

Bilateral Cerebellar Medulloblastomas: A Case Report And Review Of Literature

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Citation

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Abstract

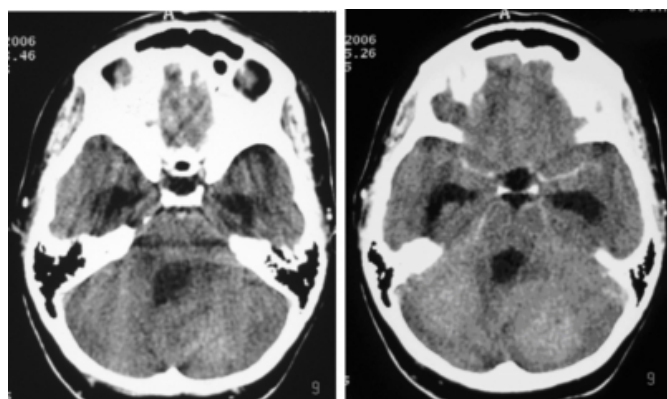
Medulloblastomas are common midline posterior fossa tumors in children, accounting for almost 30% of all pediatric brain tumors. In adults these tumors occur in the cerebellar hemisphere. Medulloblastomas occurring bilaterally in an adult patient is very rare. Present article includes a case report of one such patient. Relevant literature is also reviewed.

CASE REPORT

A 24 year old, right handed female presented to medicine outpatient clinic with complaints of painful neck movements, headache, giddiness and double vision of two months duration. Clinical examination revealed severe papilledema. The motor and sensory systems were normal and there were no cerebellar signs. She had neck stiffness and a positive Kernigs sign. There was no history of fever, convulsions, vomiting or contact with tuberculosis. India being an endemic area for tuberculosis, a provisional diagnosis of tubercular meningitis was kept. Computed Tomographic scan of brain was reported as showing moderate dilatation of ventricles. A diagnostic lumbar puncture did not reveal any infective pathology. A second close look at the CT scan however showed mild hyperdense enhancing areas, close to the tentorium cerebelli (Figure 1,2) and a MRI with contrast was then ordered.

Figure 1

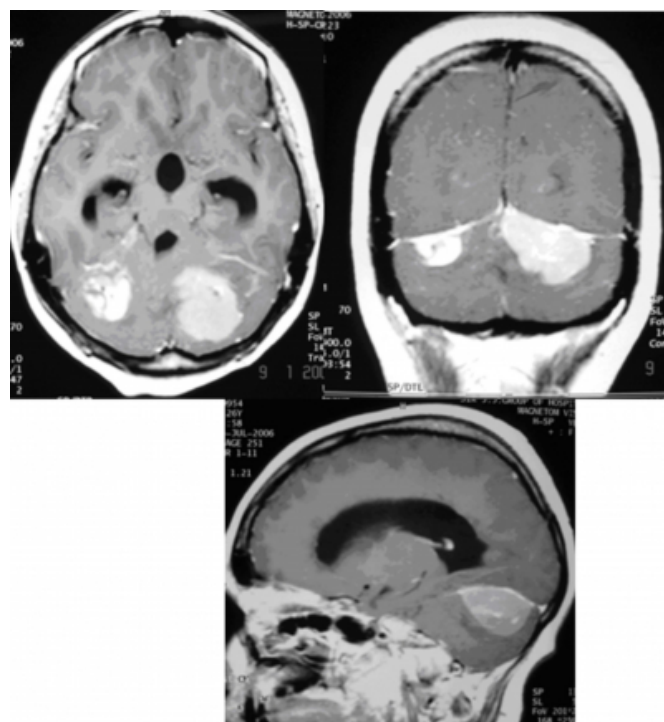
Figure 1 & 2



MRI showed two well defined extra axial masses which were hypo to isointense on T1 weighted images and isointense on T2 weighted images, with mild perilesional edema and moderate but homogenous contrast enhancement. Both the masses appeared to be arising from the tentorial leaves with broad base, causing buckling of the cerebellar folia. Tonsillar herniation and obstructive hydrocephalus were also noticeable. A diagnosis of bilateral meningiomas was thus made. (Figure 3, 4, 5)

Figure 2

Figure 3, 4 & 5



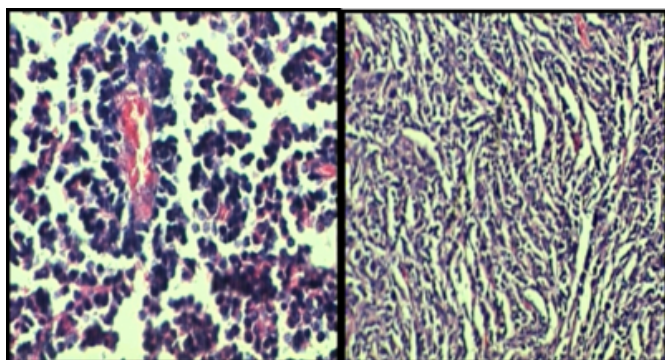
She underwent a ventriculo-peritoneal shunt followed by excision of both the tumors in sitting position.

Intraoperatively it was noted that both the tumors were separate from each other. They were fleshy, vascular and had no direct attachment to the dura. Both the tumors seemed to be arising from the superior surface of the cerebellum and were amenable to complete excision. Postoperative period was uneventful.

Histopathology of both tumors revealed highly cellular tumor chiefly comprising of round cells arranged in sheets and nodules. At few places the cells were arranged in cords and separated by collagen. Nuclei were round to oval with open chromatin and evidence of atypical mitoses and necrosis. (Figure 6, 7). Special staining with reticulin was suggestive of desmoplastic medulloblastoma. Both the tumors stained positive for synaptophysin and chromogranin and negative for EMA, GFAP and MIC-Z on immunohistochemical staining.

Figure 3

Figure 6 & 7



She received craniospinal irradiation postoperatively and is now on regular follow up with us.

DISCUSSION

Medulloblastoma is a primitive neuroectodermal tumor arising in the posterior fossa. It most commonly affects children with three fourths of patients being under nine years of age. It accounts for 20-25% of all pediatric brain tumors (1).

Cell of origin of medulloblastoma is still controversial. It has

been speculated to arise from cells of external granular layer of cerebellum or from undifferentiated cells of posterior medullary velum. Various factors like Insulin like growth factor – I, WNT and sonic hedgehog are linked to the tumor formation (2).

Location of the tumor within the posterior fossa depends on the age of the patient. These are predominantly located in the midline vermis in children. Adults are likely to have heterogeneous cerebellar hemisphere tumors, which is due to a greater prevalence of desmoplastic variant.

Medulloblastomas are known to spread by CSF pathways. McFarland noted that metastatic nodules caused by CSF seeding occurred mainly in spinal canal due to predominantly downward flow of CSF from cisterna magna (3). Gupta et al has reported a case of multicentric medulloblastoma where the metastatic nodules were located in the third ventricle (4). Glial tumors, especially high grade gliomas, are known to have multicentric origin (5).

Bilateral medulloblastomas arising separately from both cerebellar hemispheres are very rare. Malheiros et al has mentioned of one case in his article being bilateral (6). The treatment strategy for these patients should be same as in routine medulloblastoma cases, which includes postoperative craniospinal irradiation. Whether prognosis of these patients is any different from the routine medulloblastoma patients is not clear.

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