

Medulloblastoma of the Cerebellopontine Angle in a Child: A Case Report and Review of the Literature

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Abstract

Cerebellopontine angle medulloblastoma is a very uncommon presentation of medulloblastoma, especially in children. We report such a case in a 14-year-old boy. Clinical onset by acute hearing loss and MRI features were, furthermore, unusual findings. The tumour had an homogeneous signal on T1 and FLAIR sequences a high-signal intensity on diffusion-weighted images with slightly restricted ADC value ($0.85 \times 10^3 \text{ mm}^2/\text{s}$) and enhanced slightly and homogeneously after contrast. A subtotal removal of the tumour, followed by entire neuroaxis irradiation and chemotherapy were achieved. MRI data of the literature reported cases were reviewed. Features on conventional MRI sequences do not allow differentiating CPA medulloblastoma from other rare tumour of this location. We point out that ADC value and Spectroscopy might be useful differentiating CPA tumours, especially from ependymoma in children.

Keywords: Medulloblastoma; Brain neoplasms; MRI; Cerebellopontine angle tumours; Diffusion; Children

Introduction

Medulloblastoma (MB) is a highly malignant neuroepithelial tumour of the posterior fossa classified WHO IV. MB is mainly a pediatric brain tumour that account for 10% of all intracranial neoplasms and 29% of all pediatric fossa tumours in children [1-3], whereas they represent less than 1% of CNS adult neoplasms. Most medulloblastomas (MBs) are located in the vermis in children and in the cerebellar hemispheres in adults [1-3].

Typical and atypical MRI features have been reported in MBs: most of them present with well-defined margins, iso- and hyperintensity signal respectively on T1 and T2-images, and marked enhancement on T1-postcontrast images. In rare cases they present with ill-defined margins, hypointensity signal on T2, mild or nodular enhancement [4-6].

Cerebellopontine angle (CPA) is an uncommon location and only 38 cases have been reported to date [7,8]. We report a further case of CPA MB in a child and we review the cases of the literature.

Case Report

A 14-year-old right-handed healthy boy who presented with a sudden right-sided hearing loss, vertigo and gait ataxia starting 8 months before admission. An audiogram performed 5 months after onset showed a marked right-sided hearing loss and CT scan showed a rounded, homogeneously hypodense CPA lesion, without enlargement of the internal auditory canal (IAC), enhancing after contrast administration. At admission, the neurological examination revealed gait ataxia and right hypoacusis. A brain stem auditory evoked response (BAER) showed no wave of the right ear and normal wave morpho of the left ear. MRI revealed a $2.5 \times 1.5 \times 5 \text{ cm}$ (anteroposterior-cranio-caudal-transverse) mass arising at the anterior aspect of the right cerebellar hemisphere. The tumour invaded the right middle and inferior cerebellar peduncles with an exophytic component projecting in the CPA, lying within the lateral fossa of the medulla oblongata, and displacing it laterally. The lesion was well-delineated, hypo- and hyperintense respectively on T1 and FLAIR (Figure 1a). The lesion was hyperintense on diffusion-weighted-images (DWI) with slightly restricted ADC value ($0.85 \times 10^3 \text{ mm}^2/\text{s}$), in the range of high grade malignant brain tumour (Figure 1b) and enhanced slightly after

contrast (Figure 1c-1f). The tumour was located caudally to the cranial nerve VIII, but the right nerve IX was not visualized throughout its cisternal course whereas the left nerve IX was displayed. There was no extension to the internal auditory canal nor hemorrhagic area on T2 star sequence. Leptomeningeal seeding was not detected on the cerebral and spinal MRI examination. The preoperative diagnosis was astrocytoma.

The patient underwent a right suboccipital retrosigmoid craniectomy. On exposure of the right CPA, a bleeding grewish-mass with soft consistency was found to have grown exophytically from the superficial aspect of the right cerebellar hemisphere. The tumour

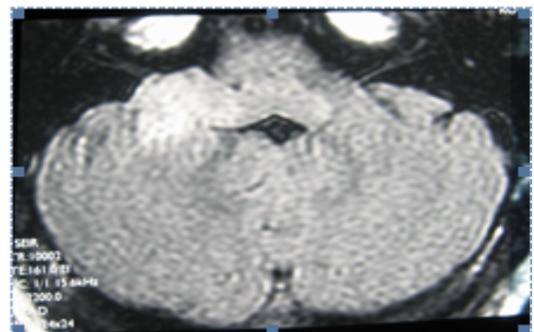


Figure 1: a) Axial FLAIR image demonstrates a well-delimited lesion at the anterior aspect of the right cerebellar hemisphere with homogeneous, moderate hyperintensity compared to normal cerebellar parenchyma. The tumour projects in the CPA and lateral medullary cisterns, is lateral-located to the medulla, compressing it. The right middle and inferior cerebellar peduncles have high signal intensity.

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completely engulfed the lower cranial nerves (IX, X, XI) and the acousticofacial bundle was lifted up but not invaded. In order to avoid any neurological injury, the resection was subtotal, with small residual tumour on the brain stem. The pathologic diagnosis was desmoplastic MB of the cerebellum (Figure 1g). In the post-operative period the patient did not develop any additional deficit and MRI examination showed the residual tumour (Figure 1h). The adjuvant therapy was started 6 weeks later. The patient received 23 Gy infratentorial irradiation, 54 Gy to local site and 30 Gy for the spinal cord and chemotherapy (carboplatin and vepeside). The latest MRI follow-up performed one year post-operation showed no additional lesion.

Discussion

MB is an embryonic neoplasm (primitive neuroectodermal tumour, PNET) (9). MBs are the most common posterior fossa pediatric tumour, with approximately 50% presenting in the first decade [1,2]. The tumour usually arises from the vermis and grows into the fourth ventricle [3]. Very rarely MBs develop in the CPA and only 38 cases have been reported in the literature to date and concerned children in only 24% of cases (9/38). Mean size at time of presentation is of 30 mm.

The symptoms tend to be shorter in duration than other posterior fossa tumours. Some clinical features that may help distinguish them

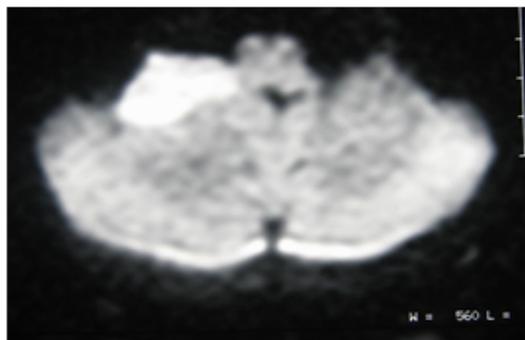
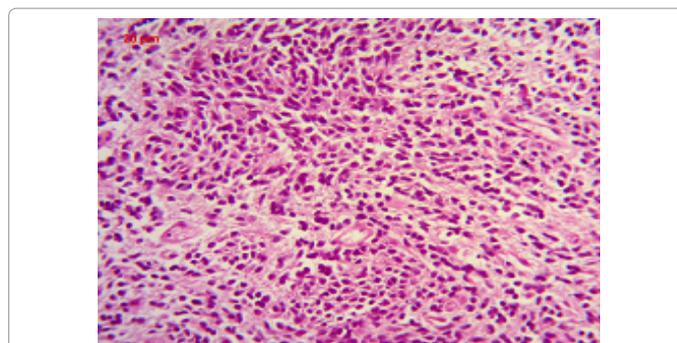


Figure 1b): DWI shows extremely high intensity in the tumour compared to normal cerebellar parenchyma.



The tumour presented large sheets with focal lobulated features, made of small cells with a high nucleocytoplasmic ratio. Cells were positive for synaptophysin.

Figure 1g): Histology of a lobulated sector of the tumour, Hematin and Eosin.

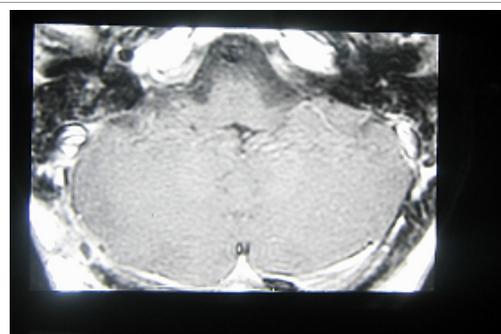
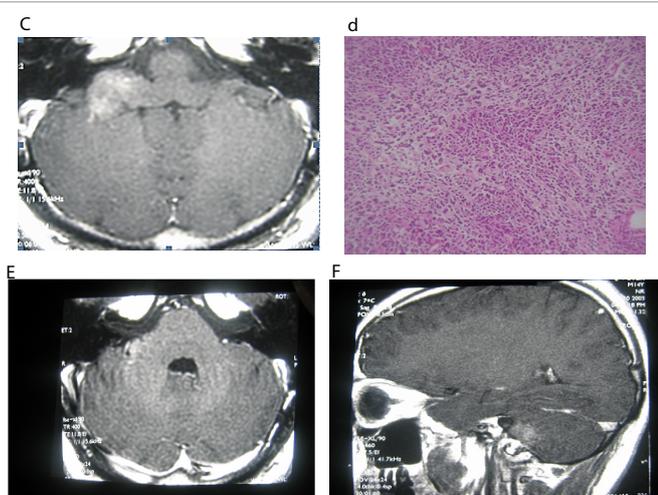


Figure 1h): Post-operative MRI: Post-contrast T1-weighted images shows the residual tumour adjacent to the brainstem.

from other CPA lesions: Vth, VI th, VII th, VIII th and lower cranial nerve involvement, and signs of cerebellar dysfunction are commonly note in CPA lesions. Hearing impairment and VII th nerve involvement is usually less common and a late feature of CPA MB. Only 5 of the 38 CPA MBs published so far presented with hearing impairment as initial symptom. Classic and desmoplastic MBs are the most common histological variants. However, most of the adult cases located in CPA were of classic type (17/26), while 9/12 reported cases in children were of classic variant [7-15]. There are no pathognomonic MRI criteria for differentiating them, and the only characteristic found is an iso- or hypointensity on T2/FLAIR sequence [15].

MB has variable appearances on MRI in both children and adults [4].

In most reported cases, CPA MBs have a homogeneous hypointensity on T1-, a heterogeneous hyperintensity on T2/FLAIR and enhance heterogeneously and strongly after contrast. Heterogeneity may be related to cystic or necrotic component and calcifications. These MRI features are those described in adult MB and don't differ from pediatric lesion, as reported recently in a series of 38 cases, not including CPA MB [16]. Another large recent series, not including CPA MBs, considered the MRI features depending on the MB variant. In most cases classic MB had a high signal on T2 and a low signal on T1. In half cases of desmoplastic MB the lesion had an isointense signal on T2. In all variants the enhancement was in most cases intense and homogeneous [6]. In the present case, the tumour was well-delineated on all sequences, finding usually reported in the pediatric population [5,17]. Although the above description apply to "typical" MBs, "atypical features" are sufficiently common (47%) to result in difficulty in confidently differentiating this tumour from others of the posterior fossa [18]. Very infrequently, MBs involving the CPA may present



Lower-cranial nerves are not identified. The acoustic-facial tract is distant from the tumour.

Figure 1 c-d-e-f): Axial (c-d-e) and Sagittal Post-contrast (f) T1-weighted images show a well-defined tumour with moderate, slightly heterogeneous contrast-enhancement.

with widening and deep invasion of the IAC, simulating an acoustic neuroma, or with a dural-based appearance, without bony involvement or hyperostosis, simulating a petrosal meningioma [11,12,19]. DWI hyperintensity has been reported in few cases of cerebellar MB in children without data on ADC values [20]. It is well known now that ADC values in PNET are low [21,22].

In the reported case, the tumour had high signal intensity on DWI and ADC value was low, in the range described in high grade tumours and PNET [21].

Our case present unusual findings. Clinical onset by acute hearing loss, and absence of edema or cystic areas are uncommon features of CPA MBs. Intra-axial tumours that may have an exophytic mass projecting into the CPA are astrocytoma and ependymoma. Occasionnaly, brain stem glioma, metastasis and lymphoma result in similar findings, but metastases are not likely children tumours. Pilocytic astrocytoma of the cerebellum may have a paravermian location and prolapse in the CPA [10]. They are usually hypointense on T1- and hyperintense on T2/FLAIR, strongly enhance after contrast and contain large cysts, whereas cysts in MB are small [10]. Rare cases of focal primary gliomas and of pilocytic astrocytoma of the brain stem can be anterolaterally exophytic, thereby occupying the CPA [23]. More rarely in children, lymphomas show similar findings as in CPA MB. Ependymoma may be radiographically indistinguishable from MB, although it more often contains recognizable foci of calcification, but have ADC value consistently higher than that of PNETs, in relation with their grading (WHO grade II) [23]. A recent study revealed that the ratio of N-Acetyl-Aspartate (NAA)/Choline and of NAA/Creatine are decreased, that the ratio of Choline/Creatine is increased, and showed a low peak of taurine [24]. If there is a lack of strict MRI features, DWI, ADC and spectroscopy findings may improve the pre-operative diagnostic accuracy of MB.

Conclusion

CPA is a very rare location of MB, especially in children. Clinical and MRI cannot help distinguish them from other CPA tumours. ADC value and Spectroscopy may assist preoperative differential diagnosis of the rare childhood CPA MB from other tumours of the same location, especially from ependymoma.

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