CASEN REPORT

CP ANGLE MEDULLOBLASTOMA WITH SUPRATENTORIAL DROP METS

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Medulloblastoma, is an undifferentiated neuroepithelial tumour of embryonic origin". It is an uncommon central nervous system tumour, and adult-onset is even rarer. It could form in an unusual or unconventional location, like the cerebellopontine angle (CPA), and such tumours don't frequently show supratentorial extension. Less than 2 per million persons are affected each year, with children being affected ten times more than adults. **Case Presentation:** We present a 17-year-old male with complaints of occipital headache and having trouble walking for three months. Right Cerebellopontine (CP) angle lesion with ipsilateral mass effect over the cerebellum was seen on MRI brain with contrast. The tumour was excised by right sub-occipital retro mastoid craniotomy and histopathology revealed medulloblastoma. Post-operatively patient underwent chemo-radiotherapy. Craniospinal MRI was done to look for any dropmets. Recovery was uneventful. Two years later, he complained about headaches and generalized tonic-clonic seizure, MRI brain with contrast revealed a lesion with extra-axial components with a mass effect in the right temporal lobe. Right temporal craniotomy was done and the tumour was excised and histopathology showed a round blue cell tumour (medulloblastoma). **Conclusion:** We report a rare case of medulloblastoma located at the CP angle along with supratentorial metastasis 2-years after treatment.

Keywords: Medulloblastoma; CP Angle tumour; Supratentorial metastasis

Citation: Javed MA, Saba AU, Ahmed N, Habib S, Razzaq S. CP angle medulloblastoma with supratentorial drop mets. J Ayub Med Coll Abbottabad 2024;36(2):443–6.

DOI: 10.55519/JAMC-02-12611

INTRODUCTION

Medulloblastoma accounts for less than 1% of adult primary brain neoplasms which frequently originates in cerebellar vermis. Cerebellopontine (CP) angle medulloblastomas are extremely rare. The majority of them are intra-axial. This tumour's extra-axial location is incredibly rare.[1] Children are afflicted 10 times more than adults, and it affects fewer than two persons per million annually. It is the second-most prevalent kind of paediatric brain tumour. Less than 2% of CNS cancers occur in adulthood.

Males (62% vs. 38% of females) have a greater rate of new cases of paediatric medulloblastoma. Most medulloblastoma patients (around 40%) have their diagnosis before the age of five, followed by 31% between the years of 5 and 9 and 18.3% between the ages of 10 and 14. Lastly, 12.7% fall between the ages of 15 and 19. Since there have only been 33 incidences of medulloblastoma in the CP angle alone, it is unlikely that they develop extra axially.[2]

The typical signs and symptoms are irritability, lethargy, nausea, vomiting, morning headaches, anorexia, diplopia, and behavioural changes. Truncal and appendicular ataxia are more frequently caused by midline cerebellar tumours. Investigations and a thorough clinical evaluation are used to diagnose medulloblastoma. These consist of MRIs, blood tests, and eye examinations to look for papilledema. The primary method for medulloblastoma diagnosis continues to be MRI of the brain and spine.

Chemo-radiotherapy may be administered separately or in combination after aggressive surgery. The recurrence of the tumour might be local or metastatic.

Infants, high-risk patients, and adults have a poor prognosis.[3] The patients with the worst prognosis are those who experience recurring disease after receiving initial treatment.[3] Metastasis typically occurs in the spinal column, posterior cranial fossa, bone, and the supratentorium.[3] However, Supratentorial intraventricular medulloblastoma metastases are not common (6% of cases).[4]

Our purpose is to report a rare presentation of an extra-axial medulloblastoma in the CP angle along with Supratentorial metastasis 2 years following therapy.

CASE PRESENTATION

A 17-year-old male complained of having an

occipital headache with trouble walking for 3 months. At the time of the presentation, he was well-oriented in time, place, and person with a GCS of 15/15. Cranial nerve examination was unremarkable; however, the patient had positive cerebellar signs on the right-side including intension tremors, positive finger nose test, and heel shin test.

MRI brain showed a contrast enhancing lesion in the right CP angle with peri-lesion oedema and ipsilateral mass effect over the cerebellum (figure 1 and 2). Right side Sub occipital retro mastoid craniotomy was done and the tumour was excised.

Histopathology showed sheet of small round blue cells showing syntical arrangement of densely packed undifferentiated cells. The individual neoplastic cells have pleomorphic, hyperchromic nuclei, very high N/C ratio and ill-defined cell boundaries. Nuclear angulation and moulding are prominent. Numerous perivascular pseudo-rosettes are seen. Many mitoses with apoptotic bodies is seen, characteristics of medulloblastoma as demonstrated in (Figure-3). The patient underwent chemo radiotherapy (vincristine during radiation therapy, and then cycles of vincristine, cisplatin, and lomustine (CCNU) and using a Cobalt-60 teletherapy system, the patient received craniospinal irradiation at a dose of 60 Gy on the posterior fossa with 36 Gy onto the spinal axis) along with a whole spine MRI for drop metastasis which was unremarkable and the patient recovered completely. The patient did not appear for follow-up.

After 2 years patient developed a headache and generalized tonic-clonic fits, An MRI brain T-2

Weighted was done, which showed a lesion in the right temporal lobe with an extra-axial lesion with mass effect (Figure 04). The tumour was excised by a right temporal craniotomy. Histopathology showed sheet of cells having round to oval nuclei, inconspicuous nucleoli and scanty cytoplasm. There is syncytial arrangement of densely packed undifferentiated cells along with homer wright rosettes formation. Mitotic figures are also noted characteristic of medulloblastoma. On immunohistochemistry synaptophysin is positive in tumour cells. Relapsing medulloblastoma has the worst prognosis in this case. The patient did not follow-up post operatively.

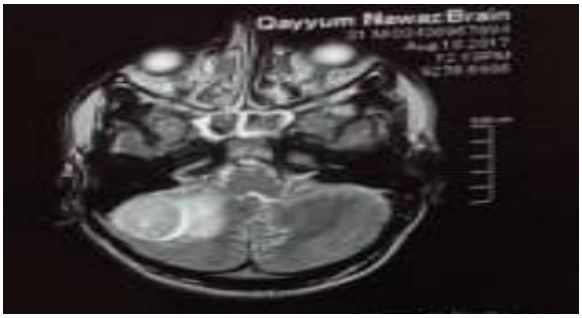

Figure-1: MRI T2-weighted image showing mixed density right CPA lesion with peri-lesion oedema and mass over adjacent cerebellum.

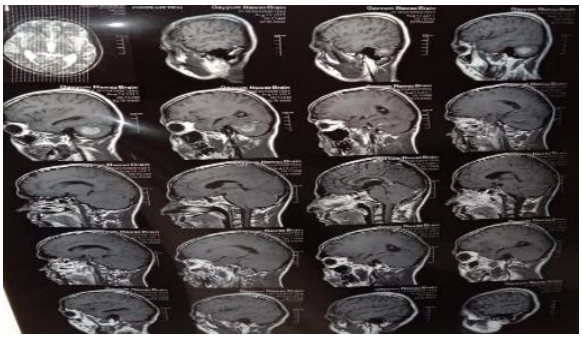

Figure-2: contrast enhancing right sided CPA lesion with mass effect.

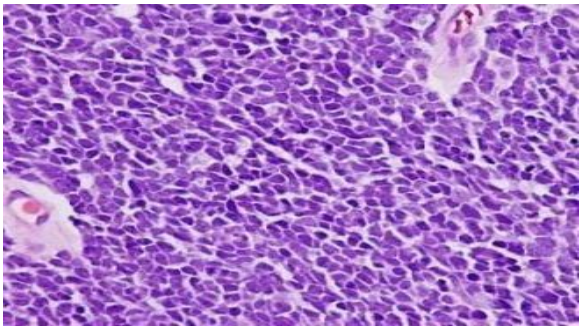

Figure-3: histopathology showing round blue cell tumour characteristic of medulloblastoma

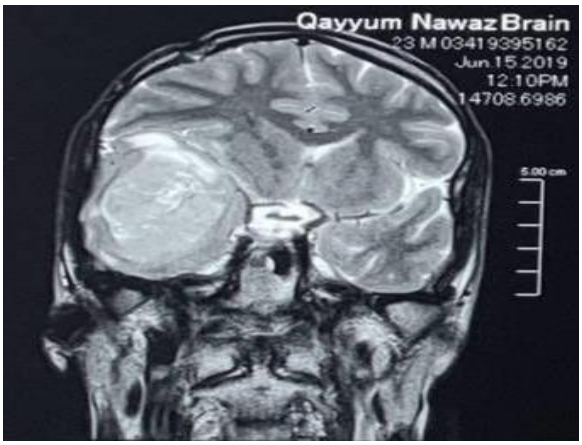

Figure-4: MRI T2-Weighted image showing extra-axial mixed densitylesion in right temporal lobe with mass effect.

DISCUSSION

The Cerebellopontine angle location accounts for 5–10% of all intracranial tumours. Vestibular schwannomas, meningiomas, and other tumours represent a majority of mass lesions in the CPA, and a large variety of atypical lesions are also found in the CPA. However, CPA medulloblastomas are relatively rare lesions[4]. Medulloblastoma accounts for 15–20% of all CNS neoplasms. It was first characterized as a primitive origin tumour growing in young children's posterior fossa by Bailey and Cushing in 1925.[5] It typically results from the disruption of several signalling pathways, including the WNT and hedgehog pathways, in the cerebellum's external granular layer. Every year, 0.5% of children under the age of 15 experience it. The traditional triad of lethargy, vomiting, and morning headaches is a common presentation; nevertheless, these symptoms are generic.There may also be cerebellar symptoms such as dysmetria, limb ataxia, or truncal ataxia. If there are facial or bulbar palsies, brainstem invasion is suspected; nevertheless, sixth nerve palsy is typically caused by hydrocephalus.[6] MRI scans of medulloblastomas frequently show hypo intense to hyperintense to gray matter on T1, show evidence of Gadolinium enhancement, and may show iso- or hyperintense to gray matter on T2. Establishing a histologic diagnosis, removing as much of the tumour mass as possible, and assisting in the hydrocephalus's resolution are the objectives of the surgery. Chemotherapy and postoperative radiation therapy are used to eradicate any remaining illness. CSF cytology and brain and spine magnetic resonance imaging are used in the patients' follow-up to look for metastases or residual disease.[6]

A localized or metastatic tumour recurrence is common. Spinal metastasis is more common compared to supratentorial metastasis, which typically affects the frontal lobe, the subfrontal region, and the cribriform plate, close to the orbital roof.[7] The most common location was found to be the posterior fossa (56%), subsequently followed by the bone marrow (25%), according to patterns of recurrence described by Kunscher *et al*.(2001).[7,8] The majority of supratentorial recurrence cases, however, have been documented in the initial two years after the initial surgery. Emmenzuel *et al*. (2006) described the metastasis of medulloblastoma in the frontobasal area twenty-one years after the complete resection of cerebellar medulloblastoma.[7,9] According to a study by Park *et al*. (1983), metastasis in the supratentorial region was reported in only 14.6% of patients, and only 12.5% of patients had spinal cord metastases.[10] Only 7% of the 768 medulloblastoma patients in a McNeil *et al*.(2002) SEER update had a Supratentorial anatomic site, and only 3% had both a Supratentorial and an Infratentorial location.[7,11] Only 1–2% of patients with medulloblastoma exhibit extra neural metastases at presentation. Nearly 83% ofpaediatric tumours are midline, compared to 49% of adult tumours that are lateral.[3] The prognosis is better in cases of metastasis when the recurrence is limitedto one site and occurs later than the first diagnosis.

Metastasis typically occurs at the posterior fossa, the spine, the bone, and the supratentorium.[3] The largest series on recurrent medulloblastoma cases, most of which involved patients under the age of 16, was published by Ramaswamy et al. in their article.[12] When compared to local relapse, they discovered that distant metastasis was the most frequent type of tumour relapse. Two adults, aged 31 (case 1) and

Twenty (Case-2), were reported by Kumar et al.[3] to have metastases in the supratentorial region at 3.5 years and 11 months, respectively, following the complete resection of the medulloblastoma in the posterior fossa.

Patients with medulloblastoma still have a poor prognosis, and the prognosis of relapsing medulloblastoma is poor and treatment is often difficult, especially because irradiation for brain stem invasion has a poor prognosis as in the present case.[4] The purpose of this study is to report a rare presentation of grade IV medulloblastoma at CP angle with drop metastasis in the Supratentorial region after 2 years of primary tumour excision.

CONCLUSION

We conclude that medulloblastoma can occur at rare locations such as CP angle and metastasis can occur in the Supratentorial region rarely. Therefore, it is advised that children with medulloblastoma continue with theirnormal treatments and 6-month follow-ups for a timely diagnosis. children with medulloblastoma should always be monitored for signs of tumour recurrence or brain metastasis.

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Submitted: November 1, 2023	*Revised: April 28, 2024*	*Accepted: April 29, 2024*

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