are rare lesions in the CPA they have to be considered in the differential diagnosis based on atypical clinical behaviour, a suspicion of such diagnosis preoperatively can alter the treatment strategy, focusing on tumor embolization, safe surgical excision and adjuvant chemoradiation, aiming towards a better quality of life, in the post-operative period.

1. Introduction

There are a very few cases of cerebellopontine angle (CPA) medulloblastomas described in the literature till date. We describe a peculiar case of CPA tumor which was diagnosed as vestibular schwannoma preoperatively and turned out highly vascular and malignant intraoperatively, which was histopathologically confirmed as medulloblastoma. The incidence of medulloblastoma is high among pediatric age group accounting for 17% of all brain tumors in the age group 0–14 years and it is less than 1% in adult population [1]. In adults the most common CPA tumor is vestibular schwannoma accounting for 85–90% followed by meningioma and epidermoid. Others like primary cholesteatoma, arachnoid cyst, metastasis add on to the list [2]. Medulloblastomas are few in number and the literature is sparse.

2. Case report

A fifty year old male patient presented with atypical symptoms of a short duration of facial asymmetry, hearing loss on right side and a weak gag suggestive of lower cranial nerve involvement on right side without any signs of raised ICP. He was investigated for the same and MRI brain done showed a large heterogeneously enhancing mass in the right CPA extending in the porus acousticus and reaching up to the foramen magnum, without any hydrocephalus (Fig. 1). Based on these imaging findings, a preoperative diagnosis of vestibular schwannoma was made. Patient was taken up for right sided retromastoid sub-occipital craniotomy and excision of the tumor and intraoperatively the tumor was found to be highly vascular and non-suckable in nature. Frozen section was suggestive of atypical malignant lesion. Due to its vascular nature the tumor could only be partially decompressed and

Abbreviations: CPA, cerebellopontine angle; ICP, intracranial pressure; EVD, external ventricular drainage; GCS, Glasgow coma scale; PCA, posterior cerebral artery; GCP, granule cell precursor cells; EGL, external granular layer; H, headache; NV, nausea & vomiting; B/L PE, bilateral papilledema; HP, hemiparesis; HA, hemianaesthesia; PE, partial excision

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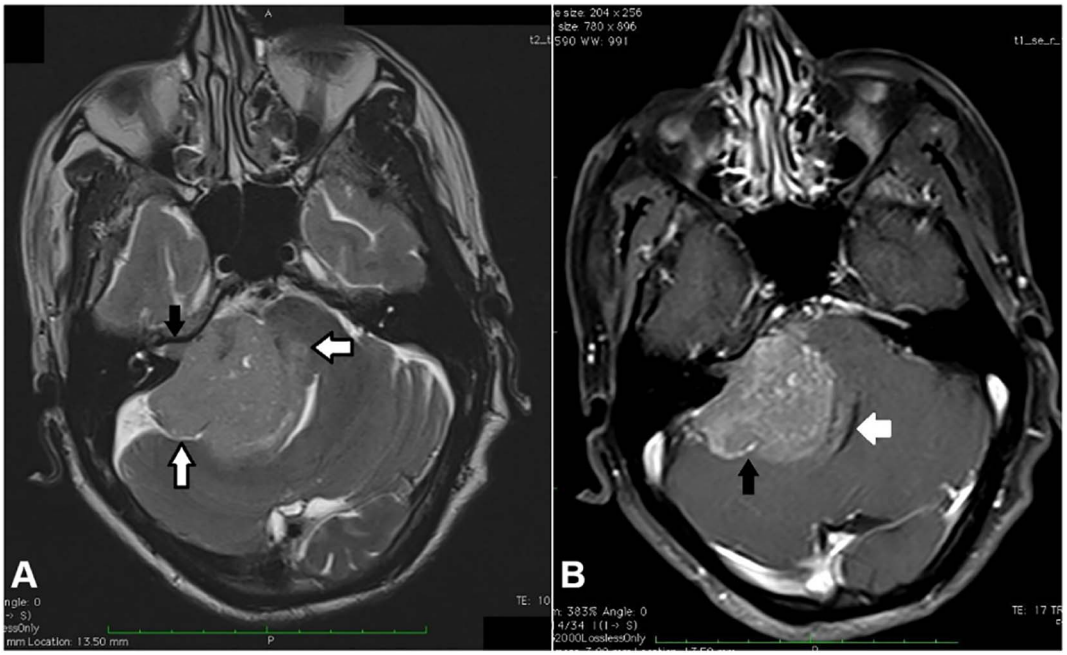

Fig. 1. A) T2 weighted axial MRI image (Te: 109 ms, TR: 5830 ms) showing a lobulated heterogeneous extra axial right CP angle mass lesion (white arrows) extending in the right acoustic meatus (black arrow) and causing mass effect and effacement of the fourth ventricle. B) Post-contrast (Gadolinium) T1 weighted images showing heterogeneous enhancement in the right CP angle mass lesion (black arrow) with mass effect and compression of the fourth ventricle (white arrow).

closed after hemostasis. In the immediate post-operative period patient developed bradycardia and a CT scan was performed which demonstrated tumor bed hematoma with developing hydrocephalus. He was taken up again for re exploration and insertion of an EVD (external ventricular drainage). After the procedure patient was put on ventilator. Subsequently patient had no improvement in his GCS and scans done later showed developing infarcts in the brainstem areas and PCA territories (Fig. 2). Later patient developed ventriculitis and deteriorated. The histopathology report showed classical type of medulloblastoma with Ki67 index of 20–25% (Fig. 3).

3. Discussion

Bailey and Cushing in 1925 were the first to describe medulloblastoma cerebelli, referring to a highly malignant small cell tumor of cerebellum in midline [3]. The WHO in 2007 has designated medulloblastoma as a distinct embryonal tumor, distinguishing it from other PNETs [3]. The granule cell precursor cells (gcp) form a secondary germinal zone called the external granular layer (EGL) during the development of cerebellum, with help of several signalling pathways like hedgehog and wnt and the disruption in this pathway results in the development of medulloblastoma [4]. Medulloblastomas in the CPA can be a result of spread from the midline cerebellum, the most common site of origin, its lateral extension through the foramen of lushka or direct exophytic growth from the surface EGL of cerebellum or pons [5]. Adult medulloblastomas arise from surface of cerebellum or pons and are laterally located intra-axially or in the tentorial region [6,7]. Extra-axial CPA medulloblastomas are rare and only few are reported in the past. The table describes CPA medulloblastomas described in the past, presentation, management and outcome comparing to the present case (Table 1).

Our patient is the oldest among the reported cases with rapid onset of symptoms of less than one month duration. Due to high vascularity the procedure was abandoned after subtotal excision and patient remained vegetative subsequently. Though the preoperative diagnosis was schwannoma biopsy revealed it to be a classical type of medulloblastoma. The short duration of symptoms and aggressive nature of the tumor goes in favour of histological diagnosis [8,9]. Though early involvement of VII, VIII goes in favour of schwannoma few cases with medulloblastomas have presented with these symptoms [10]. Our case is unique with early VII and VIII nerve involvement and lower cranial nerve involvement with no cerebellar signs. MRI findings in our case clearly showed the extra axial nature of the mass lesion and the extension into the acoustic meatus suggesting a diagnosis of vestibular schwannoma. Earlier cases have demonstrated both homogenous and heterogeneous findings in these tumors in the past [11]. Histologically desmoplastic variety is more common in adults and accounts for 20% of adult medulloblastomas [12]. They are seen in high frequency with Gorlin syndrome associated with mutation in SHH (sonic hedgehog) pathway and has a better prognosis than the classic type [13,14].

The aggressive behaviour of this tumor with short history has to prompt one to keep less common lesions like medulloblastomas as differential diagnosis in the CPA region. Medulloblastomas tend to metastasize through CSF and spinal canal being the most common site needs evaluation [15]. There was no such metastatic evaluation done in our patient.

Maximal surgical excision followed by craniospinal irradiation and chemotherapy is the mainstay in the management of both adult and pediatric medulloblastomas [16]. The tumor recurrence rate in adults is 50–60% and the median survival after recurrence is about 15 months [17].

4. Conclusion

Though medulloblastomas are rare lesions in the CPA they have to be considered in the differential diagnosis based on atypical clinical behaviour. A suspicion of such diagnosis preoperatively can alter the treatment strategy, focusing on tumor embolization, safe surgical excision and adjuvant chemoradiation, aiming towards a better quality of life, in the post-operative period.

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