Extra-Axial Cerebellopontine Angle Medulloblastoma in an Infant

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Established Facts
- Medulloblastoma is the most common malignant tumor of the brain in the pediatric population and commonly occurs in the midline (cerebellar vermis) and rarely at the cerebellopontine angle.
- Extra-axial cerebellopontine angle medulloblastoma is extremely rare, and only few cases have been reported in the literature.

Novel Insights
- This is the first report of an extra-axial cerebellopontine angle medulloblastoma in an infant.
- Generous reporting of usual tumors at unusual locations would help in the better understanding of lesions.

Abstract
Medulloblastoma is a common tumor of the posterior fossa, representing 20–25% of all pediatric neoplasms. It commonly occurs in the midline (cerebellar vermis) and rarely at the cerebellopontine angle. Most of them are intra-axial, and an extra-axial location of this tumor is very rare. Extra-axial cerebellopontine angle medulloblastoma is extremely uncommon and has never been reported in an infant. We report an extra-axial cerebellopontine angle medulloblastoma in a 1-year-old child.

Introduction
Medulloblastoma is the most common malignant tumor of the brain in the pediatric population. It is a solid, homogenously enhancing, midline tumor of the posterior fossa [1]. The tumor often occurs in the cerebellar ver-
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Case Report

A 1-year-old girl presented with a history of vomiting, decrease in feeding and intermittent fever of 1 month's duration, facial asymmetry with deviation of the angle of the mouth for 15 days and altered sensorium for 2 days. The patient presented to a local hospital with a Glasgow Coma Scale (GCS) score E2V2M4. The CT scan obtained demonstrated a large hyperdense lesion in the left CP angle region with obstructive hydrocephalus (Fig. 1a). After a cerebrospinal fluid (CSF) diversion procedure, the patient was referred to our hospital. On physical examination, pallor was seen along with bilateral coarse crepitations. On neurological examination, she was in an unconscious state and responded only to a painful stimulus (GCS E2V2M5). Right-sided facial nerve paresis with a weak cough reflex was also noted. MRI showed a large (60 × 43 × 41 mm) lobulated left CP angle extra-axial mass which appeared hypointense on T1-weighted images and isointense on T2-weighted images with intense homogeneous enhancement on gadolinium administration (Fig. 1b–d).

The lesion caused severe compression and displacement of the underlying cerebellar hemisphere, brainstem and 4th ventricle towards the right side with upstream hydrocephalus. There was widening of the left internal acoustic canal. A differential diagnosis of primitive neuroectodermal tumor/teratoid rhabdoid tumor was considered.

A left retromastoid suboccipital craniectomy with gross total excision of the lesion was performed. Intraoperatively, the tumor appeared hyperechoic on ultrasound, and 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. She did not make a good postoperative recovery and was put on ventilator support. Histopathological examination of the specimen showed densely packed tumor cells arranged in pseudorosettes as well as in a trabecular pattern. Individual tumor cells were round to oval, having hyperchromatic nuclei with scanty amounts of cytoplasm (Fig. 2). Immunohistochemistry staining demonstrated the tumor cells were strongly positive for synaptophysin. Based on histopathological examination and immunohistochemistry, a diagnosis of medulloblastoma was established. No secondary wound complications were witnessed. However, the patient remained in a low GCS state, on ventilator support postoperatively and expired on postoperative day 20. Adjuvant CT/radiotherapy was withheld in view of the persistent poor general condition in the postoperative period.

Fig. 1. a Axial contrast CT scan. b Axial T1-weighted MRI. c Axial T2-weighted MRI. d Axial contrast MRI.
Discussion

Medulloblastoma is predominantly a childhood tumor that almost always presents intra-axially. The location in the CP angle region is very rare, and nearly 40 cases have been reported in literature so far, with the largest series by Jaiswal et al. [10] where they reported 14 cases. The peak incidence is found during the first decade, and the median age at diagnosis is 5–7 years. The tumor has a male preponderance, with a sex ratio of 2:1. The age of our patient and the extra-axial location of the tumor made our case quite different from those reported in the literature.

The cell of origin of medulloblastoma is still controversial. It has been suggested that a medulloblastoma may originate either from germinal cells or their remnants situated at the end of the posterior medullary velum from remnants of the external granular layer [11, 12]. Kadin and Rubinstein [13] suggested that medulloblastomas arise from germinal cells (or their remnants) anywhere along their migratory path, and because the migratory process normally proceeds in a lateral direction, there would be a relatively higher frequency of laterally situated tumors in adults. Their development in the CP angle may be from the remnants of the external granular layer in the cerebellar hemisphere, including the flocculus which faces the CP angle.

On the other hand, some authors proposed that medulloblastomas may originate from a proliferating residue of the lateral medullary velum [14, 15].

Regarding the clinical manifestations, no specific features have been ascribed to CP angle medulloblastomas. Infants with hydrocephalus may present with irritability, lethargy or progressive macrocrania [16]. Involvement of the 5th, 6th, 7th, 8th, and lower cranial nerves and signs of cerebellar dysfunction are commonly noted in CP angle lesions [17]. However, there are some characteristics that may help distinguish them from other CP angle lesions, mainly the most common tumor, acoustic neuroma, followed by meningioma, primary cholesteatomas and epidermoid tumors [2, 18]. Facial and vestibulocochlear nerve involvement is usually less common and a late feature of a CP angle medulloblastoma; however, this feature has even been reported as initial symptom in a few cases [10]. Early onset of progressive cerebellar signs and gait ataxia may indicate an axial origin of the tumor, whereas positional nystagmus may be an early sign suggestive of an acoustic schwannoma [10]. Our case presented with signs of raised intracranial pressure with involvement of the 7th and lower cranial nerves.

Medulloblastomas are known to metastasize through the CSF to the spinal canal, leptomeninges, and supratentorial regions. Metastasis in medulloblastomas varies

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**Fig. 2.** Histopathological examination image. HE stain. ×100. Inset ×400.
between 38 and 60% in various series, with the spinal canal being the commonest site with approximately 58% [3, 19]. However, spinal metastasis from CP angle medulloblastoma has not been reported to date.

CT scan or MRI are the main diagnostic options for pediatric medulloblastomas. On CT scan, medulloblastomas are usually hyperdense lesions with homogeneous contrast enhancement. They may be partially cystic and with minimal calcification. On MRI, they are iso-or hypointense on T1-weighted images, hyperintense on T2-weighted images and intensively enhanced after gadolinium injection. However, about 10–15% of medulloblastomas are not enhanced on contrast MRI scans [20]. Britton [21] reported that the presence of a normal internal auditory canal is an important radiological finding distinguishing this lesion from an acoustic schwannoma. In our case the tumor was lobulated, well defined, and showed intense contrast enhancement with widening of the internal acoustic canal.

The treatment of CP angle medulloblastoma is still not well established because of the small number of reported cases [2, 4, 7, 10]. The therapeutic management of medulloblastoma consists of an as-radical-as-possible excision of the tumor mass, followed by irradiation. In children, chemotherapy is also applied, though routine chemotherapy in adult patients is still a matter of controversy. Ventricular shunting, before or after surgery, may be required in cases with hydrocephalus, with some cases being able to avoid shunting after primary decompression and establishment of CSF pathways.

The survival rate of CP angle medulloblastoma is not significantly different from their vermian counterparts. A total of 30% of 5-year survival has been reported for medulloblastoma of this location after surgery and radiotherapy [22, 23]. Chemotherapy has also been combined with radiotherapy with improvement in some cases [24].

Finally, the extra-axial site of this tumor is extremely rare but must be considered in the differential diagnosis of extra-axial CP angle lesions. Surgical resection followed by chemoradiotherapy offers improved survival. However, early diagnosis is absolutely necessary for a better outcome. Generous reporting of usual tumors at unusual locations would help in the better understanding of lesions.

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Ethics Statement

Patient consent was obtained.

Disclosure Statement

The authors have no conflict of interest.

References


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