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Microsurgical Resection of Cerebellopontine Angle Medulloblastoma, Case Report and Review of Literature

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Abstract

Cerebellopontine angle (CPA) medulloblastomas (MB) are rare lesions with few cases previously described in the literature. We report one more case of CPA MB. The patient was a (36)-year-old female with a mass developing in the CPA. The preoperative radiological diagnosis was (hemangioblastoma) in the case. The patient was operated in Tripoli university hospital September 2020 neurosurgical department. Operated through a (retrosigmoid) approach. The intraoperative findings revealed an extra-axial tumor and the histological diagnosis was classic type of MB in the case. We review the literature and discuss pathological, radiological features and clinical follow up of CPA MB, underlining the necessity to consider MB in the differential diagnosis of CPA lesions.

Keywords

Medulloblastoma, Extra-Axial, Cerebello-Pontine (CP) Angle

1. Introduction

Medulloblastoma is a common tumor of the posterior fossa, representing 20% - 25% of all pediatric neoplasms. The tumor often occurs in the cerebellar vermis and at the apex of the fourth ventricle. It commonly presents with rapidly progressive clinical manifestations of raised intracranial pressure and cerebellar dysfunction. There are only a few reported cases of cerebellopontine (CP)-angle medulloblastoma in the literature, with most being extra-axial and occurring in the pediatric population. We present a rare case of primary extra-axial CP-angle medulloblastoma occurring in an adult [1]-[6]. Currently, the best combination of treatment modalities for medulloblastoma is aggressive surgical resection followed

closely by radiation therapy and chemotherapy, in the other hand these modalities aimed to increase both 5 years and overall survival rate, in addition, factors that indicate a better prognosis include female, old age, and lateral situation tumors.

2. Material and Methods

Left retromastoid suboccipital craniectomy with gross total excision of the lesion was performed intraoperative, the tumor was extra-axial, soft to firm, partially suckable, moderately vascular, grayish-brown, with a well-defined plane with cerebellar surface and an ill-defined plane with the brainstem. The anterior inferior cerebellar artery and basilar artery were pushed by the tumor with a well-defined plane between the tumor and the vessels. Inferiorly it had extended to the region of the foramen magnum.

3. Follow-Up

The patient was followed at our outpatient department to acquire more information. All available follow-up data began from initial diagnosis until the end of the last follow-up. Enhanced MRI was performed for postoperative evaluation or in cases of the appearance of new neurological deficits. Facial motor function was assessed using the House and Brackmann (HB) classification. The audiometric examination was evaluated using pure tone audiometry (PTA) and the auditory brainstem response (ABR).

4. Case Reports

36-year old woman, presented with three weeks history of headache, vertigo and a tendency to sway to right side while walking, mild right 7th nerve paresis of lower motor neuron type, mild right sensorineural deafness and right sided gross cerebellar signs. She had no gross papilloedema or nystagmus. CT scan showed a hyperdense, heterogeneously enhancing mass in left CPA, compressing the pons and shifting the fourth ventricle to contralateral side. On MRI, the mass was hypointense on T1WI and heterointense on T2WI with heterogeneous contrast enhancement, isointense in axial FLAIR imaging hypointense in axial ADC map (**Figure 1**) without any intracanalicular extension. Histopathological evaluation revealed highly cellular tumor proliferation with hyprechromatic nuclei, increase amount of cytoplasm manifested by rosette formation and frequent mitotic figure, area of hemorrhage and necrosis and cystic changes are seen. Immunohistochemical; study reveal tumor is negative for GFAP, EMA, and S100, while tumor is positive for synaptophysin, CD56, and NSE, with high ki67 > 80% (**Figures 2(a)-(e)**) compatible with the diagnosis of MB classic histological subtype. After surgery, the patient had (no) neurological deficit and his audiometric findings (were) improved. The postoperative MRI showed gross total resection of the tumor (**Figure 3**). The patient received whole brain irradiation, to local site and for the spinal cord. Postoperatively and after (CTX) (radiation) therapy, she

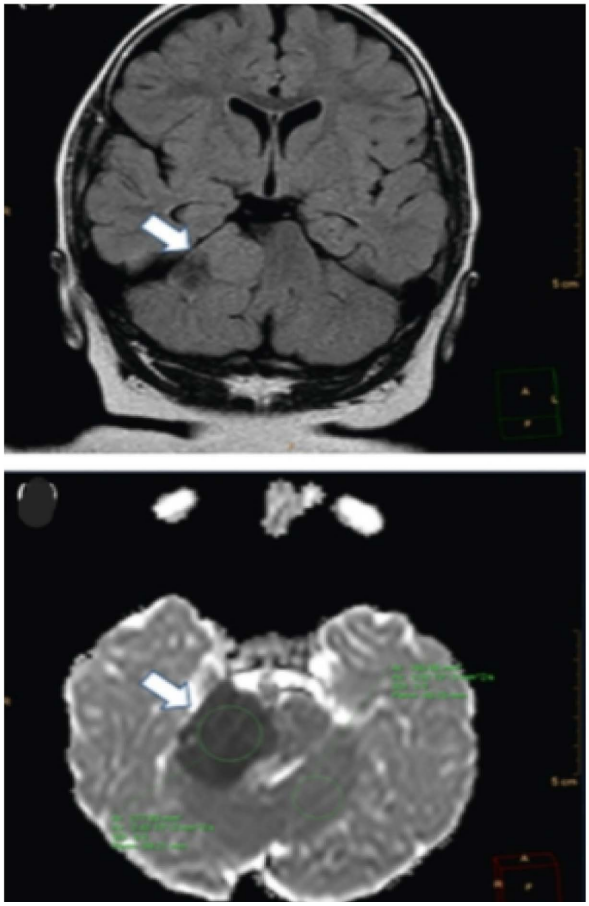

Figure 1. On MRI, the mass wasisointense in axial FLAIR imaginghypointense in axial ADC map.

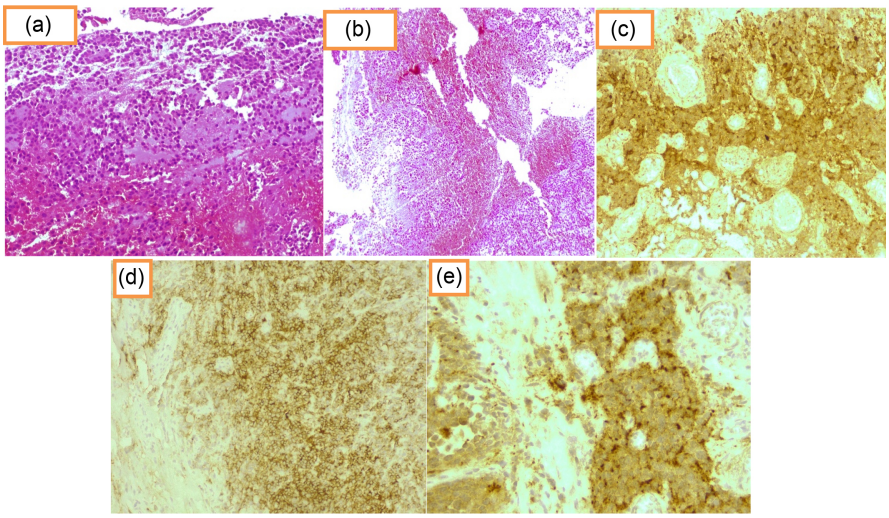

Figure 2. (a) Cellular neoplasm composed of monomorphic small cells, with round to oval hyperchromatic nuclei, minimal amount of cytoplasm. (b) Areas of hemorrhage, necrosis, and cystic degeneration. (hematoxylin and eosin stain original magnification ×400. (c) Diffuse and strong positivity of synaptophysin immune stain original magnification ×400. (d) Diffuse and strong positivity of CD56 immune stain original magnification ×400. (e) Diffuse and strong positivity of NSE immune stain original magnification ×400.

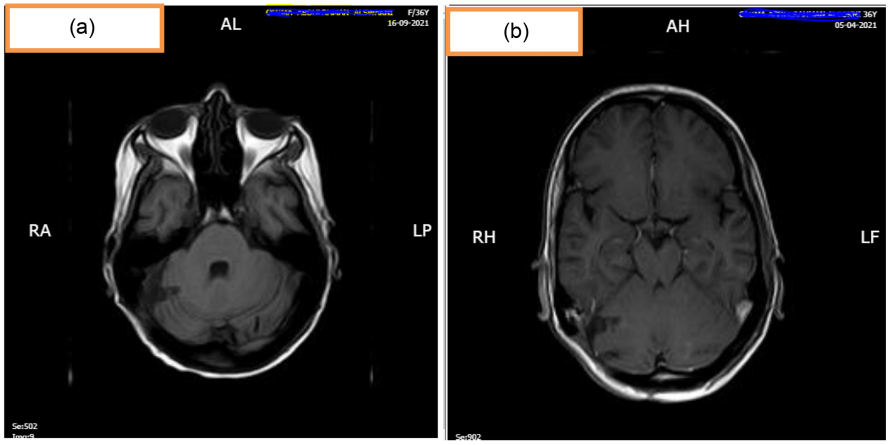

Figure 3. Post-operative MRI shows total resection of tumor with no detectable enhancement in the right CP angle promptly after surgery (a) and one-year post-operation follow-up (b).

showed improvement in neck pain, vertigo and cerebellar signs, and, after one year, in the post-operative period no neurological deterioration seen.

5. Discussion

Medulloblastoma, one of the most common paediatric brain tumors, occurs within the cerebellum, where the vermis is the most frequent site, especially in child-hood cases, but a minority of these tumors are more laterally located in the cerebellar hemispheres, especially in adults [4] [5]. A medulloblastoma with the main mass projecting into the cerebellopontine angle (CPA) is rare, and only few such cases were reported by 1993.1 A medulloblastoma presenting as an entirely exophytic CPA tumor with no apparent connection to the cerebellum or brainstem of the patient is a rarity, seldom reported in the literature [7] [8] [9] [10] [11].

Medulloblastoma may grow to occupy CPA through two pathways, lateral extension from the fourth ventricle through the foramen of loschka, or direct exophytic growth from the site of origin at the surface of cerebellum or pons [12] [13] [14] [15].

To distinguish MB from other CPA lesions, the integrity and symmetry of the internal auditory canals tend to exclude the preoperative diagnosis of vestibular schwannomas; others suggest that short duration of symptoms, the clinical absence of cranial nerve deficit and the radiological lack of bony involvement (or hyperostosis), are negative factors for preoperative diagnosis of meningioma. The dural tail sign, typical of meningioma, is not so unusual in MB and it can be associated with other benign or malignant lesions [14] [15] [16] [17].

In the present case, the patient had a preoperative diagnosis of vestibular schwannoma or CPA hemangioblastoma ass possible radiological diagnosis, subsequently; histopathological and immunohistochemistry confirm the resected lesion was medulloblastoma GIV classic type. These data confirm the lack of strict radiological and clinical diagnostic criteria for CPA MB. It is therefore necessary to consider MB in the differential diagnosis of CPA tumors.

Gross-total resection of MB is vital when it is possible, because residual tumor is associated with a worse progression free survival. In case of an extra-axial tumor the surgeon can choose to accomplish a subtotal resection in order to avoid neurological deficit and to treat residual tumor with radio surgery [6] [7] [14] [15].

In case of MB a MRI of the entire spine should be performed 10 - 14 days after surgery. In the same way, a lumbar puncture should be performed, to search for a CSF spread of the tumor. After surgery, generally the patients undergo a radiation treatment and/or chemotherapy, depending on patient' s age, extent of tumor resection, leptomeningeal dissemination and molecular factors related to the tumor [16] [17] [18] [19].

Post-operative complication up to 26% has been reported following tumor resection, in our study minimal neurological deficits appeared after we tried to preserve all neurovascular structure around the tumor, and near total recovered at the time of patient discharge.

6. Conclusion

In conclusion, our study demonstrates that MB located in the CPA is particularly rare and commonly occurs in adults. Adult patients with CPA MB may have a good prognosis, while a poor outcome is observed in some patients. Clinically it is very difficult to distinguish these neoplasms from other intrinsic CPA tumors, though they may be considered in the differential diagnosis of a mass in this region. Our findings could be helpful for developing a subsequent management plan when treating this rare disease.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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